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BOOK OF ABSTRACTS

20TH INTERNATIONAL CONFERENCE ON BEHÇET'S DISEASE

18-20 SEPTEMBER 2024
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SUPPLEMENT I

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PREPARATION AND FORMATTING OF ALL SECTIONS OF MANUSCRIPTS, COVER LETTERS, AND SUPPLEMENTARY MATERIALS

The Journal only accepts manuscripts submitted online at http://www.mjrheum.org/submit.

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Clinical and Epidemiological Characteristics of Adamantiades-Behçet Disease in a Moroccan Cohort	Tijeni Tak Tak, Casablanca, Morocco Co-Authors: Andreas Altenburg, Athanassios Kyrgidis, Richard J. Angkasa, Christos C. Zouboulis
Clinical and Epidemiological Characteristics of Adamantiades-Behçet Disease: A Comparative Study of German and Moroccan Patients	Andreas Altenburg, Staedtisches Klinikum Dessau, Brandenburg Medical School Theodor Fontane and Faculty of Health Sciences Brandenburg, Dessau, and German Registry of Adamantiades-Behçet Disease, Dessau, Germany Co-Authors: Tijeni Tak Tak, Richard J. Angkasa, Athanassios Kyrgidis, Christos C. Zouboulis
Splanchic Venous Thrombosis in Behcet's Disease: A Monocentric Study of 22 Patients	Soukaina Oumlil, University hospital Mohammed VI, Cadi Ayyad university, Marrakech, Morocco Co-Authors: Jaouad Yousfi, Mouna Zahlane, Laila Benjilali, Lamiaa Essaadouni
Management of Severe Adamantiades-Behçet's Disease with Hughes-Stovin Syndrome, Cardiac Thromboses, and HIV Infection using Pegylated Interferon Alpha- 2a: A Case Report	Andreas Altenburg, Staedtisches Klinikum Dessau, Brandenburg Medical School Theodor Fontane and Faculty of Health Sciences Brandenburg, Dessau, and German Registry of Adamantiades-Behçet Disease, Dessau, Germany Co-Authors: Lothar Krause, Michael Schirmer, Christos C. Zouboulis
Insights into Adamantiades-Behçet Disease in Germany: A Focus on Prevalence, Treatment Patterns, and Clinical Characteristics	Michael Schirmer, Medical University of Innsbruck, Austria and German Registry of Adamantiades-Behçet Disease, Austria Co-Authors: Andreas Altenburg, Athanassios Kyrgidis, Anna Kolp, Christos C. Zouboulis
Behcet's syndrome comorbid with bipolar disorder: A case report	Oumaima El Kaddouri Co-Authors: Salma Boustani
An angio-behçet in an unusual location	Omayma Ezzarouki, Centre Hospitalier Universitaire d'Oujda, Morocco Co-Authors: Zahida Aqodad, Houda Bachir, Ikram Sadki, Habiba Bennesser Alaoui, Siham Hamaz, Khalid Serraj
Neuro-Behçet: 4 pseudo tumoral forms	Echchilali Khadija, Ibn Rochd University Hospital Center, Morocco Co-Authors: Zeroual Chaïmaâ, Mourabit Safaâ, Moudatir Mina, El Kabli Hassan
Neurological manifestations of Behçet's disease	Moudatir Mina, Ibn Rochd University Hospital Center, Morocco Co-Authors: Mourabit Safaâ, Zeroual Chaïmaâ, Benzakour Merieme, Echchilali Khadija, Alaoui Fatima-Ezzahra, El Kabli Hassan, Benamour Saïda
Imaging in the neuro-Behçet: Basis of diagnosis and follow-up	Echchilali Khadija, Ibn Rochd University Hospital Center, Morocco Co-Authors: Tamir Kawtar, Zeroual Chaïmaâ, Mourabit Safaâ, Moudatir Mina, El Kabli Hassan

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Particularities of the Juvenile Form of Behçet's Disease	Lakrafi Yassir, University Hospital Ibn Rochd, Morocco Co-Authors: Barakat Leila, Moudatir Mina, Benzakour Merieme, Echchilali Khadija, Alaoui Fatimzahra, El Kabli Hassan
Clinical features and outcomes of vitreous hemorrhage associated with Behçet uveitis	Moncef Khairallah, Department of Ophthalmology, Fattouma Bourguiba University Hospital, Faculty of Medicine, University of Monastir, Tunisia Co-Authors: Nesrine Abroug, Tarek Dridi, Mootez Mourali, Sanaa Ahbeddou, Bechir Jelliti
Macular hole complicating Behçet's uveitis: a report of 5 cases	Nesrine Abroug, Department of Ophthalmology, Fattouma Bourguiba University Hospital, Faculty of Medicine, University of Monastir, Tunisia Co-Authors: Mootez Mourali, Omar Berbich, Melek Kechida, Bechir Jelliti, Moncef Khairallah
Frosted branch angiitis as a presenting feature of Behçet's uveitis	Nesrine Abroug, Department of Ophthalmology, Fattouma Bourguiba University Hospital, Faculty of Medicine, University of Monastir, Tunisia Co-Authors: Mootez Mourali, Zina Berbich, Sana Khochtali, Bechir Jelliti, Moncef Khairallah
Binocular diplopia revealing angioBehçet	Morad Chiguer, Department of Internal Medicine B, Mohammed V Military Training Hospital, Rabat, Morocco Co-Authors: Driss Said, Amal Charef, Rime Lemouaden, Assia Kadiri, Jamal Fatihi
Concomitant discovery of Behçet's disease, pulmonary tuberculosis and viral hepatitis C: Therapeutic strategy and management challenges	Imane Zahrou, Department of Internal Medicine Chu Oujda, Morocco Co-Authors: Siham Hamaz, Zahida Aqodad, Habiba Allaoui, Houda Bachir, Khalid Serraj
What a simple headache can hide	Imane Zahrou, Department of Internal Medicine CHU Oujda, Morocco Co-Authors: Siham Hamaz, Zahida Aqodad, Ikram Sadki, Habiba Allaoui, Houda Bachir, Khalid Serraj
Neuro-Behcet's disease	Loubna Tayebi, Internal medicine Co-Authors: Chadyne Taouil, Jaouad Yousfi, Laila Benjilali, Mouna Zahlane, Lamiaa Essaadouni
Infarct during Behçet's Disease: A Case Report	Farah Ahallat, Military Hospital Mohammed V Rabat, Morocco Co-Authors: Nawal Sahel, Bilal Talamoussa, Meryem Zaizaa, Zineb El bougrini, Nisrine Bahadi, Oumama Jamal, Adil Rkiouak, Youssef Sekkach
Cluster analysis of phenotypes of patients with Behçet's syndrome from a Tunisian population	Sanaa Ahbeddou, Noor Rabat Maroc, Morocco Co-Authors: Melek Kechida, Imen Ksiaa, Ansar Meftah, Imene Chaabene, Syrine Daadaa, Ines Khochtali, Bechir Jelliti, Moncef Khairallah
Behçet's Disease: The Pitfall of Unrecognized Associations	Zahida Aqodad, Centre Hospitalier Universitaire d'Oujda, Morocco Co-Authors: Siham Hamaz, Houda Bachir, Ikram Sadki, Habiba Bennesser Alaoui, Khalid Serraj
Fistulization of a subclavian artery aneurysm in the left main bronchus in the setting of Behçet's disease.	Omayma Ezzarouki, Centre Hospitalier Universitaire d'Oujda, Morocco Co-Authors: Zahida Aqodad, houda Bachir, Ikram Sadki, Siham Hamaz, Habiba Bennesser Alaoui, Khalid Serraj
Dermatological manifestations during Behçet's disease: a series of 60 cases	Morad Chiguer, Department of Internal Medicine B, Mohammed V Military Training Hospital, Rabat, Morocco Co-Authors: Amal Charef, Rime Lemouaden, Assia Kadiri, Yassine Oualehsine, Jihane Benhammou, Fatimazahra Boucham, Fadoua Mekouar, Naoual Elomri, Mohammed Jira, Jamal Fatihi
Late-onset Adamantiades-Behçet's disease	Christos C. Zouboulis, Staedtisches Klinikum Dessau, Brandenburg Medical School Theodor Fontane, Germany Co-Authors: Aristeidis G. Vaiopoulos, Mihael Samarkos, Meletios A. Kanakis, Georgios Vaiopoulos, Phedon G. Kaklamanis
Adamantiades-Behçet's disease and COVID-19	Christos C. Zouboulis, Staedtisches Klinikum Dessau, Brandenburg Medical School Theodor Fontane, Germany Co-Authors: Anthony Lim, Andreas Altenburg
Prepapillary vitreous exudate: a rare, but typical feature of Behçet disease	Sana Khochtali, Department of Ophthalmology, Fattouma Bourguiba University Hospital, Faculty of Medicine, University of Monastir, Tunisia Co-Authors: Wijden Nabi, Zina Berbich, Sana Sayadi, Hager Ben Amor, Moncef Khairallah

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A 21-year-old male with intracardiac thrombus	Farah Ahallat, Military Hospital Mohammed V Rabat, Morocco Co-Authors: Nawal Sahel, Zineb Elbougrini, Zaizaa Meryem, Talamoussa Bilal, Nisrine Bahadi, Oumama Jamal, Adil Rkiouak, Youssef Sekkach
Cyclophosphamide and Neuro-Behçet's Disease: An Old Therapy Still Effective	Zahida Aqodad, Oujda university hospital center, Morocco Co-Authors: Samia Sabri, Houda Bachir, Ikram Sadki, Habiba Alaoui Bennesser, Siham Hamaz, Khalid Serraj
Arterial involvement in Behçet's disease: Aneurysms and thrombosis	Maysam Jridi, Rabta University Hospital, Morocco Co-Authors: Tesnim Zakraoui, Tayssir Ben Achour, Ines Naceur, Fatma Said, Monia Smiti
Prevalence of Behçet's disease in 218 patients with uveitis, internal medicine consultant in a private outpatient setting, Settat, Morocco	Said El Kettani, Liberal
Hughes Stovin syndrome in Behçet's disease	Maysam Jridi, Rabta University Hospital Center, Morocco Co-Authors: Tesnim Zakraoui, Tayssir Ben Achour, Ines Naceur, Fatma Said, Monia SmitiX
Interest in the PNN/lymphocyte ratio: new marker of inflammation during vascular thrombosis in Behcet's disease?	Khibri Hajar, Ibn Sina University Hospital Center, Faculty of medecine and pharmacy of Rabat, Mohammed V university of Rabat, Morocco Co-Authors: Chadli Sarra, Fari Safae Firdaous, Messaoud Sara, Obtel Majdouline, Hajji Oumayma, Chhih Yasmine, Mouattassime Naima, Ammouri Wafae, Maamar Mouna, Harmouche Hicham, Adnaoui Mohammed, Tazi Mezalek Zoubida
Benefit and risk: the anti TNF alpha dilemma in ocular Behçet's treatment	Meryem Zaizaa, Mohammed V Military Hospital, Rabat, Morocco Co-Authors: Farah Ahallat, Nissrine Bahadi, Oumama Jamal, Zineb El Bougrini, Ilyass El Kassimi, Bilal Talamoussa, Sahel Nawal, Adil Rkiouak, Youssef Sekkach
Assessing Treatment Adherence in Tunisian Behçet's Disease Patients	Maysam Jridi, Rabta University Hospital Center, Morocco Co-Authors: Donia Dridi, Tasnim Zakraoui, Tayssir Ben Achour, Ines Naceur, Fatma Said, Monia Smiti
Prevalence of bundle retinal nerve fiber layer defects in patients with new-onset Behçet Uveitis	Moncef Khairallah, Faculty of Medicine, Monastir, Tunisia Co-Authors: Omar Miladi, Hager Ben Amor, Omar Berbich, Imen Ksiaa, Béchir Jelliti
Relapse in Behçet's Disease: prevalence and associated factors	Maysam Jridi, Rabta University Hospital Center, Morocco Co-Authors: Donia Dridi, Tayssir Ben achour, Ines Naceur, Tesnim Zakraoui, Fatma Said, Monia Smiti
Anxiety and Depression Prevalence in Tunisian Behcet's Disease Patients	Tayssir Ben Achour, Rabta University Hospital, Tunis, Tunisia Co-Authors: Donia Dridi, Tasnim Zakraoui, Ines Naceur, Maysam Jridi, Imed Ben Ghorbel, Monia Smiti, Fatma Said
Questionnaire on the Use of Biologic Therapies in Managing Behçet's Disease by Internal Medicine Residents in National Hospitals in Morocco	Oumaima Elbahhari, Cheikh Khalifa international university hospital. Mohammed VI University of Health and Science, Casablanca, Morocco. Co-Authors: Abir Allaoui, Doha Kabil, Lilia Zizi, Walid Antar, Rita Filali Aniq, Abdelhamid Naitlho
Behcet's Disease In Women Versus Men	Morad Chiguer, Department of Internal Medicine B, Mohammed V Military Training Hospital, Rabat, Morocco Co-Authors: Assia Kadiri, Rime Lemouaden, Amal Charef, Jamal Fatihi
Value of the pathergy test in Behcet's disease	Doha Kabil, Cheikh Khalifa international university hospital. Mohammed VI University of Health and Sciences, Casablanca, Morocco Co-Authors: Abdelhamid Naitlho, Oumaima Elbahhari, Lilia Zizi, Walid Antar, Rita Aniq Filali, Abir Allaoui
Neuro-Behcet's syndrome and cognitive impairment	Ugur Uygunoglu, Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty Co-Authors: Osman Aykan Kargin, Serdar Arslan, Bora Korkmazer, Sabriye Guner, Ayse Ozdede, Nursena Erener, Elif Burcu Ersungur Celik, Gulcin Baktiroglu, Rauf Hamid, Ahmet Ozz, Burc Cagri Poyraz, Emire Seyahi, Osman Kizilkilic
Symptom clusters in Behçet's disease: A case series	Young Joon Park, Heera Lee, Ewha Womans University School of Medicine, Korea Co-Authors: Eun-So Lee

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Two cases of exceptional association between Behçet's disease and Takayasu vasculitis	Fatima Ibourk El Idrissi, Ibn Sina university hospital, faculty of medicine and pharmacy, Mohammed V University, Internal Medicine, Rabat, Morocco Co-Authors: Hajar Khibri, Asmaa Taouch, Jihad Aaouira, Yasmina Chhih, Naima Mouatassim, Wafa Ammouri, Mouna Maamar, Hicham Harmouche, Zoubida Tazi Mezalek, Mohamed Adnaoui
Clinical features of Behçet's disease in Jordan	Takako Ito, Kyushu University, Fukuoka, Japan Co-Authors: Wafa Madanat, Khaldoun Alawneh, Tohru Ohta, Shigeaki Ohno, Nobuyoshi Kitaichi
Long-term Results of Interferon-alpha Therapy in Patients with Behçet's Uveitis at a Tertiary Referral Center in Turkey	Murat Oklar, University of Health Sciences Kartal Dr. Lütfi Kırdar City Hospital, Istanbul, Turkey Co-Authors: Nilüfer Zorlutuna Kaymak, Burak Tanyıldız, Mehmet Engin Tezcan
Uveitis associated with Behçet's disease in children: a case series	Wijden Nabi, Fattouma Bourguiba University Hospital, Faculty of Medicine, University of Monastir, Monastir, Tunisia Co-Authors: Sana Khochtali, Sanaa Ahdeddou, Melek Kechida, Nesrine Abroug, Moncef Khairallah
The Thrombosis Chronicles: A journey through 10 cases of Behçet disease	Samya Choukair, Faculty of Medicine and Pharmacy Rabat, Morocco Co-authors: Yasmina Chhih
Regional Register of Behçet Disease in northern Morocco	Soumaya Elbachiri, Department of Internal Medicine and Clinical Immunology, University Hospital MOHAMMED VI of Tangier, Morocco Co-Authors: Oussama Boukouk, Jihane Boucht, Rachid Smaili, Wyssal Chawad, Naoual Dalhi, Naoual El ouardi, Ikram Raaidi, Zhor Mrika, Randa Ait amran, Aabyr Al asfar, Kaoutar Daoudi, Hind Stitou, Adil Gourinda, Adil Najdi, Meriem Bahloul, Rachid Belfkih, Myriem Bourkia
Eye and Behçet's Disease: Clinical and Prognostic Features Regarding 291 Cases	Amina Jahouh, University Hospital Center Ibn Rochd of Casablanca, Morocco Co-Authors: Safaa Mhaber, Mina Moudatir, Meriem Benzakour, Fatima Zahra Alaoui, Hassan El Kabli, Khadija Echchilali
Identification of Oral Ulcer Patterns in Behçet's Syndrome through K-Means Cluster Analysis and Correspondence Analysis	Berceste Polat Akmansoy, Dental School, Marmara University, Istanbul, Turkey Co-Authors: Burcu Aksoy, Can Akmansoy, Zehra Özge Çandereli, Sarah Sacoor, Bindi Gokani, Azimoon Bibi, Adebowale Adesanya, Pareen Desai, Umit Karaçaylı, Fatma Alibaz Öner, Nevsun Inanc, Tulin Ergun, Meral Yay, Haner Direskeneli, Gonca Mumcu, Farida Fortune
Behçet without oral aphthosis: interest of new criteria	Chaymaa Sollah, university hospital centre Ibn Rochd Casablanca, Morocco Co-Authors: Chaymaa Sollah, Khadija Echchilali, Meryem Benzakour, Fatima Zahra Alaoui, Hassan El Kabli, Mina Moudatir
Epidemiological and clinical characteristics of Moroccan patients with Behçet's disease: A retrospective study.	Leila Barakat, Ibn Rochd University Hospital, Morocco Co-Authors: Hajar Omali, Ibtissam Habte, Meriem Benzakour, Khadija Echchilali, Mina Moudatir, Hassan El Kabli
The particularities of angio-behçet: a review of 160 cases	Hind Bellamine, Ibn Rochd University Hospital Centre, Morocco Co-Authors: Khadija Echchilali, Meriem Benzakour, Fatimzahra Alaoui, Hassan Elkabli, Mina Moudatir
Neuroleptic malignant syndrome simulating a neurological relapse of behcet's disease: a case report	Hind Bellamine, Ibn Rochd University Hospital Centre, Morocco Co-Authors: Meriem Benzakour, Affaf Allam, Khadija Echchilali, Fatimzohra Alaoui, Hassan Elkabli, Mina Moudatir
The articular manifestations of Behçet's disease	Amina Jahouh, CHU IBN ROCHD, Morocco Co-Authors: Soukaina Mounsif, Mina Moudatir, meriem Benzakour, Fatima zahra Alaoui, Hassan Elkabli, Khadija Echchilali
Mucocutaneous manifestations of Behçet's disease	Israa Lassouli, Ibn Rochd University Hospital in Casablanca, Morocco Co-Authors: Dounia Younes, Leila Barakat, Mariam Benzakour, Khadija Echchilali, Mina Moudatir, Hassan El Kabli
Digestive Manifestations in Behçet's Disease	Nassima Boukantar, University Hospital Center Ibn Rochd Casablanca, Morocco Co-Authors: Mina Moudatir, Khadija Echchilali, Meriem Benzakour, Fatimazahra Alaoui, Hassan El Kabli
Pulmonary Artery Aneurysms in Behcet Syndrome: A Case Series	Sarra Chadli, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco Co-Authors: Hajar Khibri, Meriem Bourkia, Naima Moatassim, Wafaa Ammouri, Mouna Maamar, Hicham Harmouche, Mohamed Adnaoui, Zoubida Tazi Mezalek

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Drug Survival of Adalimumab in 335 Patients with Behçet Syndrome	Didar Ucar, Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty, Turkey Co-Authors: Ozge Sonmez, Sinem Nihal Esatoglu, Elif Kaymaz, Yesim Ozguler, Serdal Ugurlu, Emire Seyahi, Melike Melikoglu, Ugur Uygunoglu, Zekayi Kutlubay, Gulen Hatemi
Thoracic manifestations of Behçet's disease excluding cardiac involvement	Soukaina Mounsif, Chu Ibn Rochd, Turkey Co-Authors: Chaymaa Sollah, Khadija Echchilali, Meriem Benzakour, Fatima zahra Alaoui, Hassan ElKabli, Mina Moudatir
Cardiac Involvement in Behçet Syndrome: A Case Series	Sarra Chadli, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco Co-Authors: Hajar Khibri, Meriem Bourkia, Naima Moatassim, Wafaa Ammouri, Mouna Maamar, Hicham Harmouche, Mohamed Adnaoui, Zoubida Tazi Mezalek
Withdrawal of Colchicine in Behçet Syndrome	Gulen Hatemi, Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty, Turkey Co-Authors: Basak Sirin, Sinem Nihal Esatoglu, Hasan Yazici
Frequency of Tuberculosis and Associated Risk Factors in Patients with Behçet Syndrome	Gulen Hatemi, Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty, Turkey Co-Authors: Murat Yildirim, Sinem Nihal Esatoglu, Yesim Ozguler, Melike Melikoglu, Izzet Fresko, Vedat Hamuryudan
Mimickers of Nervous System Involvement Among Patients with Behçet Syndrome	Ugur Uygunoglu, Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty, Turkey Co-Authors: Elif Dincses, Elif Buse Caliskan, Ece Kaya, Melih Tutuncu, Sabahattin Saip, Aksel Siva, Gulen Hatemi
Neuro-Behçet revealed by an hemiface edema	Doha Kabil, Cheikh Khalifa international university hospital. Mohammed VI University of Health and Sciences, Casablanca, Morocco Co-Authors: Oumaima Elbahhari, Abdelhamid Naitlho, Lilia Zizi, Walid Antar, Rita Filali Aniq, Abir Allaoui
Efficacy and Tolerability of Switching Colchicine Preparations in Patients with Behçet Syndrome	Sinem Nihal Esatoglu, Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty, Turkey Co-Authors: Sena Fidan, Yesim Ozguler, Serdal Ugurlu, Emire Seyahi, Melike Melikoglu, Izzet Fresko, Zekayi Kutlubay, Gulen Hatemi
Abdominal Surgical Interventions Among Patients with Gastrointestinal Involvement of Behçet Syndrome	Sinem Nihal Esatoglu, Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty, Turkey Co-Authors: Sabriye Guner, Sevim Guler, Gulen Hatemi, Nuray Kepil, Yusuf Ziya Erzin, Aykut Ferhat Celik, Ibrahim Hatemi
Predictive Modeling of Behçet's Disease Using Machine Learning: Insights from Clinical Data Analysis	Abire Allaoui, Mohammed VI University of Health and Sciences, Casablanca, Morocco, Mohammed VI Center of Research and Innovation, Rabat, Morocco., Clinical Immunology, Autoimmunity and Inflammation Laboratory (LICIA), Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco Co-Authors: Leila Barakat, Mina Moudatir, Khadija Echchilali, Hassan Elkabli
Crohn's Disease and Intestinal Behcet: Two Sides of The Same Coin	Rime Lemouaden, Internal Medicine B Co-Authors: Assia Kadiri, Amal Charef, Morad Chiguer, Yassine Oualehsine, Jihane Benhammou, Fadoua Mekouar, Naoual Elomri, Mohamed Jira, Jamal Fatihi
Unusual Manifestations Revealing Behçet's Disease: A Study of 43 Cases.	Achraf El Kabli, Chu Ibn Rochd of Casablanca, Morocco Co-Authors: Mina Moudatir, Meriem Benzakour, Khadija Echchilali, Hassan El Kabli
Behçet's Disease and Cancer: a study of 6 cases.	Safaa Mhaber, Chu Ibn Rochd of Casablanca, Morocco Co-Authors: Dounia Youness, Khadija Echchilali, Hassan El Kabli, Mina Moudatir
Behçet's disease and pregnancy: 23 pregnancies in 10 women	Rime Lemouaden, Internal Medicine B Co-Authors: Amal Charef, Assia Kadiri, Morad Chiguer, Yassine Oualehsine, Jihane Benhammou, Fatimazahrae Boucham, Fadoua Mekouar, Naoual Elomri, Mohamed Jira, Jamal Fatihi
Intracardiac Thrombosis in Behcet Syndrome: A Case-Control Study	Sarra Chadli, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco Co-Authors: Hajar Khibri, Meriem Bourkia, Naima Moatassim, Wafaa Ammouri, Mouna Maamar, Hicham Harmouche, Mohamed Adnaoui, Redouane Abouqal, Zoubida Tazi Mezalek

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Vascular Thrombosis in Behçet's Syndrome: A Comprehensive Analysis of 146 Cases	Sarra Chadli, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco Co-Authors: Hajar Khibri, Meriem Bourkia, Naima Moatassim, Wafaa Ammouri, Mouna Maamar, Hicham Harmouche, Mohamed Adnaoui, Rachid Razine, Zoubida Tazi Mezalek
Pulmonary artery thrombosis in Behcet Syndrome: A Case-Series	Sarra Chadli, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco Co-Authors: Hajar Khibri, Meriem Bourkia, Naima Moatassim, Wafaa Ammouri, Mouna Maamar, Hicham Harmouche, Mohamed Adnaoui, Zoubida Tazi Mezalek
Clinical and Ultrasonographic Correlates of Postthrombotic Venous Remodeling in Behçet's Syndrome: A Cross-Sectional Study	Tumay Ak., Istanbul University-Cerrahpasa, Cerrahpasa Medical Faculty, Turkey Co-Authors: Seyfullah Halit Karagoz, Omer Faruk Sarıahmetoglu, Alican Karakoc, Melike Melikoglu, Ibrahim Adaletli, Emire Seyahi
Acute myocarditis revealing Behçet's disease: a case report.	Hind Bellamine, Ibn Rochd University Hospital Centre, Morocco Co-Authors: Khadija Echchilali, Fatim-Zahra El Alaoui, Hassan Elkabli, Meriem Benzakour, Mina Moudatir
CMV Associated Uveitis in Immunocompetent Patient and Ocular Behçet's Syndrome: A Diagnostic Challenge	Nada Boumazzourh, Ibn Sina Hospital, Faculty of medicine and pharmacy, Mohammed V University, Rabat, Morocco
Unusual arterial localization revealing Behçet's disease: Splenic artery aneurysm	Ichrak Hajaj, N. Mouatassim; H. Khibri, M. Maamar; H. Harmouche, Z. Tazi Mezalak, M. Adnaoui Co-Authors: Chhih Yasnima, Khibri Hajar, Chadli Sara, Mouatassim Naima, Ammouri Wafaa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida
Unusual Stenosis in Vasculo-Behcet Disease (BD)	Ichrak Hajaj Co-Authors: Chhih Yasmina, Khibri Hajar, Chadli Sara, Mouatassim Naima, Ammouri Wafa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida
Clinical characteristics of pediatric Behçet's disease in different geographies	Ummusen Kaya Akca, Hacettepe University Faculty of Medicine, Turkey Co-Authors: Farhad Shahram, Erdem Karabulut, Massoomeh Akhlaghi, Ozlem Akgun, Mustafa Cakan, Esra Esen, Caterina Matucci Cerinic, Seyede Tahereh Faezi, Ruya Torun, Erbil Unsal, Marco Gattorno, Nuray Aktay Ayaz, Aysenur Pac Kısaarslan, Betul Sozeri, Ezgi Deniz Batu, Isabelle Koné-Paut, Seza Ozen
Cardiac involvement in Behcet disease	Rim Zribi, Rabta Hospital Tunis El manar University, Tunis, Tunisia , Rabta Hospital Tunis El manar University, Tunis, Tunisia Co-Authors: Ines Naceur, Tayssir ben achour, mayssem jridi, Imed Ben ghorbel, Mounir Lamloum, Monia Smiti, Fatma Said
Exceptional association between Behcet's Disease and Overlap Syndrome	Samya Choukair, Faculty of Medicine and Pharmacy Rabat Morocco Co-Authors: Yasmina Chhih
A heart with two tragedies: a Behçet's disease revealed by an infectious endocarditis.	Ichrak Hajaj Co-Authors: yassmina chhih, Khibri Hajar, Fari Safae, Sara Chadli, Mouatassim Naima, Ammouri Wafaa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida
A Rare Cause of Portal Hypertension: Porto-sinusoidal Vascular Disease (PSVD) and Behçet's Disease: A Case Report	Ichrak Hajaj, N. Mouatassim, H. Khibri, W. Ammouri, M. Maamar, H. Harmouche, Z. Tazi-Mezalak, M. Adnaoui Co-Authors: Yassmina chhih, Khibri Hajar, Fari Safaa, Chadli Sara, Mouatassim Naima, Ammouri Wafaa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida
Spectacular image of a Giant False Aneurysm of the Celiac Artery during Behçet's disease	Meryem Zaizaa, Mohammed V Military Hospital,Rabat, Morocco Co-Authors: Nissrine Bahadi, Oumama Jamal, Zineb EL Bougrini, Farah Ahallat, Bilal Talamoussa, Nawal Sahel, Ilyass El Kassimi, Adil Rkiouak, Youssef Sekkach
Behçet uveitis presenting with or without retinitis: A comparative study	Moncef Khairallah, Faculty of Medicine of Monastir, Tunisia Co-Authors: Nesrine Abroug, Oumaima Allagui, Wijdene Nabi, Imen Ksiaa, Béchir Jelliti
Behcet's Disease and IgA Vasculitis: A Real Assosciation or Quincidence ?	Bouzgarou Mohamed, Resident
Hughes-Stovin syndrome about 3 cases and literature review.	Fatima Zahra Boucham, Internal Medicine Co-Authors: Charaf Eddine Amezyane, Mohamed Jira, Fadoua Mekouar, Nawal El Omri, Jamal Fatihi

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The characteristics of a northern Israeli cohort of patients with Behcet's syndrome	Mohammad Naffaa, Galilee Medical Center, Naharyia, Israel, The Azrieli Faculty of Medicine, Bar-Ilan University, Safed, Israel Co-Authors: Firas Sabbah, Fadi Hassan, Rula Daood, Helana Jeries
Behcet and Hypnos: evaluation of sleep quality in Behcet's disease in a Tunisian population	Tayssir Ben Achour, Rabta University Hospital, Morocco Co-Authors: Donia Dridi, Ines Naceur, Maysam Jridi, Imed Ben Ghorbel, Monia Smiti, Fatma Said
Assessment of fatigue in Tunisian Behcet's patients: prevalence and associated factors	Tayssir Ben Achour, Rabta University Hospital, Morocco Co-Authors: Donia Dridi, Ines Naceur, Maysam Jridi, Imed Ben Ghorbel, Monia Smiti, Fatma Said
Behcet's disease: how does it begin?	Donia Dridi, Rabta University Hospital, Rabta University Hospital, Morocco Co-Authors: Ines Naceur, Tayssir Ben Achour, Maysam Jridi, Imed Ben Ghorbel, Monia Smiti, Fatma Said
Case Report of a Patient with Recurrent Clinical Neuro-Behçet's Disease and Persistently Normal MRI Findings	Douaa Elmejdoubi, Neurology Department, Mohammed VI University Hospital Centre, Marrakesh, Morocco
Association study of HLA-A and HLA-B alleles with clinical manifestations of Behçet's disease in a Japanese population	Akira Meguro, Yokohama City University Graduate School of Medicine, Japan Co-Authors: Masaki Takeuchi, Masao Ota, Nobuhisa Mizuki
Autoimmune manifestations in the clinical spectrum of haploinsufficiency A20: not to be confused with the diagnosis of Behçet	Khaoula Ikemakhen, Internal Medicine-Immunology-Clinical Hematology, Hospital center, Périgueux, France Co-Authors: Baulier Gildas
Gender influence in Behçet Syndrome: a Tunisian study	Melek Kechida, Fattouma Bourguiba University Hospital, Monastir, Tunisia Co-Authors: Imen Ksiaa, Ansar Meftah, Imen Chaaben, Ines Khochtali, Bechir Jelliti, Moncef Khairallah
Failure of Conventional Immunosuppressive Therapy Among Patients with Behçet Uveitis: Causes and Management Modalities	Melek Kechida, Fattouma Bourguiba University Hospital, Monastir, Tunisia Co-Authors: Imen Ksiaa, Omar Miladi, Oumayma Allagui, Bechir Jelliti, Moncef KhairallahX
Psycho-cognitive disorders in patients with neuro-Behçet	Khadija Baitha, Department of neurology and clinical neurophysiological explorations, Ibn Rochd University Hospital, Hassan II University, Casablanca, Morocco.
Neurobehçet revelation mode	Baitha Khadija, Department of neurology and clinical neurophysiological explorations, Ibn Rochd University Hospital, Hassan II University, Casablanca, Morocco Co-Authors: Meskaoui Jihane
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Rare Association of Behçet's Disease and Marfan Syndrome: Diagnostic and therapeutic challenge	Hajar Joulal, Mohammed VI University Hospital of Marrakech, Morocco Co-Authors: Jaouad Yousfi, Laila Benjilali, Mouna Zahlane, Lamiaa Essaadouni
Comparison of Diagnostic Criteria in Behçet's Disease in a Moroccan population	Hajar Joulal, Mohammed VI University Hospital of Marrakech, Morocco Co-Authors: Jaouad Yousfi, Laila Benjilali, Mouna Zahlane, Lamiaa Essaadouni
Aortic abdominal aneurysm and splanchnic vein thrombosis in Behçet's syndrome: an unusual association	Nada Boumazzourh, Ibn Sina Hospital, Faculty of medicine and pharmacy, Mohammed V University, Rabat, Morocco
Evaluation Of Body Image Perception and Social Appearance Anxiety in Behcet's Disease	Fatma Alibaz-Oner, Marmara University School of Medicine, Turkey Co-Authors: Rafi Haner Direskeneli, Tulin Ergun, Alihan Akdag, Damla Tanrikulu, Zeynep Tuye Cakir
Knowledge of Tunisian primary care professionals regarding Behçet disease	Houssem Abida, Mongi Slim University Hospital, Tunisia Co-Authors: Zeineb Meddeb, Cherifa Abdelkefi, Amira El Ouni, Sana Toujani, Kamel Bouslama, Thara Larbi, Saloua B'chir Hamzaoui
Comparison of inflammatory markers in Behçet's disease	Kamila Nurbaeva, V.A. Nasonova Reasearch Institute of Rheumatology, Russia Co-Authors: Tatiana Reshetnyak, Regina Goloeva, Aleksandr Lila
A rare case of Behçet's disease with severe vascular involvement.	Leila Barakat, Ibn Rochd University Hospital, Morocco Co-Authors: Meriem Benzakour, Mina Moudatir, Khadija Echchilali, Hassan El Kabli
Vascular Behçet's Disease: Focus on aneurysms	Abir Cherif, Rabta Hospital, Tunis, Tunisia Co-Authors: Tayssir Ben Achour

II. E-POSTERS

Prevalence of Behçet's disease in 384 patients with recurrent oral aphtosis followed in liberal outpatient internal medicine	Said El Kettani, Liberal
Serum calprotectin as a biomarker of disease activity in Behçet's disease	Kamila Nurbaeva, V.A. Nasonova Reasearch Institute of Rheumatology, Russia Co-Authors: Tatiana Reshetnyak, Maria Cherkasova, Regina Goloeva, Tatiana Lisitsyna, Aleksandr Lila
Serum calprotectin in Behçet's disease	Kamila Nurbaeva, V.A. Nasonova Reasearch Institute of Rheumatology, Russia Co-Authors: Tatiana Reshetnyak, Maria Cherkasova, Regina Goloeva, Galina Davidova, Tatiana Lisitsyna, Aleksandr Lila
Serum MPO-DNA complex in Behçet's disease	Kamila Nurbaeva, V.A. Nasonova Reasearch Institute of Rheumatology, Russia Co-Authors: Tatiana Reshetnyak, Ivan Ptashnik, Anna Kudriaeva, Alexey Belogurov, Regina Goloeva, Aleksandr Lila
NLR and SII in Behçet's disease	Kamila Nurbaeva, V.A. Nasonova Reasearch Institute of Rheumatology, Russia Co-Authors: Tatiana Reshetnyak, Regina Goloeva, Aleksandr Lila
Anticoagulant treatment in addition to immunosuppressives decreases the relapse rate in Pulmonary Arterial Involvement of Behçet's Disease	Fatma Alibaz-Oner, Marmara University, School of Medicine, Department of Internal Medicine, Division of Rheumatology, Istanbul, Turkey. Co-Authors: Kerem Abacar, Ayşe Elif Boncukcuoğlu, Aysun Aksoy, Derya Kocakaya, Cagatay Cimşit, Haner Direskeneli
Systemic Immune-Inflammation Index (SII) Role in Diagnosis of Behçet Disease	Faezeh Moghimpour Bijani, Shariati hospital, internal medicine department, School of medicine, Tehran university of medical sciences, Iran Co-Authors: Majid Alikhani, Tahereh Faezi, Tahereh Yavari
Betacellulin is the major ligand of epidermal growth factor receptor pathway in the exacerbation of experimental autoimmune uveoretinitis	Koichi Yokoi, Hokkaido University, Japan Co-Authors: Kenichi Namba, Miyuki Murata, Kayo Suzuki, Keitaro Hase, Daiju Iwata, Miki Hiraoka, Nobuyoshi Kitaichi, Masayuki Murakami, Susumu Ishida
CitH3 as a specific marker of NETosis in Behçet's disease	Kamila Nurbaeva, V.A. Nasonova Research Institute of Rheumatology, Russia Co-Authors: Tatiana Reshetnyak, Maria Cherkasova, Regina Goloeva, Aleksandr Lila
Does Vein Wall Thickness have prognostic value in Behçet's Disease? A prospective follow-up study	Fatma Alibaz-Oner Co-Authors: Kerem Abacar, Rabia Ergelen, Fatma Temiz, Özge Kipri, Tulin Ergun, Haner Direskeneli
Predictive model for long-term visual prognosis of Behcet's uveitis using machine learning	Nobuyoshi Kitaichi, Health Sciences University of Hokkaido, Japan Co-Authors: Ryosuke Dei, Miki Hiraoka, Ippei Yoshikawa, Kenichi Namba, Shigeaki Ohno, Susumu Ishida, Kazuo Nakajima
Articular involvement during Behçet's disease in private outpatient internal medicine in 197 patients	Said El Kettani, Liberal
Ocular manifestations of Behçet's disease (about 197 patients consulting internal medicine in a private outpatient setting)	Said El Kettani
Behçet Disease in the Elderly	Merieme Benzakour, Hassan II University of Casablanca, Hassan II University of Casablanca, Morocco Co-Authors: Khadija Ennekhali, Mina Moudatir, Khadija Echchilali, Hassan El Kabli
Clinicopathological Characteristics of Behçet disease with Severe Aortic Regurgitation	Wenjie Zheng, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences & Peking Union Medical College, China Co-Authors: Menghao Zhang, Xun Wang, Yeling Liu, JInjing Liu, Wenze Wang
ABO Blood Groups and Increased Risk for Vascular Involvement in the Patients with Behçet Disease	Erdem Bektas, Istanbul University, Istanbul Faculty of Medicine, Department of Internal Medicine, Turkey Co-Authors: Abdülkadir Büyükdemir, Yasemin Yalcınkaya, Bahar Artim Esen, Murat Inanc, Ahmet Gül
Medical practice assessment on anticoagulation in Behçet's disease (BD): Primary results from a Mediterranean survey.	Maysam Jridi, Rabta University hospital Center, Morocco Co-Authors: Tayssir Ben achour, Ines Naceur, Monia Smiti, Fatma Said
Behçet's Disease and Celiac Disease: Coexistence Case	Nisrine Bahadi, HMIMV Rabat, Morocco Co-Authors: Zineb Elbougrini, Bilal Talamousssa, Farah Ahallat, Meryem Zaizaa, Oumama Jamal, Nawal Sahel, Adil Rkiouek, Youssef Sekkach

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Analysis of two cases of false aneurysm in Behçet's disease	Meryem Zaizaa, Mohammed V Military Hospital,Rabat, Morocco Co-Authors: Ilyass El Hachimi, Nissrine Bahadi, Ilyass El Kassimi, Bilal Talamoussa, Nawal Sahel, Oumama Jamal, Farah Ahallat, Zineb El Bougrini, Adil Rkiouak, Youssef Sekkach
Online support group for Behçet Diseases patients: the Tunisian experience	Houssem Abida, Mongi Slim University Hospital La Marsa, Tunisia Co-Authors: Zeineb Meddeb, Cherifa Abdelkefi, Sana Toujani, Amira El Ouni, Kamel Bouslama, Thara Larbi, Saloua B'chir Hamzaoui
Subclinical herpesvirus activity prevents remission of uveitis in patients with Behcet's disease	Tatiana Lisitsyna, V.A. Nasonova Research Institute of Rheumatology, Russia Co-Authors: Ekaterina Sorozhkina, Galina Krichevskaya, Nataliya Balatskaya, Galina Davidova, Kamila Nurbaeva, Tatiana Reshetnyak
The efficacy of an anti-inflammatory peptide in a mouse model of Behçet's disease induced by herpes simplex virus	Seonghyang Sohn, Ajou University, South Korea Co-Authors: Bunsoon Choi, Hasan Sayeed, Hye-Myung Ryu
Cytokine signature differences in major phenotypic groups of Behçet's disease	Rabia Deniz, Marmara University, School of Medicine, Department of Medical Biology, University of Health Sciences Başakşehir Çam and Sakura City Hospital, Department of Rheumatology, Istanbul, Turkey Co-Authors: Zeliha Emrence, Şeyma Punar, Berk İleri, Kazım Yalçın Arga, Fatma Alibaz Öner, Cemal Bes, Haner Direskeneli, Ahmet Gül, Can Erzik
Efficacy and tolerability of low-dose roflumilast in refractory oral ulcers in Behçet's disease and recurrent aphthous stomatitis	Doyoung Kim, Yonsei University College of Medicine, South Korea Co-Authors: Hae June Sung, Kyung Bae Chung, Hyunwoo Jang
Screening for familial disease presence in first-degree relatives of Behçet's disease patients: Is measurement of common femoral vein wall thickness valuable for the diagnosis ?	Ayşe Elif Boncukcuoğlu, Department of Internal Medicine, Marmara University, School of Medicine, Istanbul, Turkey Co-Authors: Erhan Topal, Tülin Ergun, Rabia Ergelen, Dilara Kaş, Elifnur Alkan, Gamzenur Kaya, Şevket Girgin, Haner Direskeneli, Fatma Alibaz-Oner
Evaluation of Thrombophilia Risk Factors in Behçet's Patients with Vascular Involvement	Fatma Alibaz-Oner, Marmara University Department of Internal Medicine, Division of Rheumatology, Turkey Co-Authors: Ummugulsum Karayildiz, Tulin Ergun, Rafi Haner Direskeneli

ORAL PRESENTATIONS

CO1.1
The Relationship Between the NETosis Findings and Disease Activity in Behçet Disease

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Introduction: Neutrophil extracellular traps (NETs) have been claimed in the pathogenesis of Behçet Disease (BD). We herein aimed to investigate the potential relationship between the NETosis findings and local and systemic disease activity in BD.

Method: Serum and saliva cf-DNA, NE, MPO, and cit-H3 levels were measured as NETosis findings, and the results were adjusted according to the peripheral blood neutrophil counts (as the amount of biomarker per 1 million neutrophils).

Discussion: The study group was consisted of 30 active BD patients and 10 healthy individuals (Table 1). Patient groups and control group were similar in terms of confounder factors. In active BD, serum cf-DNA and NE levels were found to be high, whereas adjusted serum MPO and cit-H3 levels were found to be low. In inactive BD, serum NE level were higher than controls, while serum MPO, adjusted serum MPO and adjusted serum cit-H3 levels were lower than controls. No difference was found in salivary NETosis findings between patient groups and control. Serum and saliva cf-DNA levels showed a decrease in longitudinal follow-up towards remission (**Figure 1**). Quite similar results have been observed in vascular active patients. Serum and saliva cf- DNA positively correlated with C-reactive protein and erythrocyte sedimentation rate, while adjusted serum MPO and cit-H3 negatively correlated.

Conclusion: NETosis findings showed changes in association with systemic and/or local activity of BD patients in relation to the disease manifestations. cf-DNA levels potentially indicated the local and systemic disease activity, and NE level was high in both active and inactive periods. Changes of the findings following adjustment may indicate that the NETosis findings observed in serum could be related to high neutrophil turnover in the active phase of the disease. Further studies are required to clarify the mechanism of the low adjusted serum MPO and cit-h3 levels, particularly in the active patients, and the biomarker potential of NETosis findings in BD.

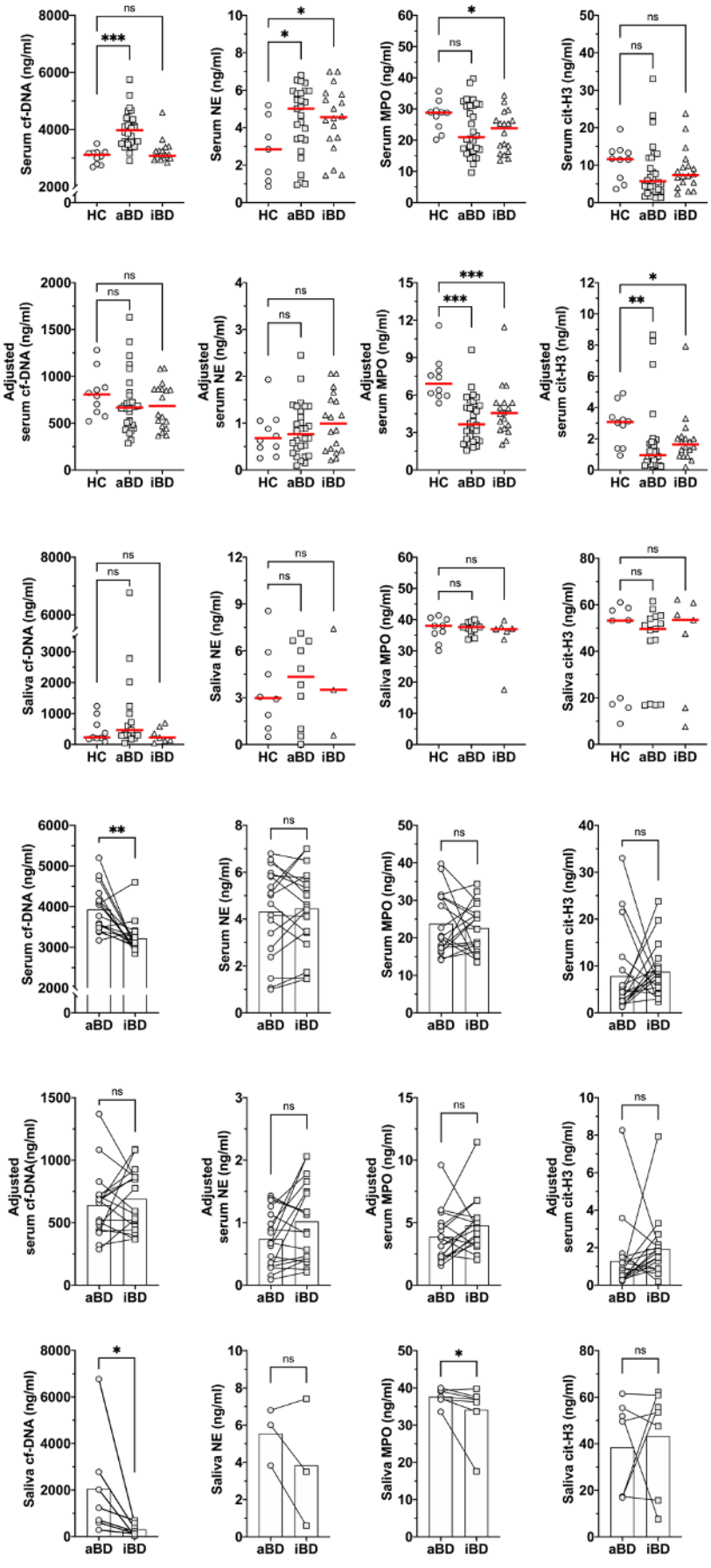

Figure 1. The levels of NETosis findings in groups and changing in longitudinally followed-up.

	Active BD (n=30)	Inactive BD (n=18)	Control (n=10)
Sex, male, n (%)	21 (70)	11 (61)	5 (50)
Age, years, med (IQR)	34 (26-42)	37 (31-42)	30 (27-48)
Smoking, n (%)	8 (27)	7 (39)	3 (30)
BD duration, months, med (IQR)	54 (10-150)	48 (10-70)	
Cumulative involvment, n (%)			
Mucocutaneous	30 (100)	18 (100)	
Arthritis	12 (40)	8 (44.4)	
Uveitis	8 (26.7)	2 (11.1)	
Neurologic	2 (6.7)	1 (5.6)	
Gastrointestinal	1 (3.3)	0 (0)	
Vascular	7 (23.3)	6 (33.3)	
Others	1 (3.3)	1 (5.6)	
Treatment			
Naive	7 (23.3)	0 (0)	
Colchicum	23 (76.7)	18 (100)	
Azathioprine	3 (23.3)	10 (56.6)	
Corticosteroid	3 (10)	9 (50)	
TNFi	0 (0)	1 (5.6)	
Laboratory values			
Neutrophil, 10^3/ml, med (IQR)	6.3 (4.7-7.7)	4.7 (3.4-6.6)	4 (3.2-4.6)
CRP, mg/l, med (IQR)	7.5 (3.16-26.5)	2.44 (0.6-4.46)	1.35 (0.55-2.3)
ESR, mm/h, med (IQR)	10.5 (7-23)	9 (3-15)	4 (4-6)

Table 1. Demographic, clinical, laboratory characteristics of patients and control.

CO1.2

The Mevalonate Pathway Metabolite Farnesyl Pyrophosphate Induces Neutrophil Hyperactivation via the TRPM2 Calcium Channel in Behçet's Disease

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Co-Authors: Menghao Zhang, Na Kang, Xin Yu, Jinjing Liu, Hua Chen, Wanli Liu

Introduction: Behçet's disease (BD) is a vasculitis of unknown etiology, with polymorphonuclear neutrophil (PMN) hyperactivation. Aberrant metabolic intermediates are endogenous danger signals eliciting innate immune responses. Farnesyl pyrophosphate (FPP) is an intermediate triggering pro-inflammatory responses and even cell death. This study aimed to reveal the immunometabolic aberrations in BD and to highlight the pathogenic implications of FPP in BD-PMN hyperactivation.

Method: We analyzed BD lipidomics and transcriptomics, and detected FPP levels, pro- inflammatory cytokines and NETs production of BD-PMN. Transcriptomics of calcium channels and TRPM2 expression in BD-PMN were analyzed pre- and post-therapy with TNF-α inhibitors.

Discussion: Our multi-omics analysis revealed elevated steroid biosynthesis in BD serum, with consistent metabolic aberrations noted only in PMN, particularly in cholesterol biosynthesis (Fig. 1A-C). FPP was confirmed as the key intermediate in the MVA pathway promoting BD-PMN hyperactivation by interfering with key enzymes upstream and downstream of FPP (Fig 1D). FPP was elevated in BD serum and PMN (Fig. 1E-F), and positively correlated with disease activity indicators like CRP, ESR and BDCAF. In addition, FPP levels were significantly decreased in remission BD after adequate treatment. Notably, FPP specifically promoted PMN to produce proinflammatory cytokines and NETs in a concentration-dependent manner (Fig. 2A- C). Our comprehensive transcriptomic analysis revealed a significantly increased TRPM2 in BD-PMN (Fig. 2C). Therapy with TNF-α inhibitors downregulated TRPM2 in BD-PMN in vivo (Fig. 2C), and TNF-α neutralizing inhibition attenuated FPP-induced BD-PMN hyperactivation in vitro, as evidenced by remarkedly decreased proinflammatory cytokines and NETs (**Figure 2D-E**).

Conclusion: Our study highlights that FPP promotes BD-PMN hyperactivation via TRPM2, and provide a novel therapeutic mechanism of TNF-α inhibitors in BD.

Figure 1

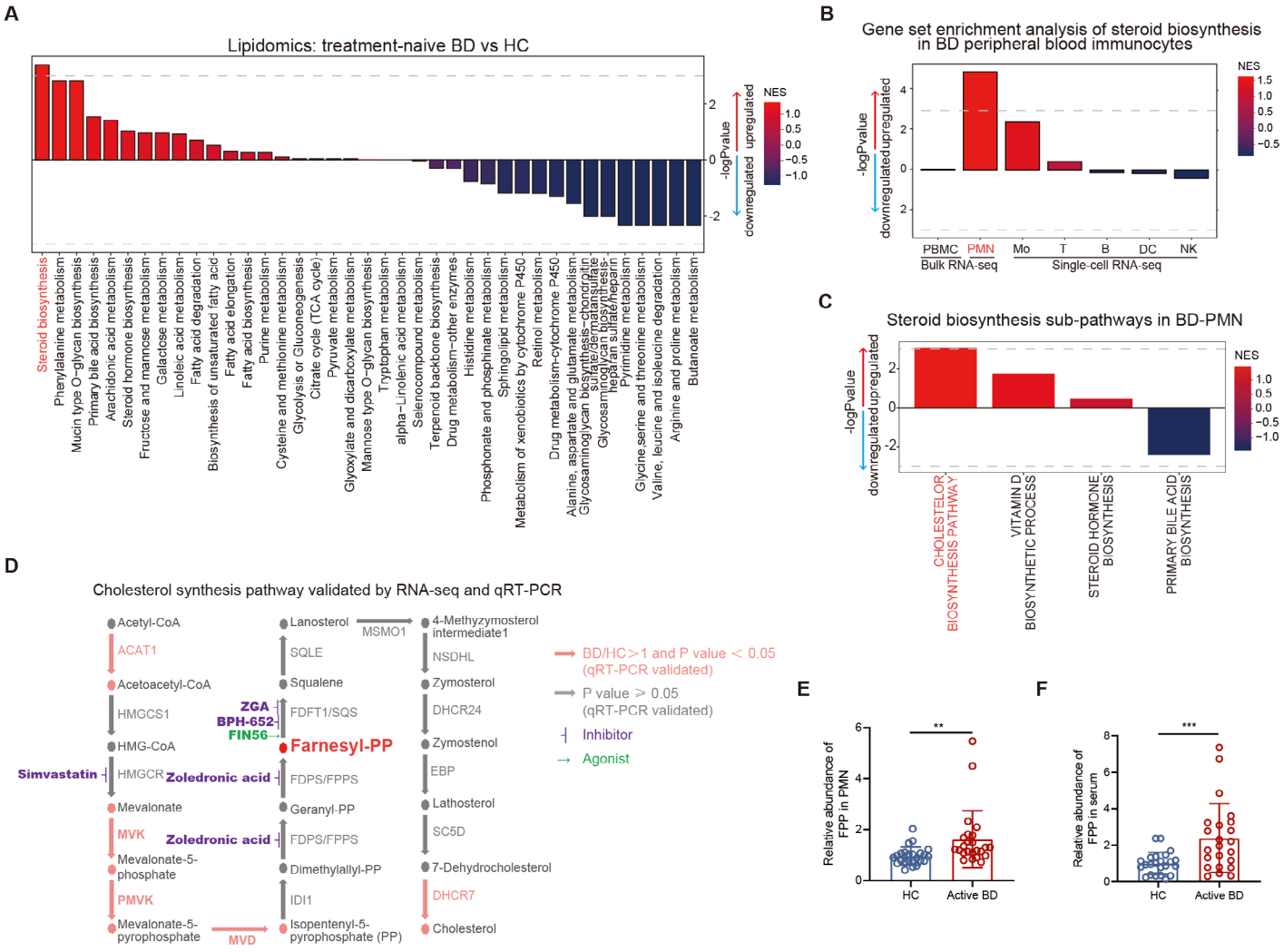

Figure 1. Multi-omic analysis revealed the elevated MVA pathway with upregulated FPP in BD (A), Lipidomic analysis between treatment-naïve BD and HC serum. (B), GSEA of steroid biosynthesis pathway in BD and HC peripheral blood immune cells. (C), GSEA of steroid synthesis sub-pathways in BD and HC PMN. (D), Schematic plot showing the intervention of the cholesterol synthesis pathway. Significantly elevated enzymes validated by both BD PMN RNA-seq and qRT-PCR are marked in red. The agonists and inhibitors are marked by green and purple arrows, respectively. (E-F), Relative abundance of FPP in PMN (E) and serum (F). **, p<0.01; ***, p<0.001.

Figure 2

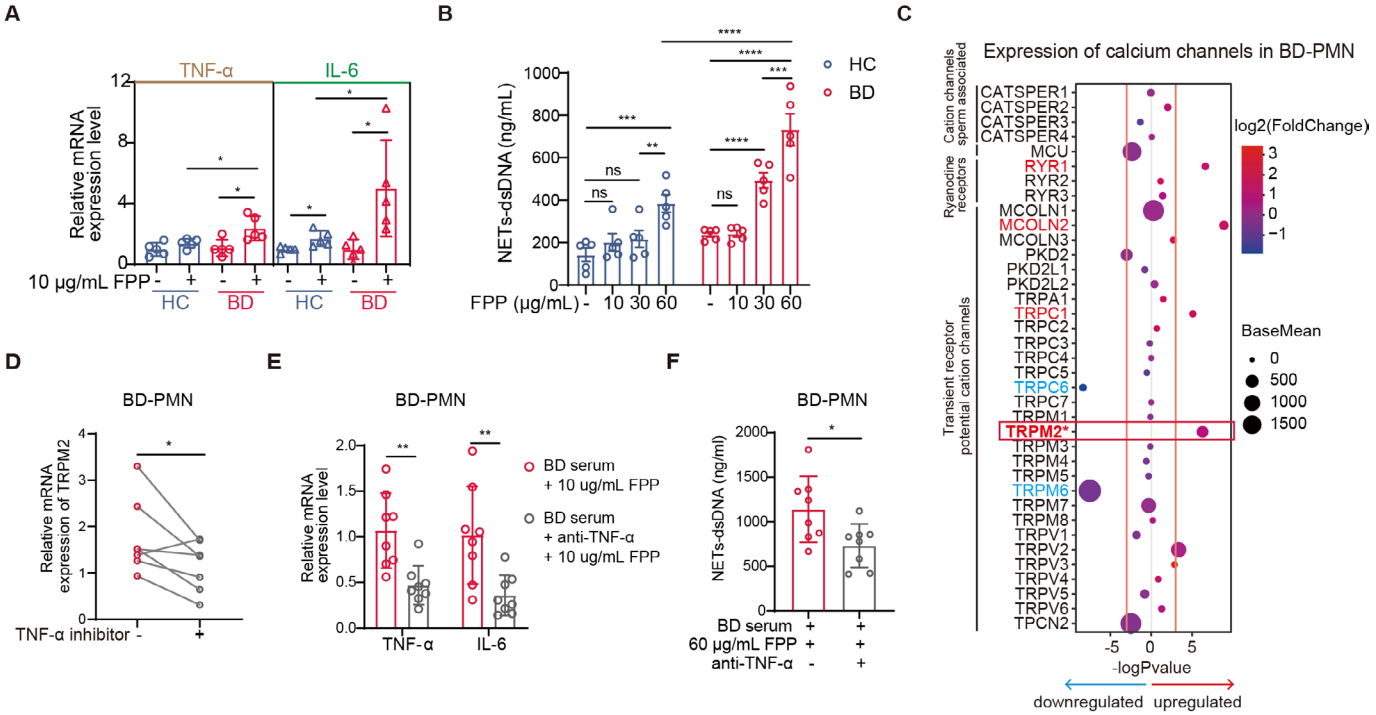

Figure 2: Serum TNF-α-induced TRPM2 overexpression is critical for FPP to promote PMN hyperactivation. (A), The expression levels of proinflammatory cytokines in 10 μg/mL FPP-stimulated PMNs. (B), Levels of double-stranded DNA in the supernatants of FPP-stimulated PMN. (C), Transcriptomics differential expression analysis of RR-sensitive calcium channels in GSE205867. (D), The mRNA expression levels of TRPM2 in PMN from BD patient before and after TNF-α inhibitor treatment. (E-F), Levels of TNF-α and IL-6 (E) and NETs-dsDNA (F) in FPP-stimulated BD-PMN after preincubation with BD serum in the presence or absence of TNF-α-neutralizing antibody. *, p<0.05; **, p<0.01; ***, p<0.001; ****, p<0.0001.

C01.3

PDE4 inhibitor mitigates activated CD8+ T cells through NF-κB signaling in Behçet's syndrome

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Introduction: Behçet's disease (BD) is a systemic vasculitis with inflammatory lesions mediated by cytotoxic T cells and neutrophils. Here, we explore the critical involvement of NF-κB signaling pathway in proinflammatory CD8+ T cells differentiation and disease progress of BD patients.

Method: We performed microarray gene expression analyses, flow cytometry, immunophenotyping, immunohistochemistry and functional assessments of CD8+ T cells from BD patients and HD.

Discussion: Transcriptionally, among the 6,595 up-regulated genes in CD8+ T cells of BD vs HD, we highlighted a great enrichment for pathways linked to NF-κB and TLR signaling (i.e. NFKB1, RELB, REL, TLR1, IRF4). Phenotypically, CD8+ T cells from BD had a higher expression of phosphorylated NF-κB (pNF-κB, 4.4 ± 0.9 vs. 1.8 ± 0.2 in MFI, p = 0.001), were more activated (higher CD11c, CD11b, CD25, CD69 and TNF-α, IFN-γ expression) and exhibited more expression of Perforin and Granzyme B (33% ± 9 vs. 9 ± 5, p = 0.009, 47% ± 8 vs.21% ± 6, p = 0.02, respectively) as compared to HD. Phosphodiesterase-4 (PDE4), an immune cell enzyme that activate the NF-κB pathway was up-regulated in blood and skin lesions of BD. In vitro and in vivo inhibition of PDE4 strongly inhibited CD8+ T cell activation, cytokine secretion, cytotoxicity and proliferation.

Conclusion: We highlighted that activated CD8+ T cells through NF-κB signaling pathway are instrumental in BD.

CO1.4

Increased granzyme K+ CD8+ senescent T cells in Behçet's disease

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Introduction: Recently, age-associated granzyme K expressing CD8+ T cells are found to be a distinct T cell population expressing exhaustion and tissue homing markers resulting in inflammaging. This T cell sub-population were increased not only in normal immune-aging process, also in rheumatoid arthritis, one of the auto-inflammatory diseases. Thus, we aimed to evaluate the intra/extracellular levels of granzymes in BD patients.

Method: First, we performed enzyme-linked immunosorbent assay (ELISA) using serum samples of age-matched 19 healthy controls, 19 active BD patients, and 19 inactive BD patients. Next, we performed stimulation assay conducting flow cytometry with the peripheral blood mononuclear cells (PBMC) in both ex vivo condition and after 72 hours of culture with anti-CD3 antibodies and anti-CD28 antibodies. Finally, we performed bulk flow cytometry with PBMC in ex vivo condition.

Discussion: First, the serum levels of granzyme K were significantly increased in active BD group compared to healthy control group (P=0.0231) and to inactive BD group (P=0.0025). The granzyme A level was decreased and the level of granzyme B was increased in active BD group than other groups, but both were not statistically significant. Next, significantly increased Granzyme K+ CD8+ CD27- CD28- senescent T cells were observed in active BD group than healthy control group (P=0.045) in ex vivo condition but not in the CD3, CD28 stimulated condition. Granzyme A and granzyme B levels were not statistically significant. Finally, also in the bulk flow cytometry, Granzyme K+ CD8+ CD27- CD28- senescent T cells were significantly increased in active BD group than HC group (P=0.047).

Conclusion: Thus, we suggest granzyme K could exhibit extracellular functions, which would be related to one of the characteristics of senescent CD8+ T cells in BD. In conclusion, increased granzyme K serum level and high granzyme K+ senescent CD8+ T cells in active BD group could be a hallmark for inflammaging process of BD.

CO1.5

Enhanced Response to Stimulation of γδ T cells induces plasma B cells in patients with Behçet's syndrome

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Introduction: The pathogenesis of Behçet's syndrome (BS) is complex and associated with dysregulated immune responses. γδ T cells are unconventional T lymphocytes that have emerged as contributors to pathogenesis in BS. Our preliminary analysis has shown that γδ T from BS patients express a T follicular helper (Tfh) cell-like phenotype with increased levels of co-stimulatory molecules associated with induction of B cell differentiation. To further our understanding of the pathogenic mechanisms in BS, we explored the role of γδ T cells in promoting B cell differentiation in patients with BS presenting with either mucocutaneous or ocular manifestations.

Method: PBMCs from 44 BS patients and 18 healthy controls (age- and gender-matched) were cultured for 1, 3, and 5 days in the presence of HMBPP (a bacteria-derived phospho- antigen that specifically stimulates γδ2 T cells). Following in vitro cultures, the induction of plasma B cell differentiation was analyzed by flow cytometry. Cytokines and autoantibody levels were measured -by Luminex cytokine array and ELISA respectively- in the cell cultures supernatants and serum samples of BS patients and the healthy control group.

Discussion: γδ T cells from patients with BS, with higher expression of Tfh markers, induced a higher frequency of plasma cells (CD19+CD27hiCD38hi), increased expression of transcription factors IRF4 and PAX5.

Conclusion: The findings suggest an enhanced interaction between γδ T and B cells, favoring B cell differentiation and production of autoantibodies that might contribute to oral tissue damage in patients with BS.

CO2.1

Epigenetic regulation of thrombo-inflammation in Behçet and antiphospholipid syndrome

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Introduction: An epigenetic regulation of thrombo-inflammation has been reported in Behçet syndrome (BS), likely driven by a unique profile of three plasmatic circulating microRNAs (ci-miRNAs) (miR-206, miR-224-5p, and miR-653-5p). We compared these ci-miRNAs expression and the thrombogram profile in BS and antiphospholipid syndrome (APS), the prototype of acquired pro-thrombotic autoimmune disease.

Method: The three ci-miRNAs expression was evaluated by Poly(T)Adaptor PCR (PTA-PCR) in 39 patients with BS (including 22 with vascular involvement), 33 with APS (of whom 22 with thrombotic APS), and 30 healthy controls (HCs). Both single marker as well as combined ROC curve analyses were performed. Thrombin generation assay (TGA) was performed on pre-collected platelet-poor plasmas from 35 patients with BS and 77 patients with APS.

Discussion: The three ci-miRNAs, taken individually or combined, did not display acceptable discriminating power between study groups. Conversely, in the subgroups of BS and APS patients with vascular involvement (n=22 each), the combined signature displayed good discrimination capability between the two syndromes [specificity 0.91, sensitivity 0.77], as well as between thrombotic APS and HCs [specificity 0.77, specificity 0.87]. Also, distinct trends in thrombograms emerged between BS and APS, with BS TGA displaying lower tLag and tPeak and higher Peak as compared to APS.

Conclusion: Despite shared elements in the molecular regulation of their pro-thrombotic tendency, distinct epigenetic factors seem to contribute to the pathogenesis of vascular events in BS and APS, possibly accounting also for the different trends in thrombograms observed in the two syndromes.

CO2.2

HLA-B51 Positivity Correlates with Symptom Completeness from Recurrent Aphthous Stomatitis to Complete Behçet's Disease

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Introduction: The objective of this paper is to elucidate the positive correlation between HLA-B51 positivity and 'symptom completeness'—as defined by the Japanese criteria—and to examine its association with various clinical manifestations in patients with Behçet's disease.

Method: We conducted a retrospective analysis on a cohort of 1119 Korean patients who were receiving care and follow-ups at a tertiary referral hospital in Seoul, South Korea. All patients had their HLA-B51 status confirmed through genetic testing.

Discussion: Among the 1119 patients, 26.54% were male and 73.46% were female. HLA-B51 positivity was found in 40.84% of the patients, while 59.16% were HLA-B51 negative. Based on the Japanese criteria, the distribution of HLA-B51(+) patients was as follows: 52.9% were classified as complete, 42.5% as incomplete, 36.4% as suspect, and 26.6% as possible. The frequency of genital ulcers and papulopustular lesions was higher in HLA-B51(+) patients (OR: 1.507, 95% CI: 0.539 - 0.645) and (OR: 1.245, 95% CI: 1.237 - 2.122), respectively. Conversely, gastrointestinal involvement was lower in HLA-B51(+) patients (OR: 0.66, 95% CI: 0.131 - 0.334), and joint involvement (arthritis) showed no significant effect of HLA-B51 positivity (OR: 0.998, 95% CI: 1.071 - 1.907).

Conclusion: Despite the limitations of being a single-center study, our research provides significant insights through the use of a large patient cohort. The findings demonstrate a high positive correlation between HLA-B51 positivity and the presence of genital ulcers and skin manifestations (e.g., papulopustular lesions), suggesting that HLA-B51 positivity increases the likelihood of achieving a higher level of symptom completeness as outlined by the Japanese criteria. This study not only replicates results observed in previous researches on symptom clustering but also reinforces the evidence of the relationships between various symptoms in Behçet's disease.

CO2.3

Association of HLA-B5, HLA-B51, and HLA-B27 with Clinical Manifestations in Iranian Behcet's Disease Patients

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Introduction: Behcet's disease (BD) is a chronic inflammatory disease with unknown etiology, likely influenced by genetic and environmental factors. HLA-B51, a gene associated with immune function, strongly associates with BD, particularly in populations along the "Silk Road." This study investigates potential relationships between HLA-B27, HLA-B5, and HLA-B51 and their clinical manifestations in BD patients in Iran.

Method: This cross-sectional study reviewed records of 1164 BD patients diagnosed using International Criteria for Behcet's Disease (ICBD). Demographic data, clinical findings including specific BD manifestations (aphthous ulcers, genital ulcers, ocular involvement, skin lesions, etc.), laboratory results, and HLA-B5/B51/B27 status were collected. Statistical analysis explored relationships between variables and HLA markers.

Discussion: The study confirmed a high prevalence of HLA-B51 (42.01%) in BD patients, supporting its established association with the disease. Interestingly, HLA-B51 positivity correlated with an earlier age of symptom onset. Additionally, HLA-B5 and HLA-B51 positivity were significantly associated with recurrent ocular symptoms. Analysis of other BD manifestations revealed no significant association with HLA-B27. Notably, patients with a positive family history of BD displayed a significantly higher prevalence of both HLA-B5 and HLA-B51 compared to those without a family history. Gender analysis showed no significant influence of sex on HLA-B5 or HLA-B51 prevalence, but HLA-B27 was more frequent in males.

Conclusion: This study strengthens the link between HLA-B51 and BD, suggesting a potential role in earlier disease onset and susceptibility based on family history. Furthermore, HLA- B5/B51 positivity emerged as a risk factor for recurrent ocular complications. Future research should elucidate the underlying mechanisms and explore generalizability across ethnicities, while also investigating the potential influence of HLA markers on other BD manifestations.

CO2.4

Influence of sex on Behçet's Disease phenotypes: Data from the International AIDA Network Behçet's Disease Registry

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Introduction: Sex impact on phenotypical expression of Behçet's disease (BD) has been specifically investigated only in a few large-scale studies. The main goal of the study was to examine sex differences in a large cohort of patients affected by BD.

Method: Data were retrieved from the AIDA Registry for BD. We assessed differences between males and females in terms of BODI, disease manifestations and cardiovascular risk. Predictive factors leading to major organ involvement were also investigated.

Discussion: In total, 1024 BD patients (567 males, 457 females) were enrolled in the study, with a male-to-female ratio of 1.24/1. Males displayed a significantly higher mean ± SD BODI (1.92 ± 2.09) at the last follow-up, compared to female patients (1.25 ± 1.87) ($p<0.0001$). Uveitis ($p<0.0001$) and vascular involvement ($p=0.0076$) were significantly more frequent among males whereas female patients were significantly overrepresented in arthralgia ($p<0.0001$), arthritis ($p=0.00025$), isolated headache ($p<0.0001$), central nervous system (CNS) involvement ($p=0.040$), and gastrointestinal involvement ($p=0.00046$). Regarding cardiovascular risk, no differences between the two groups emerged ($p=0.617$). Four variables were associated with the development of major organ involvement: male sex (OR=2.104, $p=0.001$), current treatment with biologic agents (OR=2.257, $p=0.0003$), origin from endemic countries (OR=2.661, $p=0.0009$), and disease duration (OR=1.002, $p=0.024$).

Conclusion: BD displays a more severe course among males. This subgroup develops more irreversible damage and presents more frequently ocular and vascular involvement during disease course. On the other hand, female patients are prone to experience articular involvement, headache, CNS and gastrointestinal involvement. These data suggest the existence of a sex-driven disease expression.

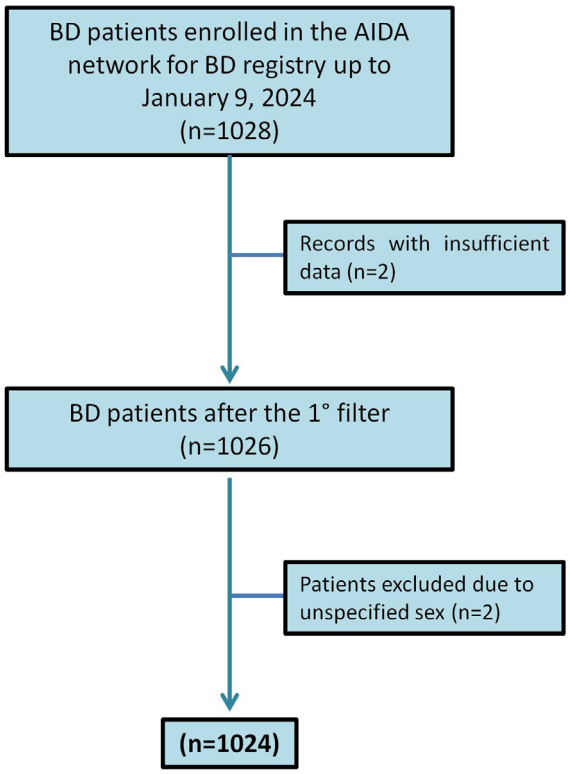

Figure 1. Chart showing the selection of the cohort.

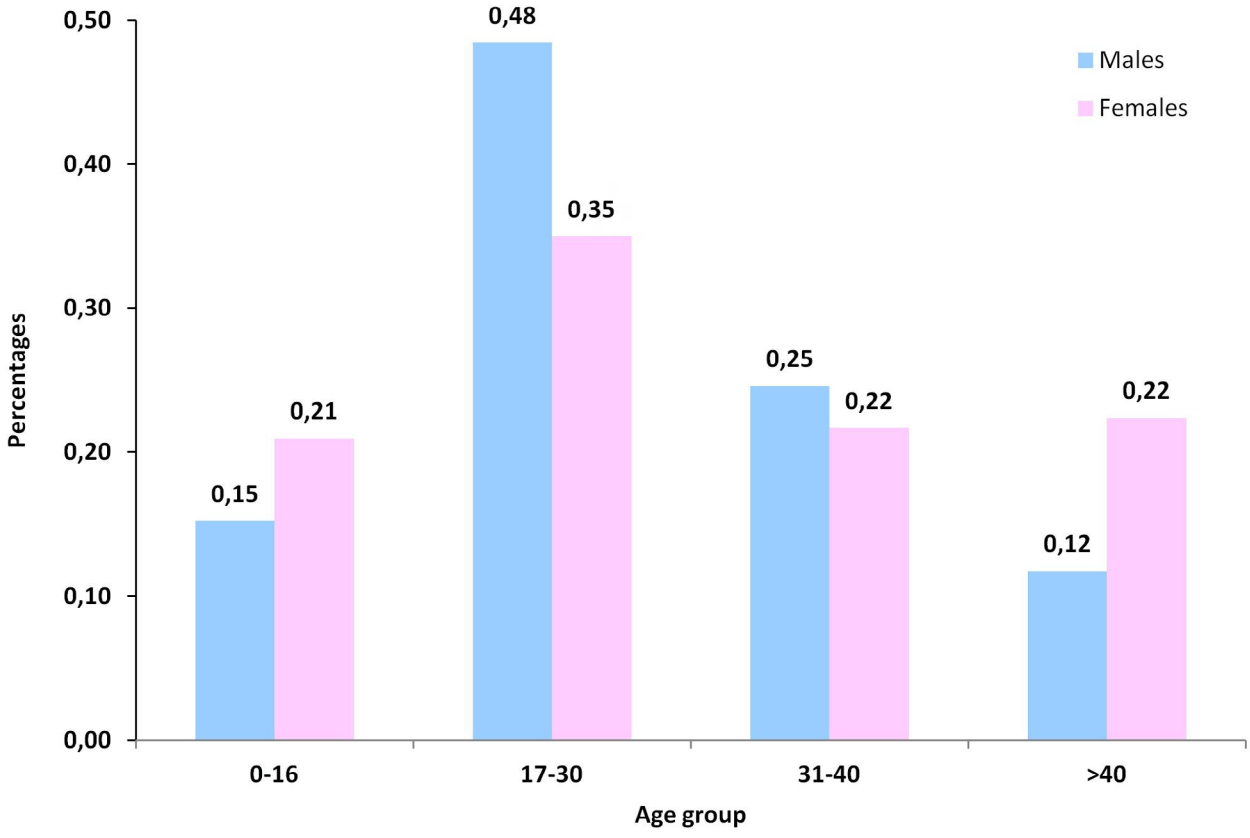

Figure 2. Distribution of age at onset (expressed in years) separated by sex for Behçet's disease patients according to different age groups.

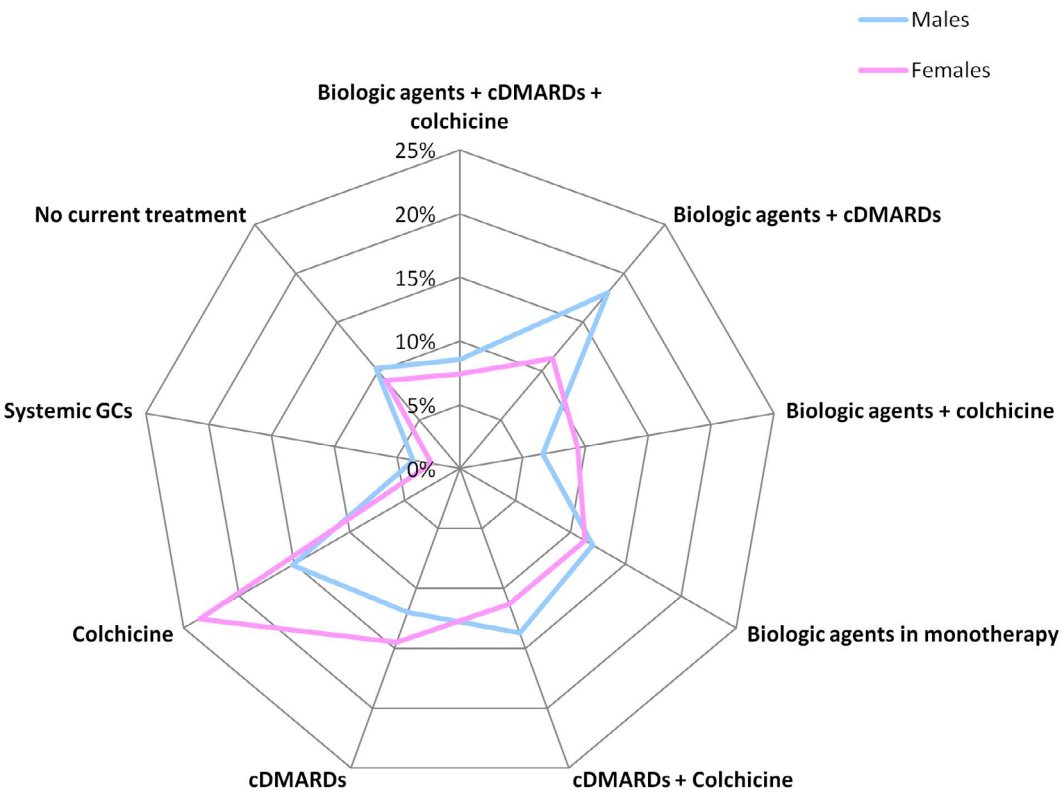

Figure 3. Treatment modalities of Behçet's disease patients recorded at the last follow-up visit, separated by sex.

CO2.5
Clinical features and phenotypic similarities of patients with familial Behcet's Disease

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Introduction: Phenotypic diversity is particularly noticeable in Behçet's disease (BD). The genetic basis of phenotypic variations in BD has not yet been elucidated. Based on the high frequency of familial BD, we aimed to analyse the familial aggregation of various manifestations of BD in this study.

Method: Patients with BD from three Turkish tertiary rheumatology outpatient clinics were evaluated. Demographic and clinical characteristics of the familial group with either first- or second-degree relative with BD and non-familial group were compared. Afterwards, patients in the familial disease group for 5 years or longer were divided into two an "index patient" and "first-degree relative patient" and the presence of BD manifestations was compared between these two groups.

Discussion: We identified 864 BD patients (mean age (SD): 47.9 (12) years, disease duration (SD): 83.7 (65.3) months with 251 (29.1%) having a BD family history. Genital ulcers (p=0.002) and papulopustular lesions (p<0.001) were detected more frequently in the familial group. Also in the familial group, statistically significant correlations were detected between the index patient and the first-degree relative-patient in terms of erythema nodosum-like lesion (r: 0.398, p: 0.016), pathergy test positivity (r: 0.561, p: 0.002), peripheral joint involvement (r: 0.563, p < 0.001) and vascular involvement (r: 0.408, p: 0.014) (**Table 1**).

Conclusion: Familial BD may difference from sporadic BD. Additionally, erythema nodosum-like lesions, pathergy test positivity, and vascular and joint involvement may tend to show familial aggregation.

	Frequency of presence in index group	*Frequency of presence in first degree relatives-group*	*Frequency of co-occurrence in both groups*	*Frequency of co-occurrence and co-absence in both groups*	*Odds ratio* (95% CI)*	*r*	*p*
Gender (Male)	26 (72.2)	27 (75)	22 (61.1)	27 (75)	6.3 (1.3-31.1)	***0.358***	***0.032***
Oral aphthous ulcers	36 (100)	36 (100)	36 (100)	36 (100)		-	-
Genital ulcers	28 (77.8)	33 (91.7)	26 (72.2)	27 (75)	1.9 (0.2-23.6)	0.081	0.64
Papulopustular lesions	21 (58.3)	25 (69.4)	14 (38.9)	18 (50)	0.7 (0.2-3.1)	-0.071	0.679
Erythema nodosum like lesions	13 (36.1)	13 (36.1)	8 (22.2)	26 (72.2)	5.8 (1.3-25.6)	***0.398***	***0.016***
*Pathergy positivity***	18 (62.1)	18 (62.1)	15 (51.8)	23 (79.3)	13.3 (2.2-82)	***0.561***	***0.002***
Major organ involvement	22 (61.1)	16 (44.4)	10 (27.8)	18 (50)	1.1 (0.3-4.3)	0.025	0.883
Uveitis	8 (22.2)	7 (19.4)	3 (8.3)	27 (75)	3.6 (0.6-21.4)	0.244	0.152
Peripheral joint involvement	17 (47.2)	13 (36.1)	11 (30.6)	28 (77.8)	15.6 (2.7-91.6)	***0.563***	***<0.001***
Vascular Behcet's Disease	14 (38.9)	6 (16.7)	5 (13.9)	26 (72.2)	11.7 (1.2-114.6)	***0.408***	***0.014***
Neuro-Behcet	3 (8.3)	0 (0)	0 (0)	33 (91.7)			
Entero-Behcet	3 (8.3)	5 (13.9)	0 (0)	28 (77.8)		0.121	0.482

*: Odds ratio of a first-degree relative having the same phenotype as the index case is given, **: Pathergy test results were found in only 58 patients' (29 in each group)

Table 1. Analysis of similarity of Behcet's disease manifestations within the families.

CO3.1

Phenotypic and Functional Analysis of NK Cell Populations in Behçet's Uveitis: Effects of Adalimumab Treatment and Comparative Inflammatory Responses with Axial Ankylosing Spondylitis

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Introduction: The aim of this study was to investigate changes in NK cell populations among Behçet's uveitis patients following adalimumab treatment.

Method: Blood samples were collected during active disease and upon clinical remission (3 months after adalimumab treatment). PBMCs were analyzed using flow cytometry, and plasma cytokine levels were measured using multiplex bead array technology.

Discussion: The study included 14 Behçet's uveitis (BU) patients, 13 axial ankylosing spondylitis (AS) patients as diseased controls, and 23 healthy individuals. Significant improvements were observed in BU patients' anterior chamber cells, angiography scores, visual acuity, and macular thickness, and in AS patients' BASDAI scores. Activator KIR NKG2D expression increased while inhibitor KIR NKG2A decreased during active disease in both BU and AS groups. After adalimumab treatment, NKG2D expression decreased and NKG2A increased in both groups. During active disease, BU patients exhibited elevated IL-10, IL-4, TGF-β, and IL-17 levels compared to healthy controls (HC), while AS patients showed higher levels of IFN-γ, TNF-α, IL-4, TGF-β, and IL-17. Active BU patients also had higher IL-2, IL-10, TNF-α, IFN-γ, and sFasL levels compared to active AS. Post-adalimumab treatment, plasma levels of IL- 2, TNF-α, IFN-γ, and sFasL remained higher in BU compared to AS patients, with a slight elevation in IL-10 levels.

Conclusion: Both Behçet's disease and AS are MHC class-I associated inflammatory diseases with overlapping pathways. Adalimumab enhanced inhibitory KIR responses and diminished activatory KIR responses. Reflecting GWAS data, active BU patients exhibited heightened IL-10 and IL-17 responses (NKreg and NK17), while AS patients were more inclined to TNF-α and IFN-γ (NK1) responses. Adalimumab reinforced IL-10 in BU, suggesting modulation of MHC/NK cell interactions. Systemic cytokine levels showed greater inflammation in BU than AS, even after adalimumab treatment.

CO3.2

The BD immune landscape exhibits a universal NF-kB-mediated hyperactivation pattern with cell-specific TNFAIP3 responses and a superimposed IFN-regulated endotype

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Introduction: The immune landscape of BD remains ill-defined, lacking a unifying picture, precise disease phenotype-endotype correlations, and an understanding of immunity at target tissue sites. Here, we aim to deconstruct peripheral and target organ-specific immunotypes of BD to define their global and endotype-specific applicability.

Method: We performed single-cell RNA sequencing and multiparametric flow cytometry on anterior chamber fluid cells and autologous and allogeneic PBMC, objected plasma to Luminex, and functional immune-phenomics studies on human monocytes and monocyte-derived dendritic cells (DC). We included 26 untreated BD and 22 age and sex-matched HD for single cell and an additional 15 SLE control and 11 HD for functional studies.

Discussion: We observed global NF-kB activation across all myeloid and T cell subsets with enrichment of classical dendritic cells (cDC) 2 and cDC1 in the eye and increased numerical representation of activated classical and non-classical monocytes in peripheral blood (PB) in BD. TNFAIP3 (A20) upregulation occurred in BD donors versus HD and in all myeloid cell types, NK, and NKT cells, but not in canonical T cells. NF-kB or IFN-mediated monocyte activation was not dependent on soluble factors from BD plasma. Plasmacytoid dendritic cells (pDC) were present in the eye during uveitis, and myeloid cells of uveitic BD patients had a superimposed IFN signature with increased numbers of pDC in PB.

Conclusion: NF-kB-dependent activation globally transcends all myeloid and T cell subsets in BD. Myeloid-centric upregulation of TNFAIP3 transcription points to lineage-dependent counter-regulation of inflammatory activity, and A20 upregulation in BD conceptually differentiates BD from A20 haploinsufficiency (HA20). We also define a superimposed, IFN-dependent activation profile that includes pDC and subsets of myeloid cells and forms a distinct uveitis endotype within BD, providing rationales for disease-wide and endotype and cell-specific therapeutic targeting.

CO3.3

Impact of HLA-B51 on Uveitis and Retinal Vasculitis: Data from the AIDA International Network Registries on Ocular Inflammatory Disorders

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Introduction: The clinical relevance of human leukocyte antigen (HLA) subtypes such as HLA-B51 on Behçet's disease (BD)-related uveitis and non-infectious uveitis (NIU) unrelated to BD remains largely unknown.

Method: Data were prospectively collected from the International AIDA Network Registry for BD and for NIU. We assessed differences between groups (NIU unrelated to BD and positive for HLA-B51, BD-related uveitis positive for HLA-B51 and BD-related uveitis negative for HLA-B51) in terms of long-term ocular complications, visual acuity (VA) measured by best corrected visual acuity (BCVA), anatomical pattern, occurrence of retinal vasculitis (RV) and macular edema over time.

Discussion: Records of 213 patients (341 eyes) were analyzed. No differences in complications were observed ($p = 0.465$). With regard to VA, a significant difference was detected in median BCVA ($p = 0.046$), which was not maintained after Bonferroni correction ($p = 0.060$). RV was significantly more prevalent in NIU-affected patients who tested positive for HLA-B51, irrespective of the systemic diagnosis of BD ($p = 0.025$). No differences emerged in the occurrence of macular edema ($p = 0.99$).

Conclusion: Patients with NIU testing positive for HLA-B51 exhibit an increased likelihood of RV throughout disease course, irrespective of a systemic diagnosis of BD. The rate of complications as well as VA are comparable between NIU cases unrelated to BD testing positive for HLA-B51 and uveitis associated with BD. Therefore, it is advisable to perform the HLA-B typing in patients with NIU or retinal vasculitis, even in the absence of typical BD features.

CO3.4

Common Femoral Vein Wall Thickness is a useful diagnostic tool to differentiate Ocular Behçet's Disease from Other Inflammatory Uveitis

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Introduction: Diagnosis of Behçet's Disease (BD) can be challenging. We showed that common femoral vein (CFV) wall thickness can be diagnostic for the cut-off value ≥0.5mm. It is crucial to make a correct diagnosis since ocular BD might cause permanent vision loss if left untreated. In this study, we focused on assessing the usefulness of CFV wall thickness measurement in BD-related uveitis (BU).

Method: 45 BD panuveitis and 72 non-BD patients diagnosed with other infectious and inflammatory uveitis were recruited. A radiologist measured CFV wall thickness and a rheumatologist for differential diagnosis evaluated them on the same day.

Discussion: The clinical features and demographics of two groups are shown in **Table 1**. The mean age was similar among patients. The majority of the patients had panuveitis (70, 59.8%), followed by anterior uveitis (32, 27.4%) and intermediate uveitis (10, 8.5%). All the BU patients had panuveitis. Among the non-infectious cases, BD was the leading diagnosis. 26 patients (22.2%) had idiopathic uveitis, 15(12.8%) had sarcoidosis,10(8.5%) had ankylosing spondylitis,9(7.7%) had HLA-B27 positive uveitis .1 patient (0.9%) had herpetic uveitis. The mean CFV thickness of the BU group was significantly higher compared to the non-BU group for both legs (for the right leg, 0.74(0.08) vs 0.45 mm(0.08); for the left leg, 0.74(0.07) vs 0.45 mm(0.08), $p<0.001$ for both) (**Figure 1**).We observed that the CFV measurements of 10 patients from the non-BU group were above 0.5 mm cut-off value. After rheumatologic examination, 5 patients fulfilled one of the diagnostic criteria sets (fulfillment of only ICBD for 4 patients and of both ICBD and ISG for 1 patient). Although 3 patients have not fulfilled one of the diagnostic criteria sets, they have been diagnosed with BD by expert opinion.

Conclusion: Our results suggest that measurement of CFV wall thickness by Doppler US can be helpful for the differentiation of ocular BD from other causes of inflammatory uveitis in daily practice.

	Behçet's Uveitis *(n =45)*	Non-Behçet Uveitis *(n =72)*
Age (years), mean (SD)	38.1 (10.3)	41.7 (12.6)
Sex, n (%)		
Male	23 (51.1)	24 (33.3)
Female	22 (48.9)	48 (66.7)
BMI, kg/m2, mean (SD)	24.9 (3.4)	28.3 (5.4)
Duration of disease (years), mean (SD)	8.8 (8.4)	6.3 (5.4)
The number of uveitis attacks, median (min-max)	3 (1-10)	2 (1-6)
Laterality of uveitis, n (%)		
Unilateral	30 (69.8)	52 (75.4)
Bilateral	13 (30.2)	17 (24.6)
Ocular symptoms at presentation		
Decreasing of visual acuity, n (%)	28 (87.5)	45 (78.9)
Hyperemia, n (%)	9 (28.1)	20 (35.1)
Painful vision, n (%)	5 (15.6)	15 (26.3)
Diplopia, n (%)	4 (12.5)	4 (7)

Table 1. Baseline demographics and disease characteristics of Behçet's uveitis and non-Behçet uveitis patients.

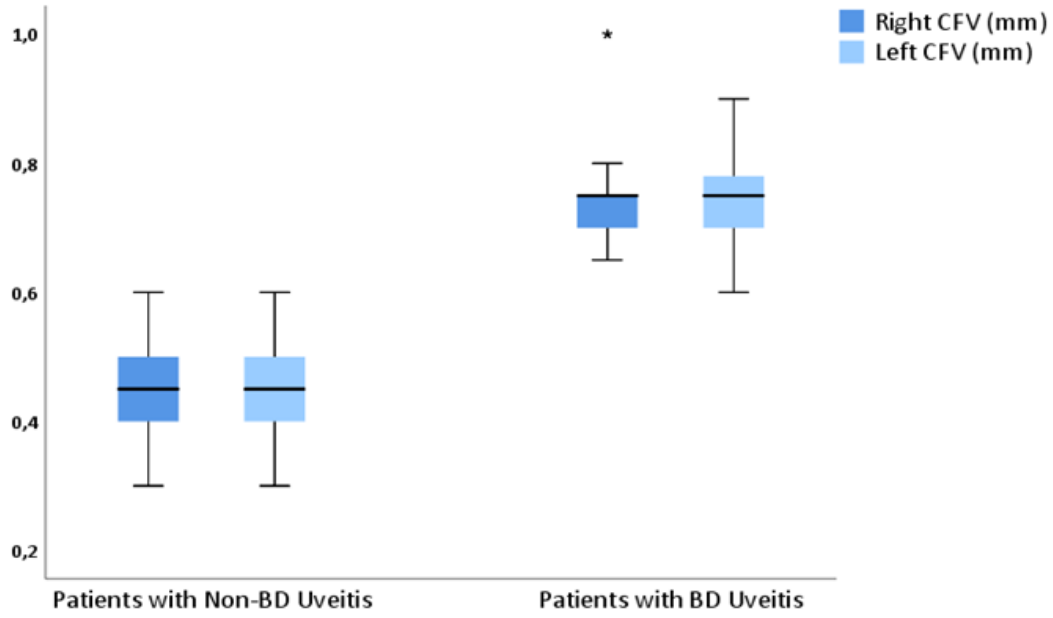

Figure 1. Comparison of common femoral vein wall thickness of the patients with Behçet's uveitis (BU) and non-Behçet's uveitis (non-BU) for both legs.
CFV: common femoral vein.

CO3.5
Long-term effect of anti-TNF therapy in patients with Behçet uveitis

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Introduction: The aim of the study is to evaluate the effect of anti-tumor necrosis factor (TNF) treatment on visual acuity and central macular thickness (CMT) in patients with Behçet uveitis.

Method: Patients who were started on anti-TNF due to Behçet uveitis were included. 52 eyes of 28 patients were included. The patients' baseline examination and their best corrected visual acuity (BCVA) and CMT at the 3rd, 6th, 12th and 24th months were recorded.

Discussion: 52 eyes of 28 patients (15 male, 13 female) were included. The average age was 38.29 years. Average follow-up time was 66.88 months. The average duration of follow-up of patients was 97.58 months; the average duration of anti-TNF use was 27.12 months. Adalimumab was started in 19 patients and infliximab was started in 9 patients. A switch was made between the treatments in 5 patients. There was a significant difference in the number of annual attacks of patients before and after the start of treatment. ($p<0.05$). When baseline BCVA was compared one by one at 3, 6, 12 and 24 months, post-treatment BCVA was better in each ($p<0.05$). The 6th month was seen as a significant breaking point in terms of treatment results. When the initial CMT was compared one by one at the 3rd, 6th, 12th and 24th months, there was a significant decrease in all visits except the 3rd month ($p<0.05$). Similar to visual acuity, the 6th month is a critical time when the median visual acuity reaches 20/20 logMAR and the central macular thicknesses reaches normal macular thicknesses.

Conclusion: When BCVA and CMT were evaluated sequentially at baseline, 3rd and 6th months, a significant increase in visual acuity and a significant improvement in central macular thickness were observed ($p<0.05$).

CO4.1

Prospective Evaluation of Neurological Presentations to a National Neuro-Behçet clinic; Migraine is the commonest neurological complaint in Behçet's Disease

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Introduction: Behçet's disease (BD) is a rare relapsing multisystem inflammatory vasculitis which is more common along the silk road. The aetiology is poorly understood. Neuro- Behçet's disease (NBD) is thought to affect 5-10% of patients. It is associated with high morbidity and mortality. NBD can affect the central nervous system (CNS) and the peripheral nervous system (PNS). CNS involvement is more common, more frequent in males and can be classified into parenchymal and non-parenchymal. As part of collaborative working between the Walton Centre NHS Trust and Aintree University Hospital, we run a dedicated monthly Neuro-Behçet's clinic (NBC). Migraine is a prevalent condition affecting 1 in 7 of the global population with chronic migraine affecting 1.4–2.2%. Severe migraine attacks are associated with significant disability and considerable socioeconomic impact, ranking in the highest disability category (class 7) by WHO (4,5,6).

Method: We conducted a prospective evaluation of all patients who were seen in the NBD clinic over a seven-month period from December 2021 to June 2022, inclusive.

Discussion: We reviewed 24 patients in the NBC (F:M 19:5, age range between 20-59, of the total of 249 (10%) patients who were seen in the Behçet's clinic. Only 4 patients (2% of total, 17% of those seen in (NBC) fulfilled the criteria for NBD. By far the commonest presentation to the NBC was headache; 20 of the 24 patients (83%). One patient had more than one headache phenotype; majority (18 patients- 90%) had chronic migraine (CM).

Conclusion: Based on our findings, NBD remains a rare complication of BD (2%). Majority of patients who presented to our NBC were diagnosed with migraine (83%). Given the prevalence of migraine in the general population, further studies would be required to ascertain the correlation to BD and whether headache could be classified as manifestation of NBD or just as a prevalent co-existing condition, possibly exacerbated by use of analgesia.

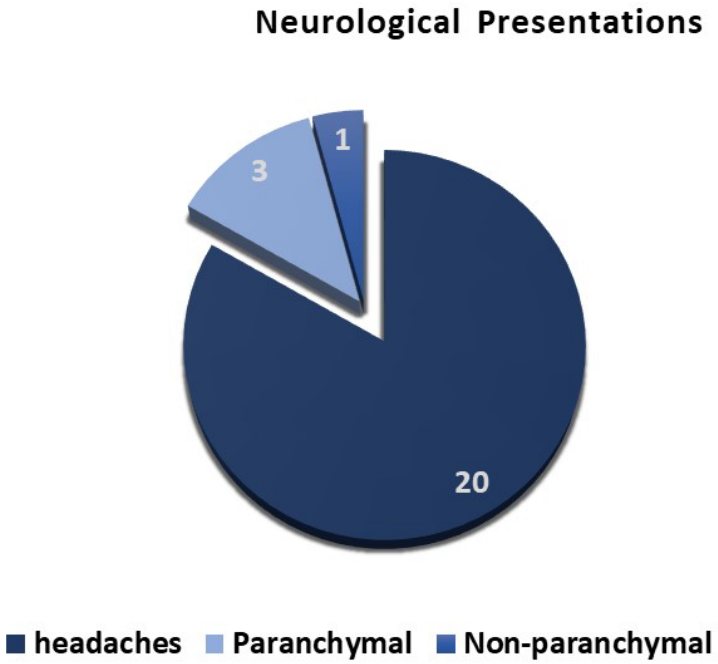

Figure 1. Presentation to the neuro- Behçet clinic.

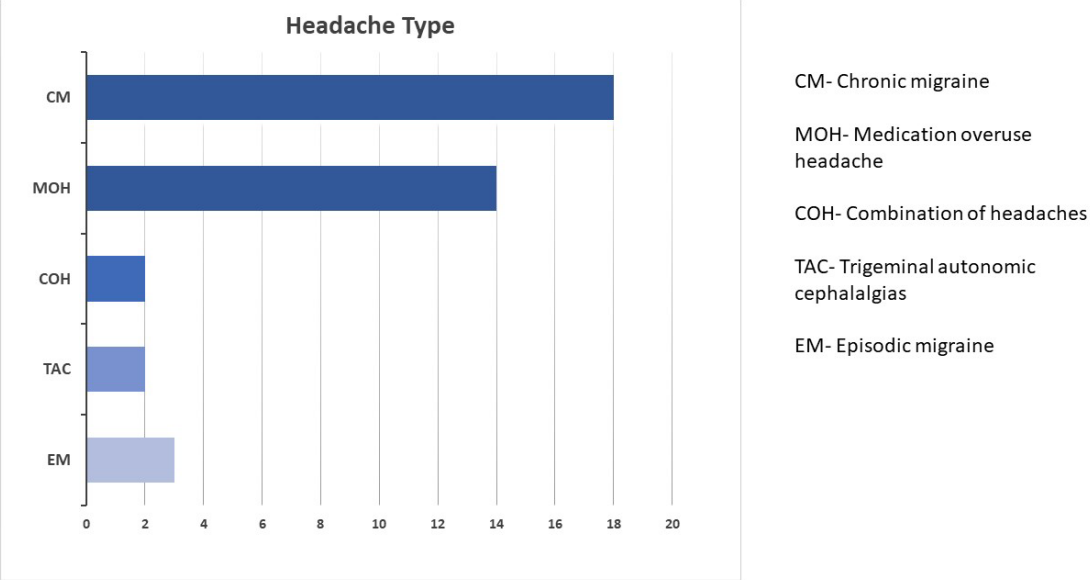

Figure 2. Headache phenotypes.

CO4.2

Neuro-Behçet in its parenchymal form: Study of clinical, radiological, therapeutic, and evolutionary profile (About 39 cases)

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Introduction: The neurological manifestations of Behçet's disease exhibit a wide clinical polymorphism and are among the most severe manifestations of the disease as they impact both the vital and functional prognosis. The aim of the present study is to analyze the prevalence, clinical manifestations, treatment, and evolution of neurological episodes in a cohort of patients with parenchymal Neuro-Behçet.

Method: This was a descriptive and retrospective study involving 39 patients followed for neuro-Behçet in its parenchymal form out of a series of 49 patients followed for neuro-Behçet at the Neurology Department of CHU HASSAN II in Fes over a period of 5 years, from January 2018 to January 2023.

Discussion: We identified 27 men and 12 women. The average age was 37 years. The onset was poly-symptomatic for 65% of patients. Motor deficit was reported by 92.30% of patients; 19 patients (48.72%) complained of gait disturbances and 15 (38.46%) of headaches. The most commonly found initial signs consisted of motor disorders (46.15%), headaches (17.95%), and balance disorders (12.82%). Serum inflammatory markers were positive in 51.28% of cases. The types of MRI lesions found consisted of disseminated demyelination, located in the brainstem (92.30%) predominantly pontine, cerebellum (38.82%), capsule (20.59%), thalamus (17.65%), subcortical hemispheric white matter, and central gray nuclei in 8.82% for each.

Conclusion: The neurological manifestations of Behçet's disease can be potentially severe, primarily characterized by involvement of the brainstem. Parenchymal forms vary and may be difficult to recognize promptly; prognosis is particularly severe, especially in cases involving the brainstem. Early management and the conduct of prospective, multicenter studies are essential to develop validated therapeutic strategies aimed at improving patient prognosis.

CO4.3

The factors associated with cognitive disorders in Behçet's Disease patients

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Introduction: the cognitive disorders (CD) is a psychopathological problem for Behçet's Disease (BD) patients. CD has high rates in BD, but their causes aren't enough investigated. The aim of the study was to determine the main factors associated with CD in BD patients

Method: 116 BD patients were enrolled in the study. The majority of patients were men (69,8%), natives of the North Caucasus (51,9%), with mean age (M±m) 33,3±0,98 years. All the patients met the ICBD 2014 criteria. The disease activity was assessed by scoring system BDCAF. Mental disorders (MD) were diagnosed by psychiatrist in accordance with the ICD-10 in semi-structured interview. CD were diagnosed with psychology and neuropsychology methods.

Discussion: CD were diagnosed in 91 BD patients (78,4%). The mechanical memory (50%) and attention deficit (80,5%) were the most frequent manifestations of CD, impairment of associative memory (31,7%) and logical thinking (36,6%) were less frequent. The presence of CD didn't depend on BD activity, severity and duration, as on patient's ethnicity, use of prednisone and immunosupressive agents. The patients with CD were older (34,3±1,07 vs 29,0±2,14, p=0,006), more often had chronic/recurrent depressive disorders (84,1% vs 50,0%, p=0,001) and chronic stressful life events (91,5% vs 62,5%, p=0,001). Then linear regression analysis was done and obtaining prognostic model showed that CD was associated with: stress factors (β=0,497), noncompliance (β=0,136), male gender (β=-0,068), higher cholesterol level (β=0,053), anxiety/depressive disorders levels (HADS-A>=8 (β=0,156) and HADS- D>=8 (β=0,02)), high disease activity by BDCAF scale (β=0,020) and older age of patients (β=0,007) (area under the ROC curve = 0,848)

Conclusion: the results demonstrated high prevalence of CD in surveyed BD patients. CD in BD is much more associated with stress factors, noncompliance, male gender, higher cholesterol level, anxiety/depressive disorders, high BD activity and older age of patients.

CO4.4

Evaluation of Optic Disk and Parafoveal Parameters in Neuro-Behçet Patients Without Ocular Involvement Using Optical Coherence Tomography Angiography

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Introduction: The study aims to examine vascular changes in the optic disc and macular region using Optical Coherence Tomography Angiography (OCTA) in patients with Neuro- Behçet's disease without ocular involvement.

Method: 36 eyes of 19 nonocular parenchymal Neurobehçet patients (NBG), 44 eyes of 22 nonocular Mucocutaneous Behçet's disease patients (MBG), and 51 eyes of 26 healthy individuals (HG) were included. OCTA measurements were performed in all groups.

Discussion: In the comparison of peripapillary retinal nerve fiber layer (RNFL) thickness, the NBG had the lowest mean, while no significant difference was observed between the groups. In the analysis of peripapillary vascular density (VD), the mean VD in the inferior quadrant was significantly lower in NBG than in the other two groups (p = 0.014). In the foveal region, both the superficial capillary plexus (SCP) and deep capillary plexus (DCP) VD means were significantly lower in NBG compared to MBG (p=0.019 and p=0.02, respectively). Additionally, NBG was observed to have significantly wider foveal avascular zone compared to MBG (p=0.035). In the assessment of choriocapillaris flow rate, the mean values for NBG in both the 1mm and 6mm diameter (subfoveal and submacular) areas were significantly lower than those in other groups (p=0.002, p=0.000, respectively).

Conclusion: The ability of OCTA to demonstrate changes in retinal and choroidal blood flow in Neuro-Behçet patients without ophthalmological symptoms or findings suggests the presence of subclinical effects and indicates the potential utility of OCTA in monitoring these patients.

CO4.5

Arterial involvement in Neurobehçet Syndrome: Vessel wall imaging as a diagnostic marker

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Introduction: The neurologic involvement of Behçet Syndrome (BS) is termed as Neuro-Behçet's Syndrome (NBS). Directly neurological involvement of BS may be classified into two forms: 1) parenchymal (p-NBS); 2) vascular involvement. Arterial involvement is rare in NBS, with limited data on this subject. This study aims to investigate vascular wall imaging (VWI) findings in intracranial arteries in NBS cases together with magnetic resonance (MR) and magnetic resonance angiography (MRA).

Method: 39 NBS patients were included in the study and VWI and contrast-enhanced cranial MRI and MRA were performed. Intracranial arteries were divided into a total of 36 separate segments and evaluated with MRA and VWI. Segments were evaluated in 5 categories in terms of vessel wall involvement: Group 1: normal appearance; Group 2: concentric wall thickening with wall enhancement; Group 2a: only concentric wall thickening; Group 3: eccentric wall thickening with wall enhancement; Group 3a: only eccentric wall thickening.

Discussion: The median age was 40 (range 16-57) and 69.2% (n=27) of patients were men. MRI was performed in 16 patients during the attack. 92.3% (n=36) of patients had parenchymal involvement and 7,7% (n=3) of patients had CVST. Intracranial artery stenosis or occlusion was detected in 15.4% (n = 6) of 39 NBS patients on MRA. Abnormal vessel wall appearance was observed in 16 patients (41%) on VWI. In 1 of 3 patients (33%) with a spinal cord lesion, a vessel wall lesion was detected in at least 1 intracranial artery segment. No vessel wall lesions were detected in the intracranial arteries in any patient with CVST.

Conclusion: This study shows that intracranial arterial involvement in NBS is more common than previously reported. VWI may play a role in the diagnosis of vasculitides that may occur during BD and can be used as a complementary test in addition to luminal imaging methods.

CO5.1

Pulmonary Involvement in Behçet's Syndrome: A Systematic Review of the Literature

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Introduction: Pulmonary artery involvement (PAI) is a significant cause of morbidity and mortality in Behçet's syndrome (BS) and most commonly affects young males. Also, it occurs early in the disease course and clusters with venous vascular involvement. We aimed to demonstrate changing involvement patterns and outcomes of PAI in BS over decades.

Method: The PRISMA-IPD checklist guided the reporting of the data. The literature search was performed via PubMed/MEDLINE, EMBASE, and Web of Science between 1980 and November 2022. (ID: CRD42022374596).

Discussion: The incidence of isolated pulmonary artery thrombus (iPAT) increases over decades, and pulmonary artery aneurysms (PAA) decrease (Fig1). Patients with iPAT have substantially better survival than those with PAA+PAT or isolated PAA. This confers iPAT may be the precursor lesion of PAA and that progression to PAA, which is a more severe involvement, can be prevented with early diagnosis and treatment. On the other hand, while surgical treatment was significantly more common before 2000 (Fig2), the use of immunosuppressive therapies (glucocorticoids, cyclophosphamide, azathioprine, and anti-TNFs) considerably increased, accounting for decreased mortality rates (Fig3). Hence, this SLR supports the notion immunosuppressive therapy is the mainstay of treatment in PAI of BS. Also, patients who do not meet the classification criteria are not uncommon (20%), suggesting that the vascular phenotype is distinct from other phenotypes.

Conclusion: This SLR shows changing trends of pulmonary involvement in BS regarding the involvement types, treatment, and outcome.

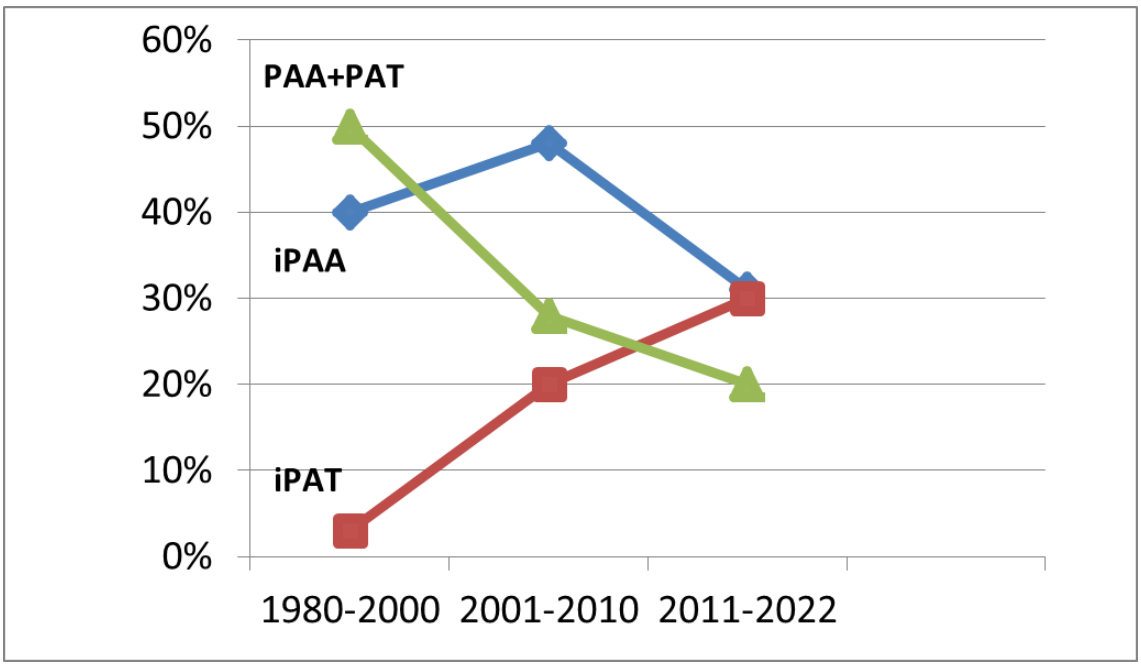

Figure 1. Changing trend of pulmonary involvement pattern over decades.

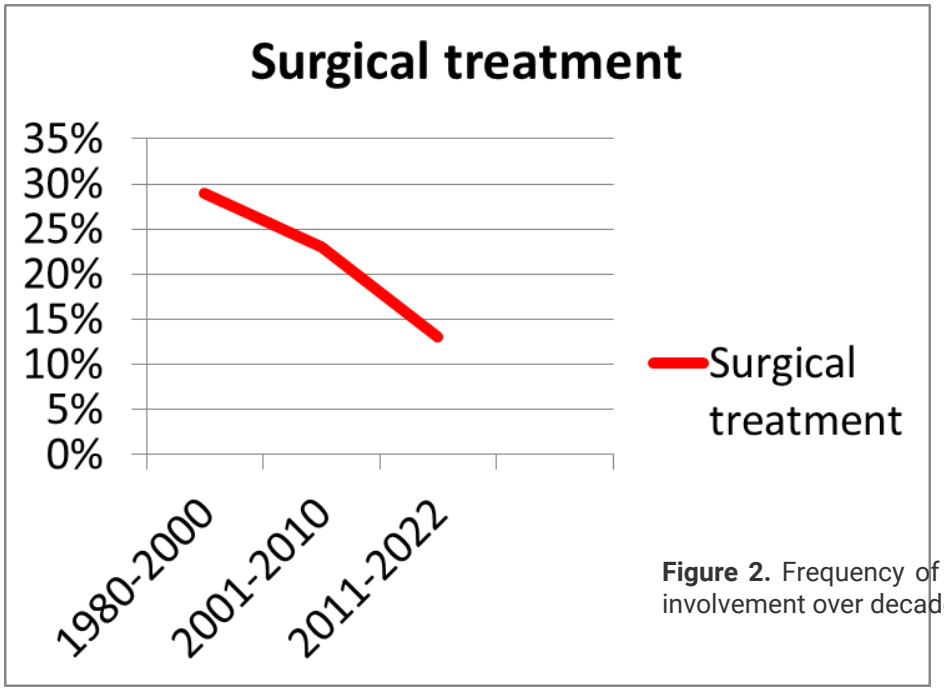

Figure 2. Frequency of surgical interventions for pulmonary involvement over decades.

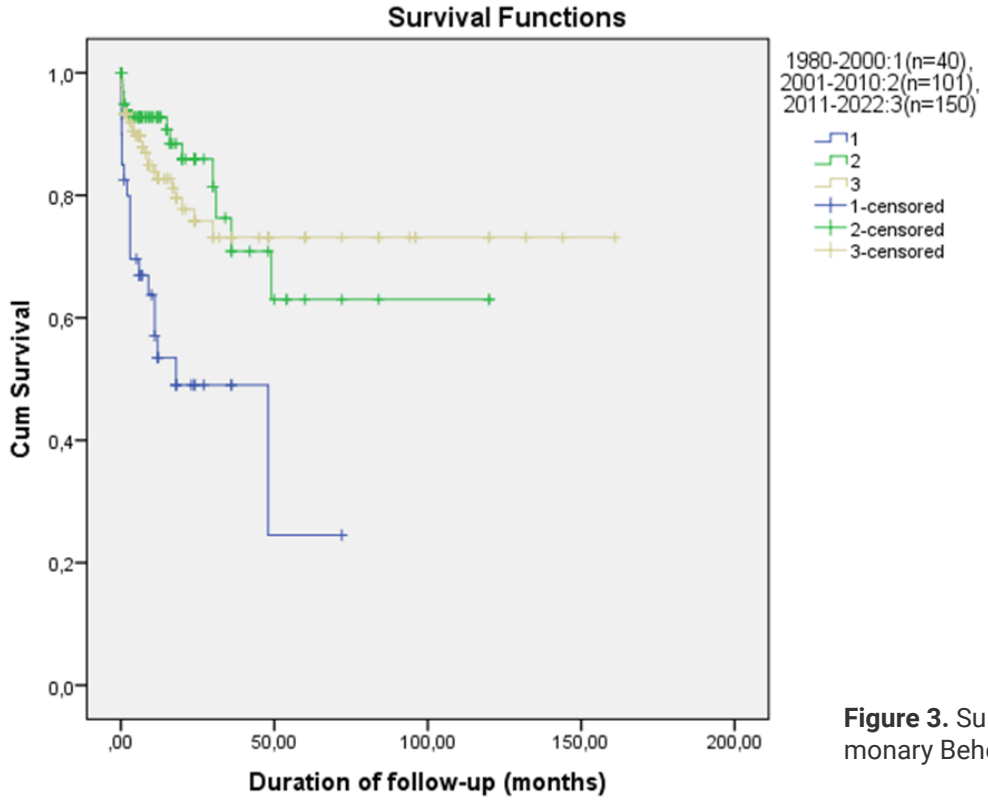

Figure 3. Survival analysis of patients with pulmonary Behçet's syndrome.

CO5.2

Diagnostic Value of Vein Wall Thickness Measurement for Patients with Suspected Behçet Syndrome

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Introduction: Increased venous wall thickness is well recognized in Behçet syndrome (BS). However, studies have shown conflicting results on the diagnostic utility of its measurement by ultrasonography. A good method for evaluating this would be measuring common femoral vein wall thickness (CFVWT) with Doppler ultrasonography among patients with suspected BS who are referred for screening.

Method: We included all individuals referred to our clinic for BS screening between May 2023 and December 2023. Thirty vascular BS patients with lower extremity deep vein thrombosis (LEDVT) and 31 healthy subjects were recruited as controls. Two radiologists independently measured CFVWT, blinded to the diagnoses.

Discussion: Among the 58 referred individuals, 30 were diagnosed with BS and 28 with other conditions (non-BS). LEDVT was present in 3/30 BS patients and in 2/28 non-BS patients. The highest median (Q1-Q3) CFVWT was observed in patients with vascular BS [0.65 mm (0.59-0.82) for right and 0.64 mm (0.56-0.76) for left], followed by those with BS [0.60 mm (0.55-0.69) for right and 0.64 mm (0.57-0.71) for left], non- BS [0.59 (0.48-0.66) for right and 0.58 mm (049-0.68) for left], and healthy subjects [0.49 (0.47-0.53) for right and 0.46 mm (0.43-0.51) for left]. Both right and left median CFVWT was significantly lower among healthy controls ($p<0.001$ for all), whereas there was no significant difference between patients with and without BS who were referred for screening. This was true even after excluding patients with LEDVT from both BS and non-BS groups. Interobserver reliability was excellent between the radiologists (ICC 0.94 for right and 0.96 for left CFV).

Conclusion: CFVWT was higher in both BS and non-BS patients compared to healthy subjects. However, its measurement did not help the differential diagnosis of new patients with BS from non-BS patients sent to our clinic with a suspicion of BS.

CO5.3

Stasis Ulcer in Behçet Syndrome: A Common but Difficult to Manage Complication

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Introduction: Post-thrombotic syndrome (PTS) and stasis ulcers (SUs) are late complications of deep vein thrombosis (DVT) in Behçet's syndrome (BS). This study aimed to determine the clinical and histopathological characteristics, treatment modalities, and outcomes in BS pts with vascular involvement and SUs.

Method: We studied 51 BS pts with SUs among 310 with vascular issues at a tertiary center. Clinical data, histopathology, imaging, and treatment details were obtained from records. Ulcer size, location, duration, and healing time were defined. Remission was defined as no active ulcer for a year.

Discussion: Patients' median age was 45 years, and age at vascular onset was 27 years. Ulcers appeared a median of 4 years after vascular involvement onset. Except for 3 pts with only venous insufficiency, all had lower extremity DVT (Table1). Active ulcers were found in 55% of the pts, and 54% had recurrent ulcers. Among the 51 pts, there were 165 ulcers; median ulcer size was 3 cm, with 41% on the medial malleolus and 25.5% on the anterior tibia. In 23 pts, ulcers healed in a median of 9 months (Table 2). Histopathological examination was available for 14 pts. In 71% (10/14), the diagnosis favored stasis dermatitis or occlusive vasculopathy. Treatment included bed rest, local treatments, and compression stockings. For SUs, 36 pts received immunosuppressive treatment (biological: 45%, non-biological: 63%), and/or glucocorticoids (28%). Anti-TNF agents were used in 23 pts (64%). Intravenous iloprost (49%), Lucilia Sericata for debridement (25%), and skin grafts (4%) were applied.

Conclusion: Ulcers were found not only on the medial malleolus and anterior tibia but also on the dorsum of the foot and lateral malleolus. Our series showed findings consistent with stasis dermatitis, unlike Jung et al. Despite treatment, ulcers remained active in 55% of pts, suggesting immunosuppressive treatment may not be effective in the late stage.

Tablo.1: The Clinical and Demographic Characteristics of Patients

Male n (%)	49 (%96)
Age	**Median:** 45, **Min:** 27 **Max:** 60
Educational Level n (%)	
- Elementary School	26 (%51)
- Secondary School	7 (%13)
- High School	13 (%26)
- University	5 (%10)
Occupation n (%)	
- Employed (Qualified Job-Unqualified Job)	2 (%4)-36(%71)
- Unemployed (Retired+Unemployed)	13 (%25)
Median Body Mass Index	**Median:** 28,8 **Min:** 19,1 **Max:** 44,2
Cigarette n (%)	
- Active Smoker	27 (%53)
- Non-Smoker	9 (%18)
- Former Smoker	15 (%29)
Median Age of Those Who Filled out the ISG (years)	**Median:** 25 **Min:** 12 **Max:** 46
Median Follow-up Duration (years)	**Median:** 15,5 **Min:** 1,7 **Max:** 29,5
Vascular Involvement Median Age (years)	**Median:** 27 **Min:** 16 **Max:** 46
Comorbidity n (%)	
- Hypertension	10 (%20)
- Coronary Heart Disease	8 (%16)
- Diabetes Mellitus	6 (%12)
- Hyperlipidemia	8 (%16)
- Others	17 (%34)
- No Comorbidity	26 (%52)
Central Nervous System Involvement n (%)	6 (%12) (5 Dural Venous Sinus Thrombosis), 1 (Parenchymal Involvement)
Eye Involvement n (%)	24 (%48) (23 Uveitis), (1 Retinal Vein Thrombosis)
Deep Vein Thrombosis n (%)	
- Bilateral	36 (%75)
- Unilateral	12 (%25)
- Femoral Vein	40 (%83)
- Popliteal Vein	35 (%73)
- Superficial Vein	22 (%46) (17 VSM, 5 VSP)
Vena Cava Inferior n (%)	12 (%25)
Iliac Veins n (%)	13 (%27)
Upper Extremity DVT n (%)	2 (%4)
Other Vessels n (%)	20 (%40) (3 VCS, 6 PAA, 6 PAT, 2 Per/AA, 2 Coronary Artery Involvement, 1 Carotis Artery Aneurysm)
Median CEAP Score	C5
CEAP: Clinic, Etiologic, Anatomic ve Pathophysiological Classification **Max:** Maximum, **Min:** Minimum, **PAA:** Pulmonary Arterial Aneurysm, **PAT:** Pulmonary Arterial Thrombosis, **Per AA:** Peripheral Arterial Aneurysm, **Upper Ex. DVT:** Upper Extremity Deep Vein Thrombosis, **VSM:** Vena Saphena Magna, **VSP:** Vena Saphena Parva, **VCS:** Vena Cava Superior	

Tablo.2: Stasis Ulcer and Its Characteristics

Stasis Ulcer			
- Median Time Interval Between Disease Onset and Initial Ulcer Development (years)	**Median:** 5 **Min:** 0, **Max:** 24		
- Time Interval Between Ulcer and Vascular Involvement (years)	**Median:** 4 **Min:** 0 **Max:** 23		
- Median Healing Time (months)	**Median:** 8 **Min:** 0,5 **Max:** 144		
- Median Cumulative Number of Ulcers	**Median:** 3 **Min:** 1 **Max:** 11		
- Ulcer Size	**Median:** 3x3 cm **Min:** 0.5x0.5 cm **Max:** 15x15 cm		
- Ulcer Location (n:165 ulcers)			
○ Medial Maleol	67 (%41)		
○ Lateral Maleol	11 (%6,5)		
○ Tibia Anterior Face	42 (%25,5)		
- Open/Healed Ulcer n (%)	28 (%55)/23(%45)		
- Median Time Interval Between Disease Diagnosis and Onset of the First Ulcer in the Group with Open Ulcers (years)	**Median:** 5 **Min:** 0 **Max:** 24		
- Median Time Interval from Onset of the First Ulcer to Present in the Group with Open Ulcers (years)	**Median:** 11,5 **Min:** 0,3 **Max:** 32		
Total Number of Patients (n: 51)	**Active Ulcer (n=28)**	**Healed Ulcer (n=23)**	**p**
Active Smoker, n(%)	14 (50,0)	13 (56,5)	0.642
Educational Status			
- Elementary-Secondary School, n(%)	18 (64,2)	13 (56,5)	0.572
Duration Disease, year (mean ± SD)	16,1 ± 6,6	14,6 ± 8,1	0.272*
Age of Disease Onset (median)(min-max)	25,5 (18-46)	24 (12-42)	0,429**
Time Interval Between Disease Onset and First Ulcer (years)(median) (min-max)	5 (0-24)	5,5 (0-24)	0,762**
BDCAF (median) (min-max)	1 (0-3)	0 (0-2)	0.071 **
Body Mass Index, (mean± SD)	29,6±4,4	25,9±4,1	0.608*
Time Spent Standing, hours (mean ± SD)	6,9±3,3	6.3±3,4	0.658
Those Using Biological Drugs, n(%)	19 (67,8)	15 (65,2)	0.842
Those Using Iloprost, n(%)	17 (60,7)	8 (34,7)	0.065
Those Proximal Deep Vein Thrombosis, n(%)	24 (85,7)	17 (73,9)	0.693
Mann-Whitney U test, * Student t test, **Min: Minimum, **Max:** Maximum			

CO5.4

Direct Oral Anticoagulants in Patients with Vascular Involvement of Behcet' s Disease: A single center experience

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Introduction: Bechet's disease (BD) is a chronic inflammatory disease that affects the vascular system in 40% of patients which is the most important cause of mortality and morbidity. Both arterial and venous systems can be involved leading to thrombosis and aneurysms. As the mainstay of therapy is immunosuppressive treatments, anticoagulants should be added in refractory thrombosis, taking into account the risk of bleeding, particularly in pulmonary aneurysms. Direct oral anticoagulants (DOACs) are commonly used for venous thromboembolism but there are no controlled studies for their use in BD. This study aims to evaluate the efficacy and safety data of DOACs in BD.

Method: Patients with Behcet's disease using DOACs for vascular involvement were evaluated retrospectively. The demographics, disease manifestation, vascular involvement, and treatments are screened from the patient files.

Results: Lower extremity DVT was the most common vascular involvement in patients treated with DOACs. Characteristics of patients are summarized in Table 1. In only one patient treated with DOACs, recurrent thrombosis was seen despite additional immunosuppressive treatment. Major bleeding events never occurred with DOACs. Only in 1 patient, gastrointestinal bleeding was observed under vitamin K antagonist that did not recur after switching to edoxaban.4 minor bleeding events occurred; 3 hemoptysis−1 with apixaban,2 with rivaroxaban, and 1 gum bleeding in a patient with rivaroxaban treatment. Discussion: Our study showed that patients with BD can be successfully and safely treated with DOACs. The most commonly used DOAC in our patients was rivaroxaban. Some case reports show similar results to our study that DOACs had an additional effect on persistent and recurrent thrombotic events.

Conclusion: While the mainstay of treatment of patients with vascular involvement of BD is immunosuppressive treatment, anticoagulant therapy may contribute to thrombus resolution, and DOACs may be effective and safe in appropriate patients.

Table 1. Characteristics of BD patients with vascular involvement

	n: 39
Age at diagnosis, median (min-max)	34 (16-56)
Male gender	21 (53%)
Disease manifestations	
Oral ulcer	37 (94%)
Genital ulcer	20 (51%)
Folliculitis	15 (38%)
Erythema nodosum	13 (33%)
Articular involvement	13 (33%)
Ocular involvement	14 (35%)
Neurological involvement	6 (15%)
Vascular involvement	
Isolated venous thrombosis	18 (46%)
Isolated arterial thrombosis	17 (43%)
Combined arterial and venous thrombosis	4 (10%)
Location of vascular involvement	
Main PA and Lobar PAs	5 (13%)
Bilateral segmental and subsegmental PAs	14 (35%)
Unilateral lower extremity DVT	18 (46%)
Other	6 (15%)
Immunosuppressive treatment for the first vascular involvement	
Azathioprine	27 (69%)
Methylprednisolone	5 (13%)
Cyclophosphamide	8 (20%)
Other (adalimumab, infliximab, interferon)	3 (0,07%)
Anticoagulant treatments	
Rivaroxaban	27 (69%)
Apixaban	9 (23%)
Dabigatran	2 (0,05%)
Edoxaban	1 (0,02%)

BD: Behcet's Disease, PA: pulmonary artery, DVT: deep vein thrombosis

CO6.1

Replication of the 23-Valent Polysaccharide Pneumococcal Vaccine-Induced Skin Pathergy Test in an Independent Cohort of Patients with Behçet Disease

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Introduction: The skin pathergy test (SPT) is an important tool in the diagnosis of Behçet Disease(BD), but its decreasing sensitivity over years has limited its use, especially after the introduction of single use disposable sharp needles. Previously, we showed improved sensitivity with preserved specificity with PS-23 antigen induced SPT, especially in active BD(aBD) as 80.3% and 100%, respectively. This study aimed to improve and replicate our previous findings with increasing the number of PS-23 induced pricks.

Method: BD patients, patients with other inflammatory diseases, and recurrent aphtous stomatitis were comprised the study group. Standard SPT was done by pricking of forearm with 20G hypodermic needles. PS23-SPT was applied by dropping a total volume of 10 ul 23-valent polysaccharide pneumococcal vaccine before 20G needle prick. Two pricks of each methods were applied in each arm. Induration (>2 mm) with erythema at 48h was accepted as positive.

Discussion: Stimulation of forearm skin by PS-23 and 20G needle prick showed 79.6% sensitivity and 100% specificity in all BD, and 91.3% sensitivity and 100% specificity in aBD patients compared to the sensitivity of 8.6% in all and 10% in aBD using a single 20G prick. For PS23-SPT of BD, both erythema and induration showed significant correlation both between first and second prick sites of each arms and between right and left forearms ($p<0.0001$ and $r=0.7$-0.8 for groups). In aBD group, 28.6% of PS- SPT negative and 6.3% of PS-SPT positive patients were under at least one immunosuppressives (IS). Fifty out of 73 PS-SPT positive aBD patients (68.5%) developed at least one pustule.

Conclusion: Duplication of prick numbers contributed the sensitivity of SPT. The association between positive result and disease activity and not using IS was confirmed. This replication study confirmed and improved our previous findings suggesting SPT induced by PS-23 antigens as a promising tool for the diagnosis of BD compared to the standard SPT.

CO6.2

Skin Microbiome Shows Differences Between Pathergy Positive and Negative Patients with Behçet's Syndrome

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Introduction: Gut, oral and genital mucosa microbiome studies in Behçet's syndrome (BS) have shown heterogeneous results including reduced bacterial diversity and decrease in butyrate-producing bacteria. A role of microorganisms and/or microbial products in pathergy positivity was proposed based on decreased positivity after surgical cleaning and increased positivity when patient's saliva and pneumococcal vaccine are applied to the pathergy site. We aimed to determine the skin microbiome of pathergy (+) and (-) BS pts.

Method: We compared the bacterial and fungal skin microbiome of 30 pathergy (+) and 30 pathergy (-) pts who were not treated with immunosuppressives. Skin samples were obtained by swabbing before the pathergy test and stored at -80C. The first 30 pathergy (+) pts and 30 pathergy (-) pts matched to the pathergy (+) ones for demographic features were included. Samples were studied using 16S and 5.8S rRNA gene sequencing.

Discussion: Bacterial diversity within each group (alpha diversity) was similarly high in both groups. Beta diversity analysis revealed that the bacteria in the two groups were different from each other in terms of genus and species. The pathergy (-) group was rich in Betaproteobacteria, Burkholderiales, and Ralstonia picketti, while the pathergy (+) group showed enrichment in Nevskia soli, Staph. haemolyticus, and Sphingomonas sanxanigenens. Fungal alpha diversity was higher in the pathergy (+) group. Beta diversity showed similar fungi in both groups. Pseudopezicula tracheiphila and Asp. heterocaryoticus were the most abundant fungi in the pathergy (+) group, while Asp. subversicolor prevailed in the pathergy (-) group.

Conclusion: The skin microbiota was different among pathergy (+) and (-) BS pts. Bacterial diversity was high and bacterial species were different among the two groups. Fungal diversity was higher in the pathergy (+) group and there were similar fungi in terms of genus in both groups. The role of these differences in pathergy positivity needs to be further studied.

CO6.3

Axial Spondyloarthritis in Patients with Gastrointestinal Involvement of Behçet Syndrome

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Introduction: Controlled studies have shown that radiographic sacroiliitis was not increased in Behçet syndrome (BS). However, gastrointestinal involvement of Behçet syndrome (GIBS) shares common features with inflammatory bowel disease which, in turn, can be associated with spondyloarthritis (SpA). We wanted to see whether GIBS patients have an increased frequency of radiographic sacroiliitis or non-radiographic axial spondyloarthropathy (nr-AxSpA) compared to BS patients with only mucocutaneous and/or joint involvement.

Method: 71 GIBS and 76 consecutive BS patients without major organ involvement were screened for axial spondyloarthritis (axSpA) using the Assessment of Spondyloarthritis International Society (ASAS) criteria. First they were questioned for chronic back pain, defined by ASAS as the presence of chronic back pain for more than 3 months and an age at onset of <45 years. Patients with chronic back pain were questioned for other spondyloarthritis features and tested for HLA-B27 status, CRP levels and X-ray and magnetic resonance imaging of the sacroiliac joints. All radiologic images were evaluated independently and blind by two radiologists.

Discussion: Chronic back pain was reported by 30 (42%) GIBS patients and 25 (33%) BS patients with only mucocutaneous and/or joint involvement (p=0.24). Five (7%) GIBS patients and 4 (5%) controls met ASAS criteria for axSpA (p=0.74). Only 1 GIBS patient had radiographic axSpA whereas 4 GIBS patients and 4 patients among the controls had nr-AxSpA. HLA B27 was positive in 3 (4%) of the GIBS patients and in 5 (7%) of the controls (p=0.72). There were no significant differences between the groups regarding other SpA features of the ASAS criteria.

Conclusion: The frequency of axSpA in GIBS patients was not higher than that in BS patients who have only mucocutaneous and/or joint involvement. This finding further suggests that, despite certain clinical similarities between GIBS and Crohn's disease, different disease mechanisms may be involved.

CO6.4

Saliva proteome analysis in Behçet's Disease: ELISA validation

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Introduction: Oral ulceration can appear as a stand-alone or be part of a systemic disease as in Behçet's Disease (BD), Inflammatory bowel disease (IBD) or mucous membrane pemphigoid (MMP). To determine specific proteins that are uniquely correlated as well as specific to each disease, the saliva proteome of this patients (10 on each) had been previously studied. When comparing diseased samples to healthy controls, 16 proteins were increased and 27 were decreased. 6, 8 and 10 were uniquely increased and 12, 16 and 13 decreased in the BD, IBD and MMP cohorts respectively. Galectin 7 and Annexin A1 were over expressed in all 3 groups. In BD, over expression of SLPI was seen. Interleukin-1 and Immunoglobulins as well as human Desmoyokin were increased MMP and IBD respectively. This project aims to validate the results by using a secondary and complementary technique – enzyme linked immunosorbent assay (ELISA) with the objective to establish the use of ELISA technique in saliva samples as well as measure the quantity of candidate protein on each sample.

Method: ELISAs were validated with healthy volunteers' saliva prior use of the samples. ELISAS were then performed according to the manufacturer's instructions and the predetermined dilutions. GraphPad Prism 10.1.0 was used for analysis. Mean rank differences in each group were compared using the Kruskal–Wallis, non-parametric statistical test.

Discussion: SLPI, Galectin 7 and Annexin 1 was decreased in BD but increased in the IBD group. Desmoyokin, Interleukin and Immunoglobulin were increased in the latter. The role of the identified proteins in BD/ IBD/MMP patients as well as the diagnostic challenges between the BD/IBD groups will be discussed.

Conclusion: As currently there are no specific diagnostic tests for BD/IBD, identifying such biomarkers could aid in improving early clinical diagnosis and enhance more targeted successful treatments. This not only could make patient's experience more positive but the use of clinicians' time more effectively.

CO6.5

Patients with Oral Ulcer Display High Levels of Salivary Acidic Glycoproteins in Behçet's Syndrome

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Introduction: The oral mucosa is subjected to antigenic stimulations by oral microorganisms, which play crucial parts in inducing immune responses and exhibiting clinical symptoms in Behçet's Syndrome (BS). The aim of the study was to assess whether salivary levels of sulfated and sialylated glycoproteins called acidic-type glycoproteins were associated with oral ulcer (OU) activity in BS patients.

Method: Patients with BS (F/M:15/18), active patients with oral lichen planus (OLP, F/M:5/5) as a diseased control group and healthy controls (HC, F/M: 18/14) were included in this cross-sectional study. Stimulated saliva samples were collected and their flow rates (ml/min) were calculated. The Alcian Blue method was used for the quantitative measurement of sulfated and sialylated glycoproteins in the stimulated saliva samples (µg/ml).

Discussion: The salivary levels of sulfated and sialylated glycoproteins in BS patients were significantly higher (51,08±26,12 µg/min) than those in the OLP group (1.63±0.75 µg/min) and HC (12,84±23.62 µg/min) ($p<0.05$). Yet, no significant relationships were observed among the levels of glycoproteins, age, gender and medication use in BS patients ($p>0.05$). In BS, the levels of sulfated and sialylated glycoproteins were categorised as the low-level group (25th percentile:29 µg/min, n=8) vs. the high- level group (50th percentile; 47 µg/min and 75thpercentile: 72 µg/min, n=25). Although 85% (n=17) of patients with active OU (n=20) were in the high-level group, only 3 patients (15%) with active OU were in the low-level group. In addition, increases in the number (2.43±1.99) and healing time of OU (6.43±5.5) were observed in the high-level group compared to another group (0.75±1.16; 2.0±2.97) ($p<.0.05$).
Conclusion: Elevated salivary levels of sulfated and sialylated glycoproteins in patients with active OU could be explained by their roles in the innate immune system that maintain oral homeostasis against the microbial ecosystem in the presence of OU in BS.

CO7.1

Prospective follow-up of patients with suspected Behçet's Disease: First results of an inception cohort.

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Introduction: Behçet's disease (BD) is characterized with diverse clinical manifestations, which can make it difficult to classify patients. Despite the lack of fulfillment of International Study Group (ISG) criteria, some patients with specific manifestations were diagnosed and treated as BD with expert opinion in experienced centers. We aimed to follow prospectively these patients that can be classified as suspected BD and conducted an inception cohort including patients diagnosed only within the last 6 months.

Method: Fifty-seven patients with suspected BD were included in this inception cohort study and followed prospectively. Fulfillment of classification criteria, new major organ involvements and immunosuppressive (IS) usage were assessed during follow-up.

Discussion: Initially, no patient fulfilled the ISG criteria, but 24(42,1%) fulfilled the International Criteria for BD(ICBD). Thirty-two (56,1%) patients were male. The median age was 37,4(31-43) years and median follow-up duration was 38(9-74) months. Initially, 47(82,5%) patients had major organ involvement. During follow-up, new major organ involvement was observed in 8(14.3%) patients and these patients needed step-up treatment with ISs. During follow-up, 5 (8,7%) (2, 2 and 1) patients developed mucocutaneous manifestations and fulfilled the ISG, ICBD and both ISG, ICBD criteria, respectively. Two (3.5%) patients with pulmonary artery aneurysm had deceased due to massive hemoptysis. The mean common femoral vein (CFV) wall thickness was 0.72 mm (SD: 0.16) for the right and 0.71 mm (SD: 0.15) for the left. In 54(94.7%) patients, bilateral CFV wall thickness exceeded the 0.5 mm cut-off.

Conclusion: In our inception cohort with 57 patients, 5(8,7%) developed new mucocutaneous manifestations and fulfilled the ISG or ICBD criteria during 38 months of follow-up. Eight patients (14.3%) developed new major organ involvement. Long term follow-up of our cohort will show the prospective course of BD patients presenting with limited clinical manifestations initially.

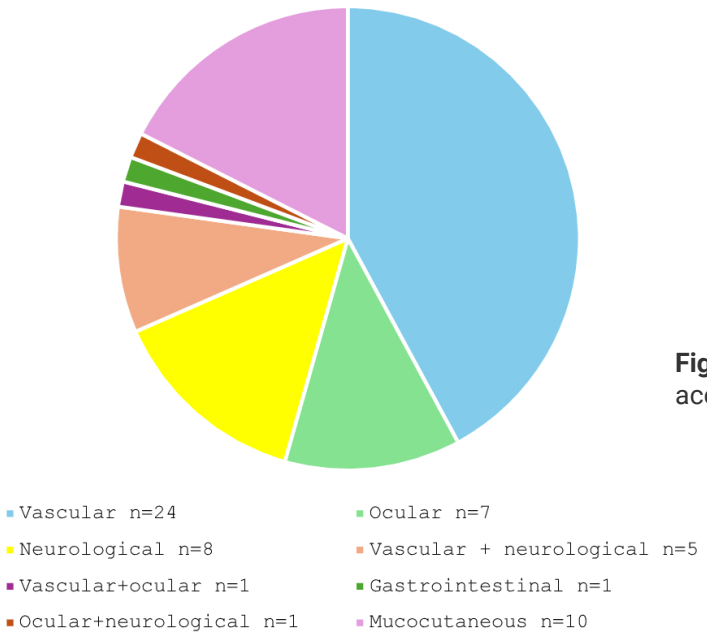

Figure 1. Distribution of suspected Behçet's patients according to organ involvements.

Table 1: Clinical features of the suspected BD patients with a new major organ involvement at follow-up

	Presentation at diagnosis	Age at Diagnosis	Disease duration when new major organ presented (months)	Treatment at diagnosis	New Major Organ İnvolvement	Treatment after new major involvement
Patient 1	OU+Vascular	40	88	Colchicine	Pulmonary Thrombus	AZA
Patient 2	OU+Vascular	43	39	AZA	Femoral artery aneurysm	CYC, MP pulse
Patient 3	Vascular	17	50	AZA	Thrombophlebitis	CsA
Patient 4	OU+Vascular+Neurological	35	78	ADA	Pulmonary Thrombus	MMF*
Patient 5	Ocular	41	44	AZA	Ocular	ADA
Patient 6	Ocular+Neurological	32	148	ADA	Ocular	TOCI
Patient 7	OU+Vascular	34	76	AZA	Pulmonary Thrombus	ADA
Patient 8	OU+Vascular	45	31	Colchicine	Pulmonary Thrombus	AZA

OU: Oral ulcer, AZA: azathioprine, ADA: adalimumab, MMF, mycophenolate mofetil, TOCI: tocilizumab, CYC: cyclophosphamide, CsA: cyclosporin-A

*When receiving adalimumab, the patient had diagnosed hepatocellular carcinoma and his treatment had been switched to MMF.

CO7.2

Major organ involvement is lower in young male patients with Behçet Disease during ten years prospective follow-up compared to retrospective cohorts

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Introduction: Major organ involvement is one of the main causes of mortality and morbidity in Behçet's Disease (BD) especially in young males. However, the prognosis and predictors of major organ involvement are insufficiently studied. We aimed to prospectively follow up with young, male BD patients with only mucocutaneous symptoms who have the highest risk for new major organ involvement.

Method: Thirty-six male patients with BD meeting ISG criteria were included. Patients with BD were assessed prospectively at 3-6 months intervals and in any urgent visits. New major organ involvements and reasons for immunosuppressive (IS) needs were assessed.

Discussion: At baseline, the mean age of the patients and the mean duration of disease were 29.6 (SD: 5.1) and 3.8 (SD: 3.3) years, respectively. At the last follow-up visit, the mean follow-up duration was 132.4 (SD: 14.2) months. The mean age of the patients was 40.7 (SD: 5) and the mean duration of the disease was 14.7 (SD: 3) years. 2 patients had lost follow-up. All patients were using only colchicine at baseline. At the end of the follow-up, 13 (36.1%) patients needed IS therapy due to major organ involvement in 5 (13.9%), refractory mucocutaneous involvement in 7 (19.4%), and articular involvement in 1 (2.8%) patient (**Figure 1**).

Conclusion: Our study which is the first prospective long-term follow-up study in BD, demonstrated a lower incidence of major organ involvement in male BD patients during prospective follow-up when compared with retrospective series. We also found that refractory mucocutaneous involvement and arthritis were more frequent reasons for IS need in young male BD patients.

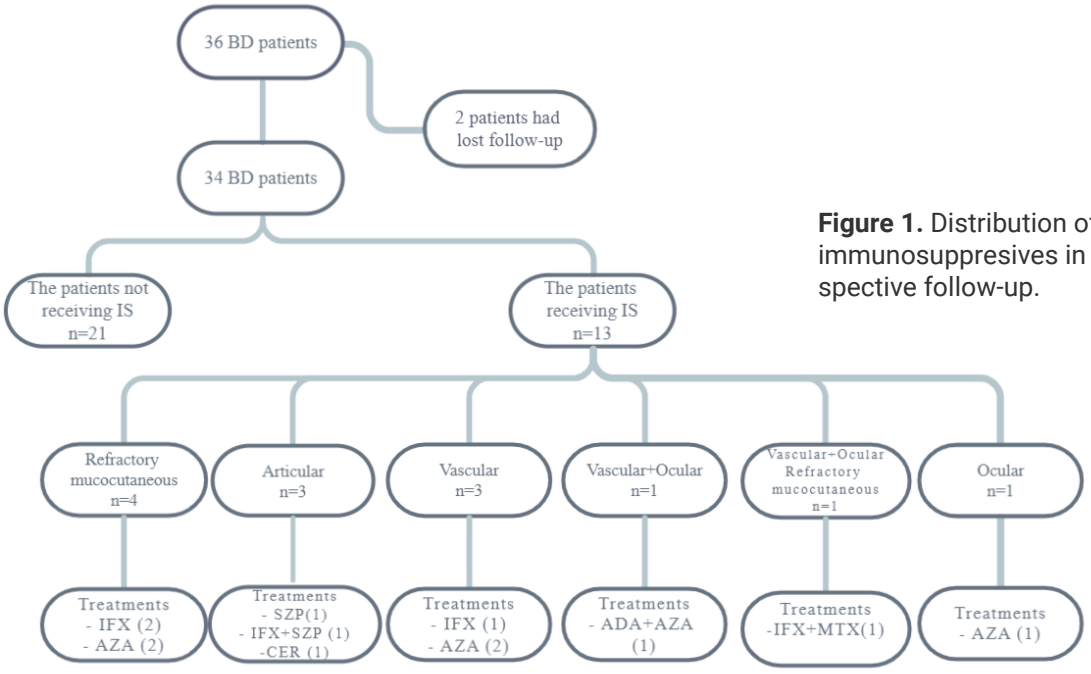

Figure 1. Distribution of manifestations needed immunosuppresives in BD patients during prospective follow-up.

CO7.3

Identifying Clinical Clusters in Behcet Syndrome: A Latent Class Analysis of a Large Retrospective Cohort

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Introduction: Behçet's syndrome (BS) is a complex, multisystemic vasculitis characterized by extreme heterogeneity, with varying disease courses among patients in terms of types and severity of manifestations. Recent cluster analysis studies in BS have yielded distinct results compared to previous phenotype studies based on factor analysis. The primary objective of the present study is to identify distinct clusters of clinical phenotypes in BS to enable more accurate classification and personalized care.

Method: We retrospectively conducted latent class analysis (LCA) to calculate the probabilities of BS patients belonging to different clusters. The optimal number of classes was selected based on multiple statistical criteria and clinical relevance.

Discussion: A total of 531 patients (411 male, 120 female), aged 30 ± 7 years (range 18-47), were enrolled. We identified 4 clinical clusters (C1-C4). In C1, all patients had mucocutaneous and articular involvement (100%), with predominant ocular lesions (70%), associated with neurological manifestations (10%). All patients in C2 had bipolar ulcers, arthralgias, and vascular involvement (100%), with frequent aneurysms (30%), cardiac (15%) and digestive involvements (10%). Patients in C3 consistently presented ocular lesions (100%), mainly associated with bipolar ulcers (50%) and vascular lesions (35%), especially deep venous thrombosis (70%). In C4, half of the patients exhibited vascular lesions and bipolar ulcers (50%), with common neurological involvement (15%).

Conclusion: This is the first clustering analysis using LCA in BS. We identified 4 clusters expressing different phenotypes with various outcomes. Our analysis may assist clinicians in accurately identifying disease subtypes to provide personalized treatment.

CO7.4

A Systematic Review of Outcome Measures Used to Assess the Core Domains for Clinical Trials in Behçet Syndrome

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Introduction: The Outcome Measures in Rheumatology Clinical Trials (OMERACT) Behçet Syndrome (BS) Working Group, a large, multidisciplinary group of experts in BS and patients with BS, proposed a core set comprising five domains mandatory for all trials in BS: disease activity, new organ involvement, quality of life (QoL), adverse events (AE), and death, as well as additional subdomains mandatory for specific organ–systems. We aimed to conduct a systematic literature review (SLR) to identify the outcome measures that match each of these domains and subdomains, for developing a Core Set of outcome measures for BS.

Method: We searched PubMed and ClinicalTrials and included randomized controlled trials, uncontrolled interventional, observational, longitudinal prospective or retrospective cohort, and case-control studies reporting any outcome measure and/or instrument (Fig).

Discussion: We reviewed the titles and abstracts of 9895 references, followed by the full texts of 752. Among these, 432 studies met our inclusion criteria. We identified 8 different instruments that were used to assess disease activity and 1 disease specific (BDQoL) and 16 generic instruments for QoL. There were few BS specific instruments for organ involvement such as Disease Activity Index for Intestinal Behçet's Disease and few instruments developed for other diseases with similar organ involvement. Several studies used their own definition for remission, response or relapse for organ subdomains.

Conclusion: This SLR revealed the diversity and variability of outcome measurement instruments that were used in BS studies, and the lack of disease-specific tools for most types of organ-specific involvement. Evaluation of the psychometric properties of the currently available instruments for selecting the best fitting instrument for each domain and subdomain and development of new instruments when necessary will lead to the development of a Core Set of outcome measures for BS.

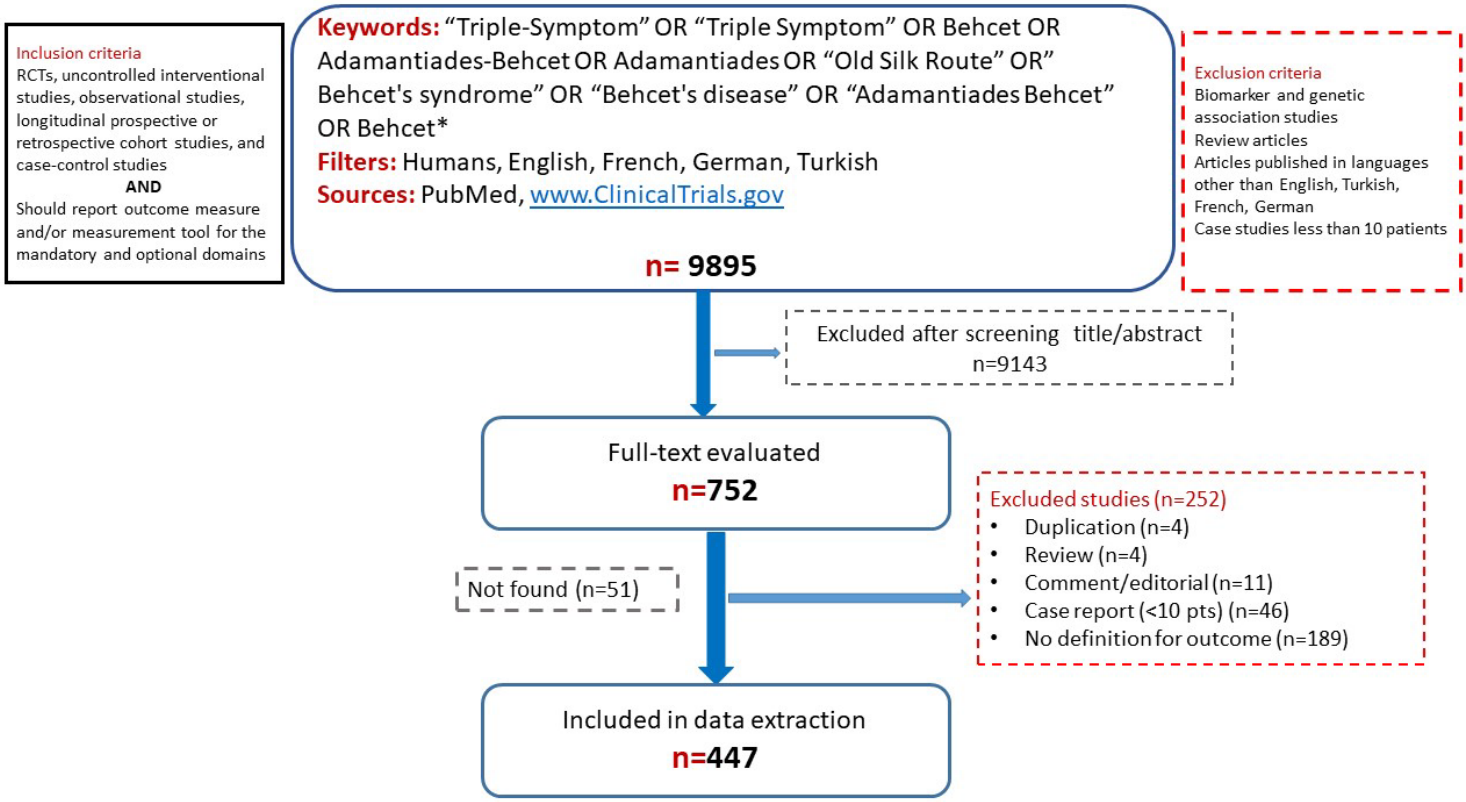

Figure 1. Flow-chart of study selection process

CO7.5

Comparison of the 2015 Pediatric Diagnostic Criteria for Behcet's Disease with the 2014 and 1990 International Study Group Diagnostic Criteria

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Introduction: Behçet's disease is a multisystem vasculitis whose pathogenesis remains unclear. Although usually described in young adults, it may begin in childhood. The diagnosis is clinical, based on international criteria. The limitations of early diagnosis are related to the progressive onset of symptoms and the variety of differential diagnoses at this age in the absence of a pathognomonic diagnostic test.

Method: To report the epidemiological features of our series and to compare the 2015 pediatric criteria with the 2014 and 1990 international criteria.

Discussion: There were 38 cases of Behçet's disease. The mean age was 10.44 years (5.5-14). The F/M sex ratio was 1.1. Eight patients (21%) were from a consanguineous marriage and 13 (34.2%) had a family history of Behçet's disease. Mucocutaneous manifestations were represented by recurrent oral aphthosis 89.4%, genital aphthosis 31.5%, pseudofolliculitis 48.4%, erythema nodosum 13.1% and acneic lesions 3.2%. The pathergy test was positive in one case. Ocular involvement was reported in 34.2% of cases, and joint involvement in 36.8%. Thromboembolic complications were observed in 18.4% of cases, and neurological complications in one case. HLA B51 antigen was present in half the patients. In our series, 44.7% met pediatric criteria, while 68.4% met 2014 international criteria and 36.8% met 1990 international criteria. The 2015 pediatric criteria had a sensitivity and specificity of 63.2% and 100% (P=0.002) respectively, compared with the 2014 international criteria taken as the gold standard, and the 1990 international criteria with a sensitivity and specificity of 52.6% and 91.7% (P=0.034) respectively.

Conclusion: Our study highlights the absence of male predominance, a high rate of consanguinity and familial Behcet. The sensitivity and specificity of the 1990 pediatric and international criteria appear better than those of 2014, with a more significant trend for the 2015 pediatric criteria (P=0.002).

CO8.1

Medication Adherence and Compliance in Patients with Behçet's Syndrome

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Introduction: Adherence and compliance with medication in Behçet's Syndrome (BS) is important in maintaining, control and preventing activation of disease which could be organ- threatening. The aim of the study was to assess the adherence and compliance of patients with their Behçet's medication.

Method: This study took place at the London Behçet's Centre where 107 BS patients were provided with a questionnaire. This enquired about medication taken, whether they miss any doses and if so, the reasons for not taking their medication as directed.

Discussion: Colchicine was the most commonly taken medication (59%), followed by azathioprine (50%) and Triorasol mouthwash (24%). Side effects were reported by 26% of patients, including gastrointestinal disturbance, hair loss, insomnia, tiredness, dry skin, weight loss and breathlessness. Additionally, 63% of patients took medications for other conditions. In terms of adherence, 84% of patients followed their doctors' instructions for medication, and 58% took their medication at the same time every day. However, 53% of patients reported missing doses, with forgetfulness, late general practitioner appointments, and busy schedules being common reasons. The majority of patients (93%) took their medications while travelling.

Conclusion: This study revealed that adhering to long term medication can be challenging for patients due to several factors such as paying for prescriptions, working in full time jobs, forgetfulness, side effects, social support and limited awareness about risks and side effects. This investigation is ongoing with future plans to better understand factors affecting the compliance and adherence rates in long term medication use in BS.

CO8.2

Characterization of a Multicenter Cohort of Patients with Behçet's Syndrome Treated with Methotrexate: Efficacy and Safety

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Introduction: The clinical heterogeneity of Behçet syndrome (BS) complicates its treatment. Methotrexate (MTX) is a drug widely used in rheumatology. Despite its potential role in BS, there is no published evidence in clinical practice. Our aim was to describe the efficacy and safety of MTX in the different clinical phenotypes of BS in clinical practice in our country.

Method: A national, multicenter, observational, retrospective study of BS patients treated with MTX. Clinical and analytical data, response, and adverse effects were collected.

Discussion: 97 patients were included: 94.8% Caucasian, 52.6% women, mean age 34.9±13 years at diagnosis and 41.9±13 years at MTX start. MTX was administered due to articular phenotype (43 patients), mucocutaneous (35), and ophthalmologic involvement (17). Previously, 37.1% had received conventional DMARDs, 5.2% apremilast, 19.6% biologic DMARDs, and 5.2% cyclophosphamide. Mean maximum MTX dose was 13.81±4.2 mg/week. MTX was administered subcutaneously in 72% and in combination therapy in 40.5%. The median duration of MTX was 2.2 (0.6-5.2) years, with equal survival rates in monotherapy and combination therapy and no difference in efficacy (32.1% vs 39%, p=0.48). MTX was discontinued in 62 patients due to inefficacy (33.8%), clinical remission (14.5%), mild adverse effects (24.7%), intolerance (16.1%). MTX monotherapy efficacy was analyzed in 62 patients with different phenotypes (Table 1). In the ophthalmologic phenotype, the time from diagnosis to drug initiation was significantly shorter (p=0.02). The articular subgroup had a better response but a higher MTX discontinuation rate due to intolerance. No link was found between GI involvement and discontinuation. No differences in efficacy and safety were observed between the ophthalmologic and mucocutaneous subgroups.

Conclusion: MTX is widely used for BS, both as monotherapy and combination therapy, with an acceptable safety profile. The articular phenotype is the main indication and most effective for MTX monotherapy.

	Mucocutaneous n=23	Articular n=29	Ophthalmologic n=10
Female sex[1]	14 (60.9)	17 (58.6)	5 (50)
Age[2]	38.9 (27.8-47.3)	42.6 (37.5-49.8)	35.6 (32-48.6)
Disease duration[2]	2 (0.3-28)	3.6 (0.4-16)	0.73 (0-2.7)
Clinical features[1]			
Oral ulcers	22 (95.7)	29 (100)	9 (90)
Genital ulcers	17 (73.9)	21 (72.4)	6 (60)
Erythema nodosum	6 (26.1)	5 (17.2)	5 (50)
Pseudofolliculitis	17 (73.9)	18 (62.1)	5 (50)
Arthritis	12 (52.2)	26 (89.7)	6 (60)
Uveitis	7 (30.4)	7 (24.1)	8 (80)
Retinal vasculitis	0	0	2 (20)
Vascular involvement	2 (8.7)	6 (20.7)	1 (10)
GI involvement	1 (4.3)	6 (20.7)	0
Cardiac involvement	0	0	0
Neurobehçet	5 (21.7)	2 (6.9)	2 (20)
HLAB51	6/12 (50)	10/21 (47.6)	3/7 (42.9)
Positive pathergy	4/14 (28.6)	4/14 (28.6)	0/1
Previous treatment			
Colchicine	20 (87)	26 (89.7)	7 (70)
Corticosteroid	19 (82.6)	25 (86.2)	7 (70)
Hydroxychloroquine	0	4 (13.8)	1 (10)
Azathioprine	4 (17.4)	2 (6.9)	1 (10)
Cyclosporine	2 (8.7)	0	1 (10)
Apremilast	1 (4.3)	3 (10.3)	0
Anti-TNF	3 (13)	4 (13.8)	0
IL-1 inhibitor	0	0	0
IL-6 inhibitor	0	1 (3.4)	0
IL-12/23 inhibitor	0	1 (3.4)	0
Cyclophosphamide	0	2 (6.9)	1 (10)

[1]Number (%), [2]Median (IQR), years,

Table 1. Clinical characteristics of 62 BS patients treated with MTX in monotherapy.

	Mucocutaneous n=23	p-value	Articular n=29	p-value	Ophthalmologic n=10
Maximum dose[2]	15 (10-15)	ns	12.5 (10-15)	ns	16.3 (10-16.3)
Efficacy[3]	11/17 (64.7)	0.04	23/25 (92)	0.03	5/9 (55.6)
Discontinuation[1]					
- Inefficacy	7 (30.4)	0.04	2 (6.9)	ns	5 (50)
- Intolerance	0	0.002	7 (24.1)	0.031	0
- Adverse effect	6 (26.1)	ns	3 (10.3)	ns	2 (20)
- Improvement	6 (26.1)	ns	3 (10.3)	ns	2 (20)
- Others	2 (8.7)	ns	0	ns	1 (10)

[1]Number (%), [2]Median (IQR), years, [3]Efficacy is evaluated in patients who did not discontinue MTX within the first 3 months.

Table 2. Characteristics of MTX treatment: Efficacy and safety.

CO8.3

Retrospective Study of the Efficacy of Apremilast for Uveitis in Behçet's Disease Ocular Lesions

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Introduction: Apremilast (APR) is a PDE4 inhibitor approved for treating refractory oral ulcers in Behçet's disease (BD). Although the efficacy of APR for lesions other than oral ulcers had been expected, there is limited evidence for its efficacy. We studied the efficacy and safety of apremilast for refractory oral aphthous ulcers in BD on ocular lesions.

Method: We retrospectively reviewed the medical records of 27 cases in which APR was introduced for refractory oral ulcers from September 2019 to December 2022.

Discussion: The mean age of the 27 patients was 45.4 ± 11.7 years, and the mean observation period from the introduction of APR was 25.6 ± 13.2 months. Patients were divided into three groups: those without a history of uveitis (group 1), those with long-term remission (group 2), and those with active inflammation (group 3). Sixteen patients in group 1 had no ocular inflammation before APR administration. Eleven patients had a history of ocular inflammation, eight in group 2 and three in group 3. All three patients in group 3 had active ocular inflammation despite receiving infliximab. All sixteen patients in group 1 did not develop new ocular inflammation during the observation period. In group 2, 7 of the eight patients remained in remission, while one had a flare-up of anterior ocular inflammation. In group 3, one patient had improved ocular inflammation, and two showed continued active inflammation. There were no adverse ocular events related to APR.

Conclusion: The results suggest that APR may have a certain effect on ocular inflammation in BD. Further studies of APR are needed to establish its efficacy on ocular inflammation.

CO8.4

De Novo Manifestations During Adalimumab Treatment in Behçet Syndrome

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Introduction: Treatment response may be variable across organ manifestations of Behçet syndrome (BS). We aimed to determine the frequency of de novo manifestations during adalimumab treatment. We conducted a chart review of all BS patients who received adalimumab in our center between 2008 and 2023. Demographic data, reasons for initiating adalimumab, concurrent medications, previous treatments, and outcomes were recorded.

Method: We defined de novo manifestations as new BS manifestations that occurred for the first time during treatment with adalimumab. For patients with vascular involvement, a new vascular event at another vessel was also considered as a de novo manifestation.

Discussion: Among the 335 patients, a de novo manifestation developed in 14 (4%) patients. De novo manifestations were vascular involvement in 5 patients, arthritis in 3, anterior uveitis in 2, nervous system involvement in 2, gastrointestinal involvement in 1, and epididymitis in 1 patient. The primary reasons for adalimumab treatment were vascular involvement in 5 patients, uveitis in 4, arthritis in 3, mucocutaneous involvement in 1, and epididymitis in 1 patient. Upon the development of de novo manifestation, adalimumab was switched to another biologic in 4 patients, dose was intensified in 3, colchicine, conventional immunosuppressives, and/or glucocorticoids were added in 5, and topical eye drops were added in 2 patients, leading to remission of de novo manifestations in all patients.

Conclusion: De novo manifestations were infrequent (4%) among BS patients treated with adalimumab. Majority (71%) were major organ involvement, mainly vascular involvement. None of the patients developed posterior uveitis.

CO8.5

Feasible drug-free remission for more than 5 years following withdrawal of successful long-term anti-TNF treatment in Behcet's disease: a re-appraisal of a single-center longitudinal outcome study

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Co-Authors: Nikos Markomichelakis, George Fragoulis, Petros P. Sfikakis

Introduction: Observational studies suggest that prognosis of vital organ involvement in Behcet's disease (BD) has remarkably improved since 2001, when anti-TNF treatment was first introduced [Lancet 2001]. Herein, we aimed to extend our previous study [Arthritis Rheumatol 2017], testing the hypothesis that long-term remission after withdrawal of successful anti-TNF treatment is feasible in severe BD.

Method: This retrospective study focused on BD patients, refractory to csDMARDs, who received successful anti-TNF treatment and subsequently discontinued. Endpoint was the proportion of patients with sustained remission for at least 5 years after withdrawal.

Discussion: From our previous BD cohort, we excluded patients lost to follow-up, as well as those who discontinued anti-TNF treatment after April 2019. Of 32 patients eligible for analysis (mean age of 47 years, 53% men), who received TNF inhibitors for a median of 2.5 years, 26 (81%) were treated for sight-threatening disease, and 3/32, 2/32 and 1/32 for severe mucocutaneous, central nervous system and gastrointestinal involvement, respectively. After anti-TNF withdrawal, 15/32 patients remained in remission for at least 5 years (range 6.5-20). Of the 17/32 patients who relapsed after a median of 7.5 months following withdrawal, retreatment with anti-TNF was effective in the long-term in 12/17. Of them, 8/12 discontinued for a second time and achieved again the study's endpoint. Overall, 23/32 (72%) remain in remission for a mean of 12 years. Of note, 17/23 are any-drug free or on colchicine only, whereas 6/23 are on azathioprine maintenance.
Conclusion: Remission lasting more than 5 years after withdrawal of successful anti-TNF treatment is feasible in patients with severe BD, suggesting that discontinuation of ant-TNF should be considered. However, since relapses that do not respond to anti- TNF re-treatment may occur, further studies are needed.

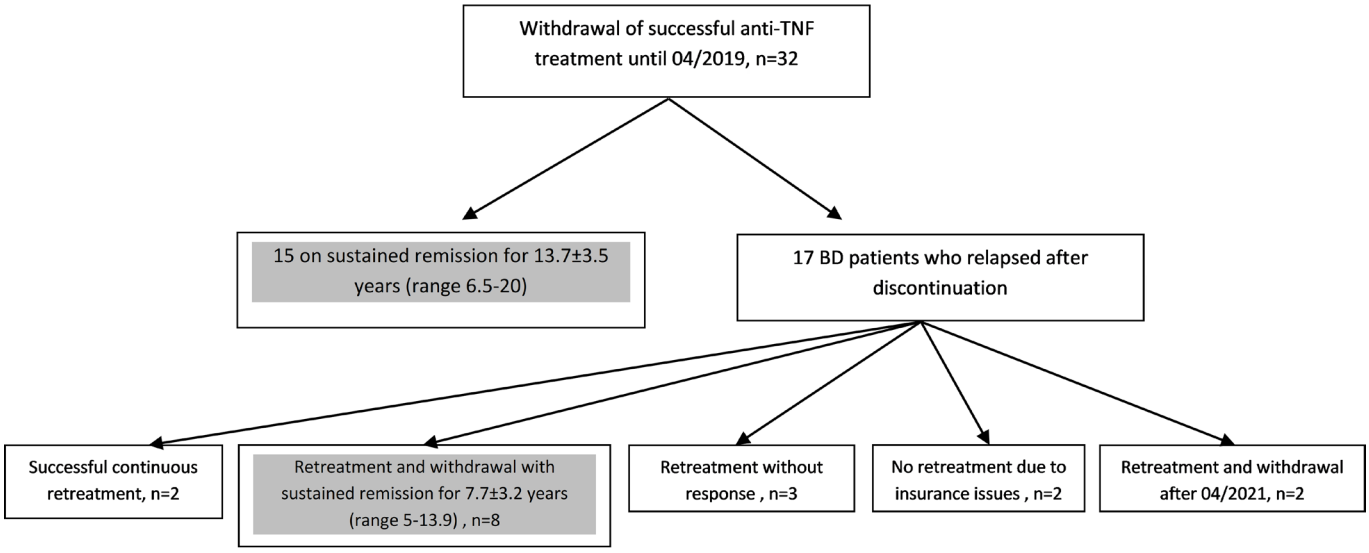

Figure 1. Flow chart of Behcet's disease patients included in the analysis.

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E-POSTERS

Uveitis in Children living with Behcet Disease: A Case Series

Kenza Bouayed, Hôpital Mère-Enfant A. Harouchi, CHU Ibn Rochd, Casablanca, Morocco
Co-Authors: Abdelhakim Youssef Benmoussa

Introduction: Behcet's disease is a multisystem vasculitis characterized by recurrent oral and genital ulcers and ophthalmological involvement. Uveitis and retinal vasculitis represent are one of the complications due to the risk of sight loss. They represent a therapeutic challenge because of their recurrence and severity

Method: 38 BD patients followed in a Pediatric Rheumatology Department,11 presented with uveitis and were retrospectively evaluated. only 6 were evaluable.The diagnosis of BD was established according to PEDBD, ICBD and/or ISG criteria.

Discussion: The mean age was 15 years and 6 months. 5 patients were male and 2 females. The mean age of onset of the first ocular symptoms was 11 years. 3 patients presented a family history. 5 patients reported ocular symptoms: ocular pain in 2 patients, ocular redness in 2 patients, decreased vision in 3 patients and sensitivity to light in one patient. 4 of the six patients were HLA B51 positive. Treatment included corticosteroids for all patients. All received bolus methylprednisolone followed by oral corticosteroids. Methotrexate was prescribed for one patient, azathioprine for all. The other molecules prescribed were colchicine for 2 patients and doxicycline for one patient in initial suspicion of bartonellosis. All patients were candidates for TNF alpha inhibitors. Only one patient received anti-VEGF injections. Average follow-up was approximately 4 years. Our patients relapsed twice on average. Currently, 2 patients have active uveitis,4 are in remission. Complications include cataract in 1 patient. Visual acuity improved to normal levels in 3 patients, and remained low in 3 cases.

Conclusion: In the literature, the mean frequency of uveitis is 45% with a range of 34% to 56%, which is similar to our series with a frequency of 38%. Uveitis in Behcet's disease is known for its recurrence and severity. Regular follow-up is essential to avoid ocular complications. Aggressive therapies with harmful side effects are often required to put the patient into remission.

Thrombosis in Children with Behcet Disease: A Monocentric Study

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Introduction: Behçet's disease is a systemic vasculitis known for its high risk of thrombosis, which is a serious and potentially fatal complication. In Pediatric BD,thrombosis is very rare.

Method: 36 BD patients followed in a Pediatric Rheumatology Department between January 2009 and April 2024 were retrospectively assessed.

Discussion: Thrombosis was found in 6 male patients (16.66 %), mean age was 10,45 years (5.7- 13). It was inaugural in 5 patients. Three patients had cerebral sinus venous thrombosis, 3 patients had deep venous thrombosis, at mesenteric veins (1case), Budd Chiari syndrome complicated with cardiac thrombosis (1 case) and internal jugular vein associated with lower lobar artery aneurysm (1case). The mean erythrocyte sedimentation rate was 30mm/1st h, and the mean C-reactive protein was 37.23 mg/L. All patients were treated with low-molecular-weight heparin and anti-vitamin K anticoagulation combined with aggressive immunosuppression using boluses of MP, relayed by oral corticosteroids combined with Azathioprine. In addition, one patient was treated with Cyclophosphamide, 2 received Adalimumab and the third is undergoing pre-biotherapy screening for Adalimumab. Outcome was favorable in 5 cases despite a case of recurrent thrombosis, we deplore 1 case of death.

Conclusion: Thrombosis is a serious condition that can reveal a vasculitis such as Behçet's disease. Male gender and high inflammatory parameters may increase the risk of thrombosis in BD. All our patients are male and present with an inflammatory syndrome. Etiological diagnosis is essential, because in addition to anticoagulant therapy, etiological treatment is fundamental to stopping the vascular inflammatory process that is the cornerstone of thrombosis genesis. The etiological problem arises primarily in inaugural forms, which is why any thrombosis in children must be investigated for evidence of BD, and if the patient is known, the family must be encouraged to consult as soon as any unusual symptoms appear.

Optic Neuritis in Behçet's Disease

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Introduction: Optic neuritis is a rare and serious manifestation of Behçet's disease, often leading to optic atrophy-induced blindness. We aimed to analyze optic neuritis in Behçet's disease in our cases series.

Method: This is a retrospective study about optic neuritis in Behçet's disease cases hospitalized in internal medicine department at Mohamed VI hospital, Marrakech from2010 to 2023. Data was collected from medical records and hospital register. Optic neuritis diagnosis was confirmed based on fundus examination findings, cerebrospinal MRI, visual evoked potentials, and visual field testing data. The diagnosis of Behcet's disease was confirmed based on ICBD criteria.

Discussion: Eight patients (14 eyes) were included, with a mean age of 25.5 years and a male-to- female ratio of 1.6. The median time from Behçet's diagnosis to ocular involvement was 2.5 years. Symptoms were dominated by visual acuity loss (80%), ocular pain (50%) and visual field impairment (26%). Visual field examination was performed in 4 cases and revealed an enlargement of the blind spot. Visual evoked potential studies were conducted in 2 cases and noted conduction disorders in the optic pathways. Cerebrospinal MRI revealed optic pathway involvement in 3 cases, including bilateral optic nerve atrophy in one patient and right lateral sinus thrombophlebitis in one patient. there were papillitis in 4 eyes, retrobulbar optic neuropathy in 8 eyes, and stasis papillary edema in 2 eyes. Treatment involved methylprednisolone pulses followed by oral corticosteroids and immunosuppressants. 11 eyes showed improvement with complete recovery in 7 eyes, but 3 eyes developed optic atrophy. Optic neuritis is underdiagnosed in Behçet's disease, often manifesting after several years. Treatment typically includes corticosteroids and immunosuppressants, with varied prognosis.

Conclusion: Optic nerve involvement in Behçet's disease warrants prompt recognition and treatment to prevent irreversible visual impairment.

Dysphonia Revealing Behçet's Disease

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Introduction: Behçet's disease (BD) is a rare and complex inflammatory disorder affecting multiple systems, including blood vessels. While oral ulcers are typical, reports of laryngeal ulcers are unusual. Here, we present a case of vocal cord ulceration due to BD.

Method: Case report: A 35-year-old female without significant medical history presented with dysphonia to the ENT department. Laryngoscopy revealed a 1 cm vocal cord ulcer. Biopsy showed nonspecific inflammatory infiltrate. History revealed recurrent oral aphthosis preceding dysphonia. Genital and oral ulcers were noted in physical examination. Infectious and malignant etiologies were ruled out. Immunological evaluation was negative. BD diagnosis was confirmed. Treatment with colchicine and prednisolone was initiated, resulting in clinical improvement.

Discussion: ENT manifestations of BD are rare but can include laryngeal ulcers, leading to acute inflammation and potential airway obstruction. Treatment typically involves colchicine and steroids, with immunosuppressants for refractory cases.

Conclusion: This case underscores the importance of considering ENT manifestations in BD, warranting vigilance for potentially life-threatening complications like airway obstruction.

Cardiac involvement in Behçet disease

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Introduction: Behçet s disease is a systemic disease that mainly affects young adults. Cardiac involvement is rare (prevalence of 1 to 6%). Pericardial involvement is the most common manifestation.. Endomyocardial damage remains exceptional with intracavitary thrombus and endomyocardial fibrosis. Our study aims to determine the cardiac manifestations of Behçets disease as well as the correlation with other specific manifestations and the disease activity.

Method: Behçets disease is a systemic disease that mainly affects young adults. Cardiac involvement is rare (prevalence of 1 to 6%). Pericardial involvement is the most common manifestation.. Endomyocardial damage remains exceptional with intracavitary thrombus and endomyocardial fibrosis. Our study aims to determine the cardiac manifestations of Behçets disease as well as the correlation with other specific manifestations and the disease activity.

Discussion: Among 399 cases of Behçe's Disease, cardiac involvement was observed in 6 cases (1.5%), 2M/4F aged 33.66 ± 16.47 years. Four patients with cardiac involvement presented intracardiac thrombosis (66.6%) involving: the right atrium (3 cases; 50%) and the right ventricle (1 case; 16.6%). Pericarditis and myocarditis were noted in 1 case or 16.6% each. All patients had mucocutaneous involvement. The most common associated disorder was oculo-behçet (2 cases; 33.3%). Treatment was based on corticosteroid therapy and colchicine in the 6 cases; it was associated with cyclophosphamide in 5 cases (83.3%), azathioprine in a single case (16.6%). Among the 4 patients who presented with an intracardiac thrombus, 3 patients (75%) had anticoagulation based on LMWH and VKA. One patient died of cardiogenic shock.

Conclusion: Most common type of cardiac involvement is pericarditis, whereas intracardiac thrombosis is rare. It's a serious complication that is fairly underdiagnosed.

Retinal vasculitis in Behcet's disease: about 45 cases (82 eyes)

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Introduction: Behcet's disease (BD) is a multisystem disorder that affect all organs, including the eye. The most frequent ocular manifestations are uveitis followed by retinal vasculitis. Our objective was to determine the clinical, therapeutic and outcome of retinal vasculitis in BD.

Method: We studied retinal vasculitis in 45 patients with ocular form of BD. All patients fulfilled the International Study Group criteria for BD. Retinal vasculitis was confirmed by fundus examination and retinal angiography

Discussion: Among BD patients (399 cases), ocular involvement was found in 92 patients (23%). Retinal vasculitis was found in 45 cases (48.9% of patients with ocular involvement) and involves both arteries and veins. The disease occur 4 times more often in men than in women Decreased visual acuity was observed in (97.7%), foggy vision in 13 cases, ocular redness in 29 cases (64%), eye pain in 16 cases and headache in 4 cases. Ocular involvement was bilateral in 37 cases (82%). Other manifestations associated with retinal vasculitis consisted of intermediate uveitis in 23 cases (51%), macular damage in 8 cases (17%), papilledema in 6 cases (13%), intravitreal hemorrhage in 1 case (0.2%),and retinal detachment in 2 cases. All patients were on colchicine and steroids. The immunosuppressive drugs (azathioprine and cyclophosphamide) was prescribed in 95% of cases. Biologic therapy (Tocilizumab and TNF inhibitors) was used in 8 cases. Blindness was observed in 8 cases (17%).

Conclusion: Early diagnosis and rapid and adequate management can improve the visual prognosis of Behcet's retinal vasculitis.

Venous thrombosis in Behçet's disease: a series of 56 cases

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Introduction: Behçet's disease is a vasculitis with a venous predilection and variable locations. The aim of our study is to specify the site, clinical characteristics, therapeutic approaches, and outcomes of venous thromboses in Behçet's disease

Method: This is a monocentric, descriptive, and retrospective study conducted within the Internal Medicine Department "B" of the Mohammed V Military Hospital in Rabat, spanning a 20-year period (2003 – 2023), collecting the medical records of patients treated for Behçet's disease. All patients met the international classification criteria for Behçet's disease of 2013.

Discussion: Among 177 patients with Behçet's disease, 56 had experienced at least one venous thrombosis, representing 31.6%. There were 48 men and 8 women (male-to-female ratio = 6). The average age at the time of vascular involvement was 37 years. Venous thrombosis occurred later in comparison to other clinical manifestations of the disease (cutaneous and articular), after an average duration of eight to nine years. It revealed Behçet's disease in 18 patients. Family history of Behçet's disease was found in 4 cases. Venous thrombosis was predominantly deep vein thrombosis. It was associated with arterial involvement in 16 cases. On imaging, thrombosis affected the iliofemoropopliteal axis in 51 cases, cerebral veins in 6 cases, inferior vena cava in 3 cases, superior vena cava in 4 cases, suprahepatic veins in 3 cases, internal jugular vein in one case, and distal leg veins in 5 cases. Venous thrombosis was complicated by pulmonary embolism in 5 cases. Anticoagulation with vitamin K antagonists (VKAs) was prescribed for all patients, along with colchicine in all cases, and corticosteroids in 51 cases. Anticoagulation was discontinued in 18 cases.

Conclusion: The venous involvement in our study is comparable to other series in the literature. The severity of this involvement lies in the extent, localization, and the risk of recurrence and pulmonary embolism.

Mucocutaneous Manifestations of Behcet's Disease

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Introduction: Mucocutaneous manifestations like oral and genital ulcers, and cutaneous lesions (papulopustular lesions, erythema nodosum-like lesions) are the most common manifestations and often the first signs to appear. Our study aimed to determine the prevalence of mucocutaneous manifestations and evaluate the mean duration between oral ulcers and the fulfilment of diagnosis criteria clinical

Method: This retrospective study analyzed patient data from January 2010 to December 2023, using the International Criteria for Behçet's Disease for inclusion (ICBD).

Discussion: Our study included 399 patients (210 men, 189 women) aged 16 to 60 years (average 32 years). Mucocutaneous symptoms appeared in 87.9% of cases, with 100% presenting oral aphthosis, 75.1% genital aphthosis, 55.6% papulopustular lesions, and 27.5% erythema nodosum. Rare cases showed Raynaud syndrome, skin ulceration, and vasculitic purpura. Systemic manifestations included vascular (24%), ocular (23%), joint (21%), neurological (14.7%), cardiac (1.75%), and digestive (1.5%) involvement. The mean duration between oral ulcers and the fulfilment of diagnostic criteria was calculated to be 4. +/- 6 years.

Conclusion: Mucocutaneous manifestations are the hallmarks of the disease, oral ulcers precede other manifestations by several years

Gender differences in Behcet's Disease: a single experience of 399 cases

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Introduction: Behcet's disease is a multisystem disorder that can affect all organs, including the skin, mucous membranes, eye, joints, brain and blood vessels.. This disease is very common in the Mediterranean basin and mainly affects young men.The objective of our studywas to compare the clinical profiles of the disease in male and female patients.

Method: This is 13 years (January 2010 to December 2023) retrospective study focusing on patients with Behcet's disease compared according to sex

Discussion: There were 399 cases, 210 men (52%) and 193 women (48%). The mean age was 37 ± 25 years for men and 42 ± 22 years for women. Skin involvement was observed in 100% of our patients. Aphthosis was found in 100% of men and women, and it was bipolar in 86% versus 70% of cases respectively. Papulopostular was present in 46% of men and 30% of women. Joint involvement was noted in 46 men with a predominance for large joints and in 81 women. 55 men had vascular Behçet, compared to 36 women (26% versus 18%). Deep vein thrombosis was the main vascular involvement, and was noted in 73 patients. Arterial involvement was present in 18 patients, 15 of whom were men. (P<0,000) Ocular involvement was noted in 88 men and 60 women. Retinal vasculitis was noted in 33 men and 9 women. (P<0,000). Neurological damage was more frequent in men without significant difference (55.9% versus 44%). The main manifestation of neuro-Behçet was headaches with a female predominance. Treatment was based on colchicine in all patients. High-dose corticosteroid therapy; immunosuppressants and biologics were indicated for more severe damage

Conclusion: Our study showed that vascular and ocular involvement were significantly higher in men, whereas joint involvement and headache were more frequent in women

Superior Vena Cava Syndrome and Psychiatric Symptoms Unveil Severe Angio- Behçet with a Giant Thrombosed Coronary Artery Aneurysm

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Introduction: Behçet's syndrome is a multisystemic vasculitis of unknown aetiology. Coronary involvement can manifest as aneurysm formation or occlusion/stenosis and occurs in less than 0.5% of cases. We report a rare case of psychiatric disorders secondary to cerebral venous thrombosis, veina cava syndrome, and giant coronary artery aneurysm as the initial manifestations of Behcet disease.

Method: 32-year-old man with no cardiovascular risk factors, presenting with symptoms of dyspnea, fluctuating behaviors, dizziness, and recurrent mild chest pain. He experienced behavioral disturbances treated as psychiatric disorders for the past 3 months. He reported recurrent oral ulcers and genital ulcers. Clinical evaluation revealed a patient with bradypsychia and behavioral disturbances, superior vena cava syndrome grade II, pseudofolliculitis, and genital aphthae. The electrocardiogram was normal. Laboratory tests indicated an inflammatory syndrome with a high CRP level of 110 mg/l. Troponin levels were negative. Thoracic CT revealed thrombosis of the superior vena cava and a cardiac mass consistent with a partially thrombosed aneurysm of the left anterior interventricular artery measuring 32 × 33 mm, with a slight pericardial effusion. Additionally, a cranial CT showed cerebral venous thrombosis. Echocardiography confirmed the presence of a 32 mm × 32 mm cardiac mass compressing the left atrium, with normal cardiac function.

Discussion: To date, there are no established treatment protocols for this complication. Immunosuppressive treatment should include corticosteroids and cyclophosphamide in combination with vitamin K antagonists. There is still controversy regarding the optimal timing for interventional procedures and whether a covered stent or coronary artery bypass grafting is the best choice.

Conclusion: The mortality rate of cardiovascular manifestations in Behçet's syndrome is 20%. There are still considerable challenges in managing coronary artery aneurysm in BD.

Arterial stenosis in Behcet disease

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Introduction: Behcet disease (BD) is a chronic systemic inflammatory disease characterized by oral aphthae, genital ulcers, skin and eye lesions, and repeated bouts of acute inflammation. Vascular involvement, including arterial stenosis, is also documented in BD.

Method: This retrospective study aimed to investigate the clinical characteristics of Behcet disease (BD) patients with arterial stenosis. BD patients who fulfilled the 2014 ICBD criteria and presented with arterial stenosis were identified from January 2011 to December 2023. Arterial lesions were detected using ultrasonography and/or computed tomography angiography (CTA).

Discussion: Among the 96 Behcet disease (BD) patients diagnosed with vascular involvement, three males were identified with arterial stenosis upon imaging. Their mean age atBD diagnosis was 37 ± 1.6 years, with arterial involvement being the initial manifestation in 66% of cases. The predominant locations of arterial lesions were the lower extremity arteries (n=2), with one patient exhibiting subclavian artery stenosis. Notably, two of these patients presented with both arterial and venous involvement, while one displayed multiple arterial lesions across different sites, including pulmonary aneurysm, thrombosis, and stenosis/occlusion in lower extremity arteries. Treatment involved steroids and immunosuppressive regimens, with two cases administered Cyclophosphamides and one treated with Methotrexate, resulting in successful outcomes for all three patients.

Conclusion: Arterial stenosis remains a rare feature of behcet disease, late diagnosis can lead to life threatening complications, effective screening protocols and early aggressive therapy are essential for favorable outcomes.

Parkinsonian syndrome as a neurological manifestation of Behçet's disease: A case report

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Introduction: Central neurological involvement in Behçet's disease (BD) is estimated at between 5 and 7%. Extrapyramidal syndromes are rarely reported in neuro-Behçet (NB). A typical parkinsonian syndrome has been reported in NB.

Method: We report a patient fulfilling the criteria of the International Study Group for Behcet's disease.

Discussion: He had recurrent oral aphthosis, genital aphthosis and pseudofolliculitis, evolving for 1 year, on colchicine. Neurologic examination revealed slight and asymmetric bilateral pyramidal syndrome, muscle rigidity involving the four limbs, bradykinesia and impaired postural reflexes. There was postural tremor in the extremities. Brain MRI showed mild Fazekas grade I cerebral and bilateral hippocampal atrophy. Intravenous bolus methylprednisolone (1 g/day) with cyclophosphamide (1g/day) was started. Progression was marked by a marked improvement in tremor and akinesia after the first bolus of intravenous methylprednisolone (1 g/day) with cyclophosphamide (1g/day).

Conclusion: The manifestations of NB are polymorphous. An extrapyramidal syndrome, although rare, may inaugurate the clinical signs of NB and provide a differential diagnosis with incipient degenerative disease.

Cardiac manifestations in Behcet's disease

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Introduction: Behçet disease (BD) is a chronic multisystem disorder. The occurrence of cardiac involvement in BD is rare, but appears to be underdiagnosed as autopsy studies in a Japanese series have demonstrated it in 16.5% of patients with Behçet's disease. Cardiac lesions affects the different tunics, dominated by pericardial involvement 20 to 40%,myocardial 20%,as well as coronary involvement and intracardiac thrombi.

Method: We retrospectively studied clinical data from patients with BD diagnosed between 2005 and October 2023 in the internal medicine department of IBN ROCHD university hospital center in 563 cases. All Patients fulfilled the International Study Group Criteria for the diagnosis of BD.

Discussion: Results: We recorded 22 cases of patients with cardiac involvement during BD (3,9 %) with a male predominance, the average age was 30 years. The average diagnosis time was 27 days. The clinical manifestations were dominated respectively by dyspnea, fever and hemoptysis. We found 17 cases of intracardiac thrombosis, 1 case with multiple intracardiac masses, 1 case of myocardial infarction, 1 case of myocarditis and 1 case of pericardial effusion and 1 case of thrombosed aneurysm of the right coronary artery. Biologically, the inflammatory syndrome was present in 80% of cases and Di-Dimers were elevated in half of the patients. All our patients had undergone a transthoracic heart echo and thoracic CT angiogram which confirmed the diagnosis. The other associated clinical manifestations were dominated by vascular involvement followed by ocular and neurological parenchymal involvement. All our patients were treated with boluses of corticosteroids and cyclophosphamide with a very good outcome with only 1 case of death from cardiac arrest. Anticoagulants were used only in cases without artery aneurysm.

Conclusion: The cardiac manifestations of Behcet's disease are diverse. Anticoagulation, immunosuppressant agents, and colchicine seem to improve the prognosis of cardiac manifestations in BD.

Successfully Switching to Adalimumab (ADA) for A Patient with Behçet's Disease Who Had Already Been Treated with Infliximab (IFX) for the Treatment of Uveitis with Secondary Failure and Experienced A Recurrence of Uveitis Accompanied by Dementia As Neuro-Behçet's Disease

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Introduction: The central nervous system involvement in Behçet's disease (BD), usually called neuro-BD (NBD) and therefore life-threatening.

Method: A 40-year-old (y/o) female urgently referred to our rheumatology division from ophthalmology with, dementia as a NBD recurring uveitis. When she was 34 y/o, her uveitis was occurred with oral aphthous ulcer with HLA-B51. Starting to treat with IFX for her uveitis of BD at her 35 y/o, she referred to the ophthalmology division in our hospital from another; however, her genital ulcers and headache using non-steroidal anti-inflammatory drugs those occurred at 38 and 39 y/o, respectively. Before around a half year, she was stopped taking colchicine with her anorexia. Nine days before, her left uveitis attack occurred; thus, a local steroid injection was performed; though, her condition worsened. At the day taking periodic IFX injection, she came to our hospital accompanied by her father and had impaired consciousness, her left eye
was index valve, and her various pathological reflexes were observed; thus, she was rushed to the emergency in our hospital. Using an interveinal methylprednisolone (IVMP) 1kg/day by 3 days for NBD, usually we called steroid-pulse, which was effective; thereafter, treatment with using oral prednisolone 1mg/kg/day and as a maximum dose of IFX (10 mg/kg/8 weeks), actually adding more IFX 5 mg/kg some days after. Though, these treatment except IVMP were no effective. Moreover, trying different types steroid and using interveinal cyclophosphamide (IVCY), there was no effect at all. Then, we decided switching from IFX to ADA, her condition was improved.

Discussion: There have been several reports on switching to biologics. Our case was similar to those reports. The patient had secondary ineffectiveness due to long-term use of IFX, and ADA change was effective.

Conclusion: We experienced a suffering case of NBD who treated with IFX for her uveitis; however, her BD developed neural lesions and switching to ADA.

National Epidemiology of Behçet's syndrome (BS) in the UK; a retrospective case control study using the Primary Care Clinical Practice Research Datalink (CPRD) and CPRD linked Hospital Episodes Statistics (HES)

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Introduction: UK CPRD and linked HES can be used to explore 'non endemic' BS epidemiology. Conditions with phenotypic and genetic overlap can also be captured with demographic factors during BS phenotype evolution toward a BS diagnosis code.

Method: After Ethics approval, using CPRD and CPRD linked HES (2001 to 2020) with Read code BS diagnosis, yearly point prevalence/incidence was derived. Retrospective matched case control design was used for covariates of interest. Controls (1:4 ratio; age, gender and practice matched) were used to examine phenotypically related MHC Class 1 Opathy conditions at baseline on BS risk using Logistic regression to calculate unadjusted odds ratios. Median time was calculated from onset of manifestations of interest to BS diagnosis.

Discussion: 4810 BS cases were coded. Prevalence increased; 12.5 (2006) to 19.07 per 100,000 (2020); 95% CI 19.93, 18.23. Incidence was stable, 0.89 per 100,000 (95% CI 1.09, 0.71). Both highest in women and mixed-race. Prevalence/incidence was highest in age 31–40 in 2020. 1752 BS cases in CPRD were compared with 5716 age/sex controls. Significant, increased risk of GU (OR 88.21), uveitis (OR 28.57) and OU (OR 27.50) in BS subjects was found compared to controls. Coded conditions with phenotypic and genetic overlap including Crohn's, UC, AS, ReA, PsA and enteropathic arthritis were significantly associated with BS compared to controls at baseline. Chronological appearance of phenotypic element prior to BS was derived. Shortest median time to BS was for GU appearance (85 days:IQR 0, 9412) and for OU (886 days;IQR 274.75, 16613).

Conclusion: UK BS prevalence in CPRD is higher than previously thought, consistent with our previous observations in THIN (1). Misclassification could be a factor. In CPRD/HES, coded BS phenotypes may emerge via multiple origins through conditions with genetic and phenotypic overlap. Improving key element recognition (AI) and a UK BS Registry could help. (1) Rheumatology (Oxford). 2020;59(10):2785-2795.

Unusual arterial presentations of Behçet's disease: 3 cases

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Introduction: BD: chronic systemic vasculitis + vascular tropism Arterial involvement 4 to 17%: mostly aorta. We report 3 cases with a vascular involvement expressed by aneurysms and stenosis.

Method: All our patients: male. Mean age at the diagnosis: 37 years old. Clinical manifestations: paraesthesias, claudication, dyspnea, dry cough, and dysphagia. Biological examinations: no inflammatory syndrome 3 patients. The diagnostic confirmation: CT scan. The locations presented in the study: a carotid artery aneurysm, occlusion of the subclavian artery, stenosis of the left internal carotid artery: 1st patient/ thrombosed fusiform abdominal aortic aneurysm, stenosis of the iliac artery + the superior mesenteric artery: 2d one /aneurysm of the aortic arch and a stenosis of the renal artery 3d one. Therapeutic side: 1 patient azathioprine AZA / 2 patients with aneurysms: methylprednisolone bolus, anti-Tumor necrosis factor TNF alpha (infliximab/ adalimumab). All patients: oral prednisone 1mg/kg/day+colchicine 1mg/day. A carotid-carotid bypass surgery: 1 patient. Favourable evolution: 3 patients. No deaths recorded.

Discussion: Arterial involvement: occlusions (38% to 80%), aneurysms (45% to 70%), pseudoaneurysms, stenoses (13%), aortitis (3%). Doppler ultrasound, scanner, magnetic resonance and Fluorodeoxyglucose-PET-CT angiography: imaging methods. The Arterial involvement's prognosis: severe. Isolated occlusive or stenotic peripheral arterial involvement: medical treatment often sufficient (corticosteroids+ immunosuppressants: AZA). Treatment of potentially life-threatening conditions (aneurysms): cyclophosphamide (CYC) and glucocorticoids. TNF blockers (infliximab and adalimumab): spectacular efficacy with severe refractory involvement to CYC. Surgery delicate+ frequent rate of complications. Endovascular treatment of aneurysms: increasingly recommended.

Conclusion: Arterial involvement in BD: rare Management: a major concern (high recurrence rate).

Sudden Memory Loss Revealing Neurobehçet Disease: Case-Report

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Introduction: Behçet's disease is a very common large vasculitis among mediterraneans and known for severe forms including the neurological and vascular involvement, this case-report highlights the importance of neuroimaging in diagnosing the severe central nervous system involvement and underlines its key feature that help rule out differential diagnosis.

Method: Our patient is a 44 years old female with known history of behcet's disease complaining of memory loss and convulsion, examination revelaed involuntary mouvements with torpor, biology consisted of inflammatory anemia, a brain MRI showed sub-cortical edema with heterogeneous enhancement of the internal left parieto-occiptal love with the splenium of the corpus callosum and other demyelenising supra tentorial regions. all were plausible with Neurobehcet's disease, ruling out then multiple sclerosis and neurosarcoidosis, she received 0,5 mg/KG of corticosteroids and monthly flush of IV cyclophosphamide (6Months) we noticed an impressive improvement of memory, muscle coordination and pain symptoms on follow-up and absence of flare-ups, and no opportunistic infections.

Discussion: The Diagnosis of Neurobehcet's disease is quite intuitive when the patient already underwent multiple complications like Angiobehcet and has been treatment- resistant, MRI and Lumbar Puncture reinforce the diagnostic hypothesis, the guidelines are case-based and follow the lines of Neurolupus, More research is to be done in this field of internal medicine and auto-inflammatory immunology.

Conclusion: More research is to be done in this field of internal medicine and auto-inflammatory immunology, and clinicians should be fully aware of ruling out neurobehcet's disease as a diagnosis of sudden memory loss bibliography: Corpus callosum lesion as the main clinical and radiological expression of Neurobehçet: A case report, DOI: 10.1016/j.neurol.2021.07.020

A Case of Pseudo-Behçet's in association with respiratory tuberculosis

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Introduction: One of the cardinal clinical manifestations of Behçet's Syndrome (BS) is orogenital ulceration, Oral and genital ulcers constitute a major criteria for the diagnosis of BS. However, orogenital ulcers can be seen in a variety of other conditions such as Herpes Simplex Virus (HSV) infection, Crohn's disease and reactive arthritis. Here, we present a case of a 24-year-old woman of Somali origin who was referred to the Behçet's Centre of Excellence with a working diagnosis of BS due to severe orogenital ulceration as a dominant symptom, which occurred at a time she was found to have latent tuberculosis.

Method: The patient presented with severe orogenital ulceration of several months, acne, and skin lesions suggestive of pustules and erythema nodosum. At the same time, she developed fevers, night sweats and was diagnosed with atypical pneumonia. Later in the course of the disease, the patient developed an episode of anterior uveitis, polyarthralgia and severe fatigue. The persistence of the constitutional symptoms resulted in the diagnosis of latent tuberculosis.

Discussion: Prior to attending the Behçet's clinic she had attended various clinical services including the Emergency Department, gynaecology, dermatology, rheumatology, and an emergency eye department. She was subsequently given a diagnosis of BS. However, initiation of treatment for tuberculosis resulted in the resolution of her symptoms. Whilst BS may not be entirely excluded, the absence of symptoms prior to the diagnosis of tuberculosis and after completion of treatment is against this possibility.

Conclusion: This case report highlights the challenges in the diagnosis of BS and the multifaceted presentation of tuberculosis. The possibility of alternative underlying pathology should be considered before the definitive diagnosis of BS and thorough assessment and investigation should be carried out by clinicians with appropriate expertise.

Behcet deep vein thrombosis in a patient with known Factor V Leiden mutation: The importance of clinical history

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Introduction: Behcet disease is often overlooked during thrombophilia investigations in deep vein thrombosis. Its presence can influence therapeutic strategy as immunosuppressive therapy is often required to prevent recurrence.

Method: We present the case of a patient with Factor V Leiden mutation and a history of deep vein thrombosis who presented a recurrence under anticoagulation.

Discussion: Ms B.K is 46 years old patient who was admitted for a chronic fever.History reveal a myocardial infarction 2 years prior which required stenting, and a deep right femoral vein thrombosis 1 year prior that was put on acenocoumarol, thrombilia testing at that time reveal a factor V Leiden mutation.Moreover the patient reported a recurrent bipolar aphthosis that started 1 year ago. Physical examination revealed a fever at 38 degrees celsius, a bipolar aphthosis, and a swollen left leg. Microbiology samples were negative. A doppler ultrasonography revealed a left deep femoral vein thrombosis. The diagnosis of a vascular Behcet disease was established and the patient was put on prednisone, azathioprine and acenocoumarol with a positive follow up Behcet disease related deep vein thrombosis (DVT) represent a challenge as studies have shown that anticoagulation alone may not prevent recurrence, which requires the use of immunosuppressive therapy as azathioprine and cyclophosphamide. This particularity sets it apart from other thrombophilia. As was shown in our case and reveal by patient's history, the first episode of DVT is related to Behcet disease, as was also confirmed by DVT recurrence under anticoagulation

Conclusion: Ruling out Behcet disease in deep vein thrombosis is an important step as its presence would require immunosuppressive therapy to prevent recurrence

Somatic JAK2 V16F mutations and the emergence or activation of a Behçet's phenotype (BS) in HLA B51 positive individuals developing in association with Polycythaemia Rubra Vera (PCV)

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Introduction: JAK kinases modulate inflammation through STATS. JAK2 V617F is a common mutation in PCV and ET, causing constitutive kinase activity, cytokine hypersensitivity and hematopoietic cell proliferation. Type 1 cytokines eg interferon alpha, signal through JAK2. JAK-STAT is key in inflammatory pathways, in cellular and antibody mediated response and in BS and IBD a target for JAK inhibition

Method: We report cases with JAK2 V617F mutations linked to PCV, both B51 positive. Case 1 (Female, 58) developed a BS phenotype with severe OU and GU prior to PCV diagnosis. Case 2, (Male, 45) with prior severe multi resistant BS age 19 (OU & GU, EN, pyoderma, thrombotic events) withdrew immunomodulators except for steroids, was lost to follow up but OU & GU remained in remission for 6 years until 1 year before PCV and severe OU & GU relapse despite prednisolone 20 mg daily. Peginterferon alpha 45 mcg weekly gave good haematologic responses, complete BS suppression and in case 2, steroid taper to 5 mg.

Discussion: BS is linked to primary myelofibrosis and myelodysplastic syndrome and trisomy 8 (1). We are unaware of BS cases with JAK2 V617F positive PCV and synchronous emergence or relapse of a BS phenotype and improvement with interferon alpha which causally links V617F mutation, myeloid cells and JAK/STAT signalling to a common pathway for both conditions. Galimberti found 34 patients with systemic autoimmune disease (IMIDS) amongst 435 patients with MPNs (86% JAK2 mutation positive) though none had a BS phenotype (2).Interferon alpha 2 b is established in treatment of ET and PV with remission in 70% and molecular response by mutant allele burden in 55% (ET) and 19% (PV). Mechanism remains unknown in PCV and BS.

Conclusion: Our cases indicate common mechanisms link both conditions. Targeting JAK is established in MPNs and our cases provide further rationale in BS. The facilitatory role of B51 and other MPN linked epigenetic changes are unknown. 1.Intern Med 61: 1713-1719, 2022 2.Clin Exp Rheum 40: 49-5517 2022

Local Vitamin D metabolic genes in Behçet's Syndrome

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Introduction: Vitamin D (Vit.D), originating from diet (Vitamin D2) or synthesized in the skin from cholesterol via UV radiation (Vitamin D3), is hydroxylated to 25-hydroxy vitamin D (25-OHD) by cytochrome P450 enzymes CYP2R1 and CYP27A1. Serum levels of 25- OHD reflect Vit.D status. 25-OHD is further hydroxylated by 1α-hydroxylase CYP27B1 into the active form of Vit.D. Active Vit.D (1α,25(OH)2D) binds to and activates the Vit.D receptor (VDR) in various tissues, mediating immune-inflammatory and metabolic processes. Both 25-OHD and 1,25(OH)2D3 are metabolized by CYP24A1 to less active compounds. This study aims to investigate the differences in local levels of CYP27A1, CYP2R1, CYP27B1, CYP24A1, and VDR in immune cells between Behçet's Syndrome (BS) patients and healthy controls (HCs).

Method: A total of 415 patient samples were investigated. Circulating 25-hydroxy vitamin D levels, T and B cell subsets, and immunoglobulin levels were measured. Total RNA isolated from patient and control PBMCs was analyzed using qPCR to measure the relative levels of CYP2R1, CYP27A1, CYP27B1, CYP24A1, and VDR genes.

Discussion: Serum Vit.D levels were significantly higher in 280 non-active BS patients compared to 135 active patients ($p = 0.032$). VDR levels were significantly higher in BS patients than in HCs ($p = 0.023$). CYP27B1 expression was significantly lower in the BS group compared to HCs ($p = 0.011$). These gene expressions did not significantly differ based on gender, BS activity, or phenotypes. Regression analysis revealed that azathioprine significantly suppressed CYP24A1 levels ($p = 0.030$), whereas patients on mycophenolate mofetil had significantly higher levels of VDR, CYP27A1, and CYP2R1. Adalimumab had a significant positive impact on CYP27A1 ($R2 = 0.44$, $p = 0.042$, $B = 0.29$).

Conclusion: The data indicates that BS patient medication impacts Vit.D metabolic genes. This suggests a role for the local Vit.D metabolic pathway in BS activity.

Inflammatory markers and their correlation with disease activity in Behçet's disease

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Introduction: Behçet's disease (BD) is an autoimmune systemic vasculitis of unknown origin. There are currently no markers available to assess disease activity in Behçet's disease, making its management more difficult. The aim of this study was to determine neutrophil/albumin (NAR), C-reactive protein (CRP)/albumin, platelet/lymphocyte (PLR), and lymphocyte/monocyte (LMR) ratios, and investigate their associations with clinical outcomes in patients with Behçet's disease.

Method: Eighty-one patients were recruited, and their IBDDAM and BDCAF scores were recorded. Patients were considered active if BDCAF> 2. The degree of severity was calculated according to the IBDDAM score. Blood samples were then taken. Statistical models were used to assess relationships between clinical findings and biological parameters, in order to search for possible correlations.

Discussion: This study included 74 patients in active phase and 7 patients in inactive phase. Active patients showed a non-statistically significant increase in mean PLR, NAR compared with the inactive group (P= 0.26 and P= 0.98) respectively according to the BDCAF score. Similarly, NAR, PLR and CAR levels tended to be higher in vascular, ocular, articular and neurological disease compared to patients without these conditions. There was a moderate positive correlation between PLR and CAR (p= 0 .008) and the degree of vascular damage. Similarly, MRL was strongly correlated with the degree of neurological damage (p=0.006). Regarding skin involvement, inflammatory markers were weakly correlated with its severity.

Conclusion: These potential inflammatory markers can be used to assess disease activity and severity in BD patients, and aid management.

Ocular involvement in behcet's disease and biotherapies

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Introduction: Behcet's disease is a multi-systemic vasculitis of multifactorial etiology. Its main symptoms are mucocutaneous, articular and ocular, as well as other disorders that can be life-threatening. Biotherapy has revolutionized the treatment of certain disorders, notably ophthalmological.

Method: This is a 10-year retrospective descriptive study of 10 patients with behcet's disease, selected according to international criteria, with ocular involvement who had received biotherapy

Discussion: The products received were infliximab and adalilumab. Indications were resistance to conventional first-line treatments and the severity of the clinical picture. Eight patients had a very good outcome, defined as regression of clinical signs and improvement in visual acuity. A relapse was observed in 1 patient. The condition of 2 patients remained unchanged. Treatment was ineffective in 1 patient. No intolerance was observed. Biotherapies used to treat the ophthalmological manifestations of inflammatory diseases include anti-TNF, anti CD20 and IFN-alpha. Several studies have demonstrated the efficacy of IFN-alpha (78-98% complete remission) in treating severe uveitis in behcet's disease. Anti-TNF agents require prolonged prescription or replacement by another immunosuppressant once ocular inflammation has been controlled, as their action is suspensive. The risk of infection, particularly tuberculosis in our context, associated with this biotherapy should limit its use to refractory forms.

Conclusion: Biotherapy is proving highly beneficial in the management of ocular forms of behcet's disease, which until now have been associated with irreversible functional sequelae.

The Socioeconomic Influence on Behçet's Syndrome Patients's Quality of Life

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Introduction: Employment contributes to patients' socioeconomic status and wellbeing, and the unemployed often face socioeconomic disadvantages. Behçet's Syndrome (BS) is a condition that affects young adults in the most productive period of their life. There is evidence to show that morbidity and mortality at a younger age are higher in people with lower education, lower occupational class and lower income. The objective of this study is to evaluate work-related outcomes associated with BS patients' self- rated health quality of life (HQoL) in both genders.

Method: A cross-sectional study using a digital survey method of 369 consented BS patients was carried out; females 77%: males 23%, mean-age= [41.1±23.3:38±13.2]. Data was collected from participants on their socio-demographic status, employment status, reasons for working, motivating factors at work and EQ-5D scores.

Discussion: BS females had significantly higher organ involvement than BS males; p= 0.014, and had significantly lower QoL than BS males; p value was < 0.001. BS patients in employment had significantly better QoL compared to the unemployed group. The average number of sick days due to BS was 39 days/6 months. There was a significant correlation between the importance of work and patients' QoL; R= 0.124, p=0.04. Most work motivation factors showed a significantly positive effect on patients' QoL. The main reasons that BS patients value employment were having a source of income, gives their lives daily structure, and prevents boredom. K-means analysis revealed; Cluster 1: older age group, casual and lowest grade workers who were most probably in remission. Cluster 2: BS severe group (blue collar); Cluster 3: well-educated (white collar) group.

Conclusion: Flexible and suitable employment for both genders of BS patients is important for patients to maintain physical and mental wellbeing, and self-esteem through their contribution to society. This improves their HQoL outcomes by reducing health inequalities related to their socioeconomic status.

Behçet disease, pulmonary tuberculosis and viral hepatitis C discovered concomitantly: Therapeutic strategy and challenges of treatment by the internist.

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Introduction: Behçet's disease (BD), independently of any immunosuppressive therapy, represents a state of immunosuppression favoring the occurrence of infection. In this observation, we report the circumstances of discovery of a Behcet disease associated to pulmonary tuberculosis and chronic hepatitis C,as well as their specific treatment methods and the therapeutic strategy rationally adopted.

Method: Mr. E. M., aged 33, single, with no notable pathological history apart from a notion of recurrent bipolar aphthosis, was hospitalized in Internal Medicine for unquantified weight loss and fever that had been going on for 3 months. The clinical examination was unremarkable apart from genital ulcer scars. The paraclinical assessment revealed the following results: normal CBC, CRP 64 mg/l, ESR = 43 mm, Positive BK sputum,positive anti-HCV antibodies positive with viral load = 6 log. Presence of aneurysmal dilatation of the thoracic aorta on CT angiography, normal liver and fibroscan studies. The diagnosis of vascular Behcet disease with pulmonary tuberculosis and chronic hepatitis C was retained. Therapeutically he was put on: anti tuberculosis drugs regimen, colchicine, corticosteroids and monthly bolus of
endoxan. Treatment of viral hepatitis C was deferred until antibacterial drugs were stopped.

Discussion: Although The diagnosis of BD can sometimes be straightforward, caution should be taken not to attribute every clinical manifestation as part of it. As was shown in our case, the patient clinical finding would have been compatible with a behcet disease,however investigations revealed a pulmonary tuberculosis that could have been

worsened by the immunosuppressive therapy required for Behcet disease.

Conclusion: Thinking of BD when faced with bipolar aphthosis is a useful reflex. However, when general signs are at the forefront of clinical manifestations, infection must remain the doctor's first fear, not only because of the underlying immunosuppression but also because of immunosuppressive treatments.

Clinical and Prognostic Profile of Behçet's Disease (A Series of 111 Cases)

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Introduction: Behçet's disease (BD) is a complex systemic vasculitis of unknown etiology. It is characterized by various clinical manifestations, including cutaneous-mucosal, ocular, articular, vascular, and neurological manifestations.

Method: This was a retrospective study involving 111 cases of Behçet's disease meeting the revised 2014 International Criteria for Behçet's Disease. Patients were recruited from the internal medicine department of Oued Eddahab Military Hospital (Agadir) and Hassan II Regional Hospital (Agadir).

Discussion: A total of 111 cases were included in this study. The male-to-female ratio was 1.5. The mean age at the time of Behçet's disease diagnosis was 35.2 years (range: 14–73 years), and the mean time to diagnosis of BD was 5 years. Cutaneous-mucosal manifestations were found in 100% of patients. The distribution of other manifestations was as follows: • Vascular involvement, affecting 33 patients (29.7%) • Ocular involvement in 31 patients (27.9%) • Articular involvement in 29 patients (26.1%); • Neuro-Behçet in 6 patients (5.4%) and 28 cases of neuropsychiatric manifestations observed mainly in male subjects; • Four cases of bilateral optic neuropathy (3.6%); • Digestive involvement in one patient (0.9%). All patients were on colchicine (100%). 69 patients (62.1%) received systemic corticosteroid therapy. Azathioprine was prescribed in 18 cases (16.2%), cyclophosphamide in 16 cases (14.4%), anti-TNF alpha agents for 8 patients (7.2%), and anticoagulants for 19 patients (17.1%).

Conclusion: Our study confirms epidemiological observations made in other countries of the Mediterranean region regarding Behçet's disease. The results confirm the male predominance of the disease, the high prevalence of cutaneous-mucosal manifestations, and half of the patients presenting with severe manifestations, especially ocular and vascular. However, parenchymal neurological and digestive involvement were less frequent in our series.

Comparison of activity scores in the follow-up of patients with Behçet's disease

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Introduction: Behçet's disease (BD) is a multi-systemic vasculitis whose activity remains difficult to assess. As a result, accurate assessment of this activity remains essential for proper therapeutic management. The aim of this study was to determine the concordance between the IBDDAM, the BDCAF, the Turkish score and the expert physicist's global assessment (PGA) in the evaluation of changes in BD activity.

Method: Patients with BD were evaluated by IBDDAM, BDCAF and Turkish score at two consecutive follow-up visits. The interval between the two visits varied from 3-6 months. The change of disease activity was determined (increased, unchanged or decreased) according to the PGA. We used the ROC curve to determine an appropriate cut-off point for change in disease activity.

Discussion: Eighty-six patients were evaluated at their two consecutive follow-up visits. There was a highly significant positive correlation between Turkish score, BDCAF ($p < 0.001$) and IBDDAM ($p < 0.001$) in both consecutive visits with a stronger correlation between Turkish score and IBBDAM ($p < 0.001$). This positive correlation ($p < 0.001$) was also objectified in the change in score activity with a better correlation between Turkish score changes and IBBDAM. Comparison of the area under the ROC curve showed a statistically significant difference between the 3 scores ($p = 0.007$). The best cut-off for the assessment of activity change was 1.5 for all 3 scores, with better sensitivity (82%) and specificity (66%) for the IBDDAM. A better statically significant agreement was also observed for IBDDAM with PGA compared with the other scores; while considering the calculated cut off.

Conclusion: IBDDAM is the preferred method for assessing the evolution of disease activity in our BD patients, compared with PGA.

Investigating the Role of Nutrition and Olfactory Function in Behçet's Syndrome Patients

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Introduction: Nutrition and immune-related conditions are being studied to determine how dietary macronutrients contribute to health. Olfactory function, which affects diet and food choices, may be linked to immune-related disorders. The relationship between diet and olfactory function in Behçet's Syndrome (BS) patients and its impact on disease activity is not well understood. This study aims to explore the influence of diet on inflammation and olfactory function in BS patients.

Method: A 24-hour dietary recall software was used to compare the diets of 75 BS patients and 35 healthy controls (HC). Serum short-chain fatty acids (SCFA) were measured using ELISA. GPR41 and GPR43 receptor levels were quantified by qPCR. The UPSIT smell test assessed olfactory function in the cohort.

Discussion: Dietary intake and olfactory function were not significantly different between BS patients and HCs. However, a significant majority (73%) of patients consumed pro- inflammatory foods, and 75% did not meet the recommended fibre intake. Severe microsmia was observed in BS patients with major organ involvement. An inadequate ω-3/ω-6 ratio was found in 80% of patients. Notably, ω-3 and ω-6 levels were significantly lower in patients with moderate and severe microsmia compared to those with mild microsmia (P = 0.04 and 0.01, respectively). There was no significant difference in SCFA levels between BS patients and HCs. BS patients exhibited lower gene expression of the GPR41 receptor compared to HCs (P = 0.04). Active BS patients had significantly lower GPR41 and GPR43 gene expression than non-active BS patients (P = 0.02 and 0.015, respectively). HCs did not show significant microsmia.

Conclusion: Most BS patients consumed a pro-inflammatory diet, which may be connected to BS activity. Impaired GPR41 and GPR43 function in BS patients warrants further study. Maintaining ω-3/ω-6 levels is important for controlling BS disease activity and enhancing olfactory function. Those with major organ involvement exhibited significant microsmia.

Routine Immuno-Haematology Profile Results in Behçet's Syndrome

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Introduction: There is little published work on routine immuno-haematology results in Behçet's Syndrome (BS). Immunology and Haematology blood tests are performed on samples taken from all patients attending the London Behçet's Centre. In this study we aim to determine the relevance of measuring lymphocyte subsets, full blood count and Immunoglobulins in patient management, and the correlation with clinical features.

Method: 909 samples were taken from 635 patients, 60% female and 40% male. 43% were longitudinal samples. Lymphocyte subsets, i.e., absolute counts and percentages of CD3+ T cells, CD3+ CD4+ T cells, CD3+ CD8+ T cells, CD19+ B cells and CD16+CD56+ NK cells, full blood count and Immunoglobulins (Ig) G, A and M were requested over 3 years. Testing and analysis were performed by Immunology, Haematology and Clinical Chemistry departments in the East and South East London Pathology Partnership at the Royal London Hospital.

Discussion: Preliminary analysis compared results to published reference ranges. At least one parameter was outside the references ranges in 92% of all samples and 90% of first samples. Lymphocyte subsets: T cell results were raised in 18%, and B and NK cells results were reduced in 19% and 33% of samples. Full blood count: Neutrophils were raised in 17%, and lymphocytes were reduced in 12% of samples. Immunoglobulins: IgA and IgM were often raised. Comparison of longitudinal samples demonstrated stable lymphocyte subsets with no significant difference between first and second results. There was a significant difference between lymphocyte and neutrophil counts in the full blood count panel. Levels of IgG and IgA were significantly different.

Conclusion: The high number of samples with results outside reference ranges suggests that routine enumeration of Immuno-Haematology profile is relevant to management of BS. Further analysis will aim to analyse the association of lymphocyte subsets with symptom severity scores and investigate the impact of clinical outcomes on panel results.

Pulmonary embolism revealing an intracardiac thrombus: an exceptional complication in Behcet's disease

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Introduction: Pulmonary embolism (PE) is an unusual event in Behcet's disease, and the presence of an intra-cardiac thrombus (ICT) should always be suspected. It is a serious, life- threatening complication.

Method: Our retrospective study collected four cases of pulmonary embolism complicating ICT among 50 patients meeting the criteria of the ICBD 2013, followed up in the Internal Medicine Department of CHU SOUSS MASSA between 2020 and 2023.

Discussion: All our patients were male, and the mean age at diagnosis of PE was 38 years. PE revealed MB in 2 patients. PE was secondary to ICT, which was discovered by transthoracic echocardiography. PE manifested as chest pain and hemoptysis in all patients, and dyspnea in 3. Diagnostic confirmation was based on thoracic angioscan. PE was associated with multiple pulmonary artery aneurysms in 2 patients and Hughes-Stovin syndrome in one. In terms of treatment, all patients received a monthly bolus of corticosteroids and cyclophosphamide, one patient recieved infliximab, and anticoagulation was started in 2 patients only (the other 2 had associated aneurysms). The clinical course was marked by the disappearance of symptoms and the absence of recurrences in all our patients. Radiological monitoring after 6 months of treatment in 2 patients showed arterial repermeabilisation and complete resorption of thrombi. No deaths were reported. PE is most often the consequence of an in situ thrombosis, or a complication of a ICT, which should always be detected by echocardiography in the case of PE in BD. It may also be associated with pulmonary aneurysms, which makes anticoagulant treatment a challenge, given the high risk of hemorrhage. The course of the disease is favorable with medical treatment, subject to early diagnosis.

Conclusion: Our work highlights the potential benefit of echocardiography in the assessment of BD patients presenting with PE, so that a TIC is not missed.

Angio-Behçet

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Introduction: Vascular involvement constitutes a clinically, prognostically, and therapeutically significant manifestation in Behçet's disease (BD).

Method: We collected 111 cases of BD over a 5-year period. 33 patients had angio-Behçet. The study seeks to analyze their epidemiological and clinical characteristics to identify potential risk factors and therapeutic outcomes.

Discussion: The male/female sex ratio was 3.7. The average age at diagnosis was 30.6 years. In 87.9% of cases, vascular involvement revealed BD. Concerning venous involvement (n=28), thrombosis was the most frequent: lower limb deep vein thrombosis (n=18), unusual locations (n=10) superficial vein thrombosis (n=2). Arterial involvement mainly included pulmonary aneurysms (n=3), one renal artery aneurysm, and one coronary artery aneurysm. Intracardiac thrombus (n=3) were all complicated by pulmonary embolism. One case of Hughes-Stovin syndrome was noted in our series. Corticosteroids and colchicine were the mainstay of treatment, often combined with other immunosuppressive agents. Infliximab was used in three patients: One as a first-line treatment and for the others, it was initiated after the failure of immunosuppressants or during a relapse. The prognosis of patients with angio- Behçet varied, with most of them responding well to medical treatment, while others required surgical intervention.

Conclusion: Angio-Behçet is a mode of revelation of BD. Arterial involvement, although not rare in our context, is often associated with venous involvement and vascular involvement is often associated with genital aphthae, highlighting the importance of syndromic groupings within BD. Further research is needed to better understand the pathophysiology of vascular involvement in BD and to develop more effective therapeutic strategies.

Rare and unexpected case of Ocular Behçet's disease

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Introduction: Ocular involvement in Behçet's disease is polymorphous, severe and life-threatening, affecting all tunics of the eye. Many types of ocular involvement have been described, but granulomatous involvement is exceptional.

Method: We report the case of granulomatous uveitis in a young patient with Behçet's disease.

Discussion: A 21-year-old girl presented with a red eye and bilateral severely decreased vision. Ophthalmological investigations revealed a visual acuity at 6/10 with granulomatous anterior uveitis and retinal vasculitis. Extraocular signs included recurrent oral aphthosis, without polyarthralgia or skin rash. Infectious work-up (viral and bacterial serologies, phthisiology assessment), immunological work-up and angiotensin- converting enzyme assays were negative. Thoracic imaging was normal. The HLAB51 antigen test was positive, and the diagnosis of Behçet's disease was accepted. Our patient received corticosteroid and immunosuppressive therapy.

Conclusion: The eye is one of the main targets of Behçet's disease, dominated by uveitis and vascular complications. The existence of granulomatous character is exceptionally reported in the literature. This clinical case opens the discussion to think of Behçet's disease in front of a granulomatous uveitis especially in the presence of an evocative clinical and epidemiological context, but the need for larger studies is imperative.

The role of monocytes in resolving inflammation in Behçet's Syndrome

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Introduction: Behçet's Syndrome (BS) is an immune mediated vasculitis. The cells of the innate immune system including neutrophils, NK cells and monocytes have all been implicated in the pathogenesis of BS. Monocytes have been reported to play a significant role in driving inflammation in BS however, very little is known about how monocytes resolve inflammation in BS. We therefore hypothesize that monocytes are not effective at propagating resolution in BS.

Method: Isolated monocytes from 49 BS patients and 24 Healthy controls (total monocytes, its subsets (Classical (CM), Intermediate (IM) and non-classical (NCM)) and a few surface markers (CCR2, CX3CR1, CD11b, HLA-DR) were phenotyped by flow cytometry. Chemotactic and phagocytic ability of BS monocytes to take up microbes and apoptotic cells (efferocytosis) were investigated by Boyden chamber and flow cytometry respectively.

Discussion: Results show reduced expression of chemokine proteins CCR2 and CX3CR1 on monocytes in BS, which are relevant in the recruitment of monocytes in the inflammatory phase and resolution phase respectively. Results from chemotaxis assay shows aberrant migration of monocytes in BS patients towards recombinant CCL2. Most importantly, we reported a reduced ability of classical monocytes in BS to effectively phagocytose apoptotic cells (Efferocytosis).

Conclusion: Efferocytosis is a key driver of resolution, and our study suggests that monocytes in BS are defective at initiating resolution. With further mechanistic investigation of efferocytosis by monocytes, a new insight on approaching the management of inflammation in BS may be unraveled by harnessing the ability of the immune system in dampening inflammation by resolution.

Epidemiological, clinical, and therapeutic characteristics of arterial involvement in Behçet disease: a study in an internal medicine department

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Introduction: Behçet's disease (BD) is a vasculitis affecting venous and arterial vessels of all calibers. Arterial involvement, which is rarer, can be severe and life-threatening, warranting urgent treatment. The aim of our study is to describe the epidemiological, clinical, therapeutic and evolutionary characteristics of arterial involvement in this disease

Method: This is a retrospective, descriptive, monocentric study ruled in the department B of internal medicine at Military Hospital of Instruction of Rabat from 2013 to 2024. The diagnosis of BD was based on Behcet's Syndrome International Study Group Criteria and arterial involvement was documented by arterial duplex and/or CT angiography.

Discussion: Among 115 cases of MB, 40 cases of vascular involvement were recorded (34%), of which 10 (9.5%) patients with arterial involvement. These included 9 men and 1 woman. The average age at diagnosis was 25 years, with extremes of 22 and 56 years. Aneurysms were located in the pulmonary artery in 4 cases, the common femoral artery in 2, the common carotid artery and the splenic artery in 1 case. Arterial thrombosis occurred in 3 patients, with 2 cases of intracardiac thrombus and 1 case of pulmonary artery thrombosis. Vascular involvement was mixed in 2 patients. The associated systemic signs included bipolar aphthosis in all patients, ocular involvement in 4, joint involvement in 5 and neurological involvement in 1. All patients were treated with corticosteroids combined with immunosuppressive immunosuppressive (cyclophosphamide or azathioprine). Complications included the death of one of our patients following rupture of his pulmonary artery aneurysm with cataclysmic hemoptysis.

Conclusion: Arterial involvement in Behçet's disease is rare, and can lead to serious complications with life-threatening consequences. It is therefore crucial to be aware of it and, above all, to search for it, so that appropriate treatment can be offered quickly.

Comorbidities and their impact on Behçet's Syndrome activity, Quality of Life, Mental Health and Social Adjustment of the Patients

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Introduction: Comorbidities in Behcet's Syndrome (BS) present a challenge to clinicians to assess and manage BS patients. The objectives of this study were to investigate the prevalence and comorbidity clusters in BS patients. In addition, to investigate the impact of comorbidities on patients' mental wellbeing and Quality of Life (QoL).

Method: Data was collected from a single-centre cross-sectional study of 515 consented BS patients and included patient demographics, number and type of comorbidities, surgical procedures, BS clinical parameter outcomes, patients' lifestyle, mental and psychological scores, and EQ-5D scores.

Discussion: 81% of BS patients had comorbidities; 61% had 1-2, 29% had 3-4 and 10% had 5-6. The number of comorbidities were significantly higher in females compared to males; p=0.006. The distribution is in the vein with the general population. However, some comorbidities were distributed to a larger extent in males than females including cardiovascular conditions, Raynaud's phenomenon, and chronic obstructive pulmonary disease. Those in the older age group had significantly more cardiovascular, and immune-related diseases than younger groups; p=0.001 each. Hysterectomy, cholecystectomy and appendicectomy were the most common surgical procedures. BDCAF scores increased significantly with comorbidity number, and were associated negatively with patients' QoL. The lowest EQ-5D scores were found in BS who had immune-related disease, psychological conditions and cancer. A significant positive relationship was found between the number of comorbidities and PHQ9 and Work and Social Adjustment scores; $R2=0.75$, $p<0.05$ and $\beta=1.0156$; $R2=0.814$, $p-<0.01$ and $\beta=2.27$ respectively.

Conclusion: This study sheds a light on comorbidities as key factor in BS management, and affects the interpretation of clinical research findings. Comorbidities in BS have an impact on disease activity, patients' QoL, employment status and depression. This suggests that better control of comorbidities improve BS outcomes.

A giant coronary aneurysm complicating Behcet's disease

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Introduction: Behcet's disease (BD) is a systemic vasculitis that can affect arteries and veins of all calibers. Cardiac involvement in Behcet's disease has been described, but is clearly associated with a poor prognosis. Among the cardiac manifestations of BD, coronary involvement is considered rare, if not exceptional.

Method: We report a new case of coronary aneurysm complicating Behcet's disease.

Discussion: Young patient aged 27, with no cardiovascular risk factors apart from his male sex, followed for Behçet's disease since 2019 associated with ankylosing spondylitis. BD was revealed by thrombophlebitis of the left lower limb, and re-interrogation revealed recurrent bipolar aphthosis. He was admitted for management of an acute coronary syndrome, with a very high troponin level, a thoracic angioscan showed a giant aneurysm measuring 5.42 cm in the coronary arteries. In view of the severity of the vascular damage, a bolus of methylprednisolone followed by anti-TNF alpha- based biotherapy was introduced, and the patient is currently on his fourth course of infliximab. No cardiovascular events occurred during this follow-up period, with a reduction in the size of the aneurysm during a control CT scan performed at the third course. Coronary involvement in Behcet's disease is rare. It most often affects young male subjects under the age of 40, with no risk factors for atherosclerosis, as in the case of our patient. Treatment of this particularly serious condition is based on corticosteroids and immunosuppressants, or even biotherapy, and must be introduced rapidly to avoid a fatal outcome.

Conclusion: Coronary aneurysm is a rare manifestation and a turning point in the evolution of MB. The diagnosis must be strongly suspected in the event of a myocardial infarction in a young patient and in the presence of atypical coronary lesions, particularly aneurysms.

Patient adherence and compliance with Triorasol Mouthwash

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Introduction: Oral ulcers are a common feature of Behçet's Syndrome (BS), significantly impacting patients' quality of life. Triorasol mouthwash is an effective topical agent for managing oral ulcers in BS patients, improving their quality of life. The aim of the study was to assess the adherence and compliance rates of Triorasol mouthwash usage amongst patients with BS.

Method: An anonymous, prospective questionnaire was given to 50 BS patients. Demographic data including age, sex, ethnicity, and employment status was collected. Patients were queried on their utilisation of Triorasol mouthwash, perceived ease of use, and any challenges encountered.

Discussion: Of the 50 patients surveyed, 46% reported using Triorasol mouthwash. Factors influencing adherence and compliance included understanding of usage instructions and accessibility to instructional materials. For example, 19% patients use the mouthwash four times daily when they have ulcers in accordance with the mouthwash instructions, 22% use the mouthwash three times daily, 22% use it twice daily, 8% use it once daily and 29% do not use it at all. 4% of patients swallow the mouthwash which they are advised not to do.

Conclusion: Adherence and compliance with Triorasol mouthwash among BS patients is crucial for optimising treatment outcomes and enhancing quality of life. Novel patient education initiatives including an instructional video and information leaflet have enhanced patient's understanding of the use of Triorasol mouthwash, and its importance in managing their disease.

Behçet's disease: experience in an internal medicine department (60 cases)

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Introduction: Psychiatric involvement in Behçet's disease (BD) is relatively common (20% to 30%). It may include depression, anxiety, mood disorders, psychoses and cognitive disorders. Linked either to the impact of the chronic, disabling disease, the side effects of treatment, or neurological damage due to vascular inflammation affecting the central nervous system. The aim of our work is to describe the clinical aspects of psychiatric disorders in BD.

Method: Retrospective study including 7 patients hospitalized in our internal medicine department, between 2019 and 2024 presenting with BD with psychiatric disorders, whose diagnosis was established following a specialized psychiatric consultation.

Discussion: In our series, there were 3 men and 4 women, giving an M/F sex ratio of 0.75. The mean age of onset of MB was 30 years. Psychiatric involvement was not indicative of MB in any of our patients. The mean age of onset of psychiatric disorders was 37 years. All patients had skin involvement, predominantly oral aphthosis, 57% had joint involvement, 28% ocular involvement, 14% vascular involvement and 42% neurological involvement. Four cases of depression were noted (57%), including one depression associated with an anxiety disorder, two cases of moderate depressive syndrome and a single case of melancholic depression. Two cases showed agitation (29%). One case showed psychomotor excitement (14%). None of the patients in our series presented with hallucinations or hysterical seizures. Neurological etiology was considered in 43% of patients. Psychiatric disorders were treated with neuroleptics, anxiolytics and antidepressants. The course of treatment was favorable in all our patients.

Conclusion: Psychiatric disorders in Behçet's disease show great clinical polymorphism. They occur mainly in patients with neurological impairment. However, they may also be related to the emotional stress, anxiety and depression experienced by patients suffering from BD, necessitating appropriate psychological management.

Safety and efficacy of Infliximab in patients with Behcet's disease

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Introduction: The manifestations of Behcet's disease (BD) can be life threatening and functionally challenging. The aim of our study was to describe the clinical and paraclinical characteristics and evolution of patient with BD treated by Infliximab, and to compare them with literature.

Method: This is a retrospective cross-sectional study of five patients with BD treated with infliximab from 50 cases of BD, between October 2019 and January 2024. The cases were collected From the Internal medicine department of HASSAN II HOSPITAL AGADIR. All of our patients met the international criteria of the ICBD 2013.

Discussion: All patients were male with a mean age of 30,6 years. Mucocutaneous involvement was present in all patients. Ocular manifestations in one patient (refractory uveitis), vascular involvement in four patients including three multiple pulmonary artery aneurysms and one coronary aneurysm, with no articular or neurological involvement. The indication of infliximab (IFX) were ocular involvement n= one (20%), vascular involvement n= four (80%); they were indicated as first-line therapy in two patients (40%); and three (60%) were already on immunosuppressives when the vascular lesion requiring IFX developed. The evolution was marked by remission in four patient (80%); and death in one patient with a profuse hemoptysis. No side effects was observed.

Conclusion: Infliximab seems to be effective in majority of BS patients with severe involvement, even in those who are refractory to immunosuppressives and glucocorticoids. However, other studies are needed in terms of length of infliximab regimen, whether or not it should be discontinued, and, if so, whether or not immunosuppressants should be given as maintenance after discontinuation.

Angio-Behcet: thrombosis in unusual locations, about 09 cases

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Introduction: Vascular involvement is common in Behçet's disease (BD), affecting up to 40% of patients depending on the series. Deep vein thrombosis (DVT) of the lower limbs predominates. Our aim is to describe thrombosis in BD, excluding DVT of the lower limbs.

Method: We retrospectively collated nine cases of vasculo-Behçet of unusual location in our department over a period from 2019 to 2023.

Discussion: Nine cases of unusual thrombosis were selected. There were two female and seven male cases, with a clear male predominance. The mean age at diagnosis was 34,5 years (range 16-53 years). The diagnosis of vascular involvement was confirmed by Doppler ultrasound and/or angioscan. Bipolar aphthosis was present in all patients. Intracardiac thrombosis was found in four cases (44.44%), cerebral thrombosis in 2 cases (18.18%), Budd-Chiari syndrome in 1 case, jugular thrombosis in 1 case and portal vein thrombosis in 1 case. All our patients received corticosteroid-based treatment in combination with other therapies: immunosuppressants, anticoagulants and colchicine. Four patients received anti-TNF alpha biotherapy. The evolution was marked by stabilisation and even remission for our 8 patients, one patient was lost to follow-up. No deaths were reported.

Conclusion: Thrombosis in unusual locations is a complication of BD, affecting young men in particular, and in the early years of the disease in most cases. Far from being rare, these types of incolvement are particularly serious, and the prognosis for the disease is very poor. These manifestations require the use of immunosuppressants, or even biotherapy, particularly for venous disease of the large trunks, arterial disease and cardiac disease.

Ocular Manifestations of Behçet's Disease

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Introduction: Behçet's disease (BD) is a chronic, multi-systemic inflammatory vasculitis characterized by the variability of cutaneous and articular manifestations, and the potential severity of visceral involvement, particularly ocular involvement. Ocular Behçet's disease can be severe and visually life-threatening.

Method: We conducted a retrospective descriptive study of patients with confirmed BD admitted to the internal medicine department at Agadir University Hospital from January 2019 to January 2024. The study included 111 patients with BD, of which 31 had oculo-Behçet. The aim of this study was to investigate the epidemiological, clinical, and evolutionary characteristics of oculo-Behçet.

Discussion: A total of 111 patients were enrolled in the study, of whom 31 (27.9%) had oculo- Behçet. The median age of onset of ocular involvement was 34.6 years (range: 11- 65 years). The male-to-female ratio was 1.21. The mean interval between the first sign of Behçet's disease and the onset of ocular involvement was 4.2 years (range: 1 month-20 years). Ocular symptoms were dominated by decreased visual acuity and/or visual blur (87% of cases), followed by ocular redness (74.2%) and peri-ocular pain (25.8%). Ocular involvement was bilateral in 22 patients (70.9%). Uveitis (anterior, intermediate, and posterior) was the most prevalent ophthalmic manifestation, affecting 93.5% of patients, with 48.4% presenting with pan-uveitis. Retinal vasculitis was present in 12.9% of cases. Most patients (70.9%) developed ophthalmic complications, including blindness (29% of cases), cataract (16.2%), papillitis (6.4%), and maculopathy (19.3%).

Conclusion: Uveitis, particularly pan-uveitis, is the most frequent and potentially vision- threatening ocular manifestation of Behçet's disease. Early diagnosis and appropriate management are crucial for improving the ocular prognosis and preventing vision loss.

Shifting patterns in new Behçet's patients: two snapshots five years apart

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Introduction: The UK has seen unprecedented social upheaval over the past five years: the Covid- 19 pandemic, Brexit, global conflict, and economic recession. This has a potential impact on new patients attending the London Behçet's Centre of Excellence.

Method: Convenience samples of 80 patients newly diagnosed with BS at the London Behcet's Centre of Excellence between May 2023-2024 and May 2018-2019 were taken. Retrospective record analysis was carried out. Oral ulcer severity and arthritis pain score at first assessment were also recorded from a sample of new patients each year from 2018-2023. Data was analysed using the Z-test for two proportions.

Discussion: A significant decrease in the proportion of new BS patients identifying as White British was seen in 2023-24 compared to 2018-19 (9% vs. 22%, z= -2.93, p= 0.003). This was accompanied by an increase in patients identifying as Middle Eastern, African or South Asian in 2023-24. There were no significant changes in age or gender. There was an increase in working-age patients reporting unemployment in 2023-24, although this was non-significant (23% vs. 6%, z= 1.50, p= 0.13). An increasing trend in ulcer severity score at first assessment was seen from 4 in 2018 rising to 18 in 2023. Arthritis pain score also changed over this time-period.

Conclusion: Oral disease severity in newly diagnosed BS patients may be increasing. An increase in ethnic minority groups being diagnosed with BS in 2023-24 may reflect increasing migration, improved access to healthcare and BS awareness in minority groups. Economic recession and national unemployment may also have an effect on BS patients' health outcomes. Clinicians should be aware of the changing needs of BS patients to provide holistic care.

Behçet's Syndrome and Recreational Drugs

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Introduction: Some patients take recreational drugs in addition to their prescribed Behçet's medication, which if not disclosed to healthcare professionals may adversely impact their management. The aim of this study was to evaluate the prevalence and patterns of recreational drug use amongst Behçet's patients, along with exploring associated mental health comorbidities.

Method: Data was collected from 52 Behçet's patients at the London Behçet's Centre of Excellence at The Royal London Hospital. A pilot study showed patients' reluctance to share information on drug use. Therefore, an anonymous prospective questionnaire was given to patients in either paper or digital format. This included questions on demographics, drug usage rates, administration methods, concurrent mental health conditions, and the presence of comorbidities.

Discussion: The majority of participants were female, aged 26-33, employed full-time, homeowners, and married. Among the respondents, 9 out of 52 admitted to recreational drug use, with cannabis being the most commonly used substance (88.9%), followed by cocaine and ecstasy (55.6% each). Oral ingestion (66.7%) and smoking (66.7%) were the primary routes of drug administration, while 44.4% reported snorting drugs. Additionally, 44.4% used multiple drugs simultaneously, and 55.6% reported feeling dependent on these substances. Most users consumed drugs infrequently, with 60% using them once or less a month. Notably, all patients who attempted to quit successfully did so, and 60% reported no concurrent mental health issues.

Conclusion: The findings highlight a significant proportion of Behçet's patients engaging in recreational drug use. Further investigation is warranted to understand the implications of drug use on Behçet's Syndrome and potential drug interactions. Moreover, interventions to encourage patients to disclose drug use and provide drug cessation support are recommended to enhance patient care and outcomes and ascertain the impact on their health.

Awareness of risk of elective cosmetic treatment amongst Behçet's patients

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Introduction: Many patients at the London Behçet's Centre have been noted to have undergone surgical and non-surgical cosmetic procedures, which may lead to activation of their Behçet's Syndrome (BS) due to a hyperactive immune response to injury. The aim of the study was to assess the prevalence and types of cosmetic procedures Behçet's patients are having, and their understanding of the risks involved.

Method: Data was collected prospectively from patients at the London Behçet's Centre of Excellence. They were asked to complete an anonymous questionnaire either in paper or digital form. Questions were asked on demographics, co-morbidities, previous cosmetic procedures, desire to have treatment in future and awareness of risks.

Discussion: 42 patients completed the questionnaire. 10% of patients had previously undergone elective cosmetic procedures, with breast augmentation being the most common (40%). Despite potential risks, none of the patients consulted their Behçet's specialists before their procedures, although they did not report any post-operative disease activation. 75% would consider further elective cosmetic procedures in future.

Conclusion: The decision for patients to undergo cosmetic procedures is complex, and involve psychosocial, behavioral and physical components. These results highlight the importance of patient education to ensure that any cosmetic procedures are carried out safely, with optimal medical control of their BS to prevent disease activation.

Behçet's disease: experience in an internal medicine department (60 cases)

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Introduction: Behçet's disease (BD) is a multi-systemic vasculitis of unknown etiology affecting vessels of all calibers, both veins and arteries. The aim of our study is to describe the epidemiological, clinical, therapeutic and evolutionary profile of this disease.

Method: Retrospective and descriptive study conducted in our internal medicine department, covering all patients hospitalized for BD over a 5-year period from 2019 to January 2024.

Discussion: A total of 60 patients were included in our study, 48 males and 12 females, sex-ratio m/f: 4. The mean age at diagnosis was 30 ± 8 years, with ages ranging from 12 to 50 years. In addition, we report 2 cases from the same family (twin sisters). Clinical manifestations were dominated by mucocutaneous involvement, mainly oral aphthosis, present in all patients (100%) and inaugural in 75% of cases. Joint involvement came second with a frequency of 56.7%, followed by ocular involvement in 45% of cases, angio-Behçet in 40%, neuro-Behçet in 18.3%, entero-Behçet in 15% and psychiatric involvement in 11.6%. In terms of treatment, almost all our patients benefited from colchicine (97%), while 80% required corticosteroid therapy. Anticoagulants were used in patients with thromboembolic manifestations (31% of patients). The use of immunosuppressants, mainly azathioprine, followed by methotrexate and cyclophosphamide, was noted in 40%, 5% and 5% of patients respectively. Biotherapy, notably anti-TNF, was prescribed in 10% of cases. In our series, the evolution was favorable, with lesion stabilization in 70% of cases, whereas it was unfavorable, marked by frequent relapses in 18% of cases. Two patients (men) with severe vascular damage died during the course of the disease. Five patients were lost to follow-up.

Conclusion: Our study confirms the clinical polymorphism of BD, which encompasses various systemic disorders, the most serious of which are vascular, ocular, neurological and digestive, justifying the use of immunosuppressive drugs with varying degrees of mortality.

Correlation Between Mucocutaneous Manifestations, Systemic Involvement, and Disease activity in Behçet's Disease

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Introduction: Introduction Behçet's disease, a systemic inflammatory vasculitis of unknown etiology, commonly presents with cutaneous and mucosal symptoms crucial for diagnosis. Our study explores these manifestations and their correlation with systemic involvements in patients at our internal medicine department.

Method: This retrospective study analyzed patient data from January 2010 to December 2023, using the International Criteria for Behçet's Disease for inclusion (ICBD).

Discussion: Our study included 399 patients (210 men, 189 women) aged 16 to 60 years (average 32 years). Mucocutaneous symptoms appeared in 87.9% of cases, with 100% presenting oral aphthosis, 75.1% genital aphthosis, 55.6% pseudo-folliculitis, and 27.5% erythema nodosum. Rare cases showed Raynaud syndrome, skin ulceration, and vasculitic purpura. Systemic manifestations included vascular (24%), ocular (23%), joint (21%), neurological (14.7%), cardiac (1.75%), and digestive (1.5%) involvement. Our findings suggest mucocutaneous involvement does not necessarily progress to other systemic manifestations. Colchicine treatment significantly reduced recurrence of aphthosis.

Conclusion: Mucocutaneous manifestations are pivotal in diagnosing Behçet's disease, showing significant variation and necessitating ongoing monitoring and individualized management.

Managing Arterial Aneurysms in Behçet's Disease: Insights from a Retrospective Cohort Study

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Introduction: Arterial Aneurysms are severe complications of Behcet disease, can rupture, leading to grave and often fatal outcomes. Effective management of aneurysms includes a range of recommendations depending on the patient's condition.

Method: This study is a retrospective, descriptive, analytical,monocentric and observational investigation, from January 2012 to January 2022.

Discussion: Among the 531 patients followed for Behçet's disease, The sex ratio was 5:1. HTA was present in 10.23%, diabetes 2.3%, alcohol 7.4%, and tobacco 10.6%. Arterial aneurysms accounted for 35% of all vascular involvement with venous thrombosis comprising 65%. clinically, pain was in 90%, skin erythema, and warmth in 60%. Ulcerations were observed in 5%, vascular bruit in 80%, and a pulsatile mass in 2% of aortic aneurysms. Claudication was present in 60%, but no patients exhibited ischemia, arterial thrombosis, stenosis, or occlusion. Arterial aneurysms were located in the pulmonary artery and its lobar branches (66%), femoral artery (7%), the aorta (6%), and the axillary and subclavian arteries and false aneurysms (5%). Most patients with vascular manifestations presented with anemia and elevated CRP (92.5%)and ECG abnormalities. Corticosteroids were given to 90%, followed immunosuppressants. Surgical treatment was performed in only 15 cases (1.5%), mainly for arterial aneurysms (aneurysmectomy and bypass) and the placement of cava filters. A favorable evolution was observed in 78% of patients, with 16.5% relapses. 4.7% dying from vascular causes, primarily pulmonary embolisms and aneurysm ruptures. Statistical analysis revealed significant associations between aneurysms and diabetes, posterior uveitis, as well as relapses and deaths ($p < 0.001$).

Conclusion: arteriel aneurysms in Behçet's disease are rare but serious. Glucocorticoids and immunosuppressants are crucial in limiting recurrences. Endovascular treatment with stent grafts is an alternative to open surgery, reducing recurrences.

Vascular Behçet's syndrome

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Introduction: Behçet's disease (BD) is a systemic auto-inflammatory vasculitis of unknown etiology, characterized by mucocutaneous manifestations, including recurrent oral and genital ulcers, ocular manifestations, and systemic vasculitis involving all types of vessels, regardless of size. The aim of our work is to evaluate clinical and therapeutic characteristics of vascular Behçet's in the department of Internal Medicine.

Method: We conducted a retrospective descriptive analysis of a cohort of patients with vascular BD evaluated between January 2004 to December 2023, according to the criteria of the International Study Group for BD, (ICBD) 2014. Patients with cerebral venous thrombosis were excluded

Discussion: Among 399 patients diagnosed with BD; 131 patients (32,8 %) with vascular Behçet's disease (BD) were included in the study. A male predominance was noted, with a sex ratio of 2.4. The mean age at diagnosis of the vascular involvement was 34.3 years. Venous thrombosis was reported in 108 patients (82.4%). The most frequent site of deep vein thrombosis was the lower extremities, occurring in 70 cases (53.4%), followed by the inferior vena cava, superior vena cava thrombosis and portal vein thrombosis in 17 (13%), 11 (8.4%) and 12 (9.2%) patients, respectively. Arterial involvement was reported in 28 patients (21.4%). The pulmonary artery was involved in 9 cases (6.9%), while extra-pulmonary arterial involvement was noted in 12 cases (9.2%). All patients with deep vein thrombosis were treated with colchicine, steroids, immunosuppressants, and anticoagulants, whereas those with arterial involvement received similar regimen excluding anticoagulation. Three cases warranted arterial embolization, while one patient underwent a pulmonary lobectomy.

Conclusion: Our study highlights the prevalence of vascular involvement in BD and demonstrating a male predominance. Consistent with previous studies, our findings indicate a higher prevalence of venous involvement compared to arterial involvement.

Digestive Manifestations of Behçet's Disease: A Case Study and Clinical Implications

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Introduction: Affecting individuals aged 20 to 40 in the Mediterranean region, Behçet's disease is a systemic vasculitis presenting a wide range of symptoms, including cutaneous- mucosal, vascular, ophthalmological, and rarely gastrointestinal manifestations.

Method: A retrospective observational study: 531 patients diagnosed with Behçet's disease. 21 exhibited gastrointestinal symptoms. The average age of these patients was 38 years, with a significant male predominance, comprising 19 men and 2 women.

Discussion: Our clinical case below emphasizes the importance of considering all symptoms, even seemingly insignificant ones, in individuals with Behçet's disease, as the consequences can be life-threatening. It involves a 45-year-old patient diagnosed with Behçet's disease. Her initial symptoms included bipolar ulcers, pseudofolliculitis, and deep vein thrombosis, successfully treated with colchicine and anticoagulants. However, over the course of three years, she began experiencing significant digestive issues such as diffuse abdominal pain, bloating, and transit disorders: diarrhea/constipation. These symptoms were accompanied by fatigue, unexplained weight loss, and loss of appetite. The progression was marked by the sudden onset of acute abdomen: peritonitis requiring immediate surgical intervention due to perforation. After the operation, blood tests showed increased CRP levels and leukocytosis, but indicated no malabsorption issues. As a result, the patient received corticosteroid therapy and cyclophosphamide remotely from her surgery.

Conclusion: To prevent serious complications such as peritonitis, it is crucial to conduct a comprehensive assessment, including colonoscopy, in the presence of possible digestive symptoms associated with Behçet's disease. Taking early measures can improve treatment outcomes and enhance overall patient quality of life.

Clinical practice guidelines for Behçet's Disease 2020 by the Japan Society for Behçet's Disease: Comparison with the other sets of guidelines and recommendations

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Introduction: The Japan Society for Behçet's Disease (JSBD)and Behçet's Disease Research Committee of Japan generated Clinical practice guidelines for Behçet's Disease in 2020. We here show the JSBD guidelines compared to currently used guidelines and recommendations in other countries.

Method: Recommendations in the JSBD guidelines were generated according to the Medical Information Network Distribution Service (Minds) and the GRADE system. The evidence and agreement levels determined the statements' strength for each clinical question (CQ). Recommended therapies for each BD symptom in the JSBD guidelines are compared with 2018 update EULAR recommendations, French recommendations, and UK guidelines for the management of BD.

Discussion: The JSBD guidelines consist of 150 CQs concerning the management of mucocutaneous, joint, ocular, gastrointestinal, vascular, and neurological symptoms. The JSBD guidelines recommend colchicine, topical and systemic glucocorticoids (GC), immunosuppressants, and anti-TNF antibodies as standard treatments for individual symptoms, like the other sets of guidelines and recommendations. In addition, the French and UK also suggest IL-6 inhibitors and IL-17 inhibitors as options for refractory cases. Major differences are found in ocular and neurological involvement treatments between the JSBD and the others. In the JSBD guidelines, the first line therapy for ocular lesions is colchicine which is not recommended in the others. For acute ocular attacks, topical GC administration is preferred over systemic administration, unlike the others. In the neurological involvement, the JSBD guidelines recommend GC for acute type and methotrexate for chronic progressive type as the first-line therapies. Anticoagulation is recommended for deep vein thrombosis except for patients having bleeding risk.

Conclusion: The JSBD guidelines optimize recommended therapies for Japanese BD patients. It is awaited that new treatments are established based on standardized efficacy and safety assessments in BD.

Female thrombosis in Behcet disease

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Introduction: Thrombosis is a common clinical manifestation during angiobehcet. The objective of this study is to describe the clinical and epidemiological aspects of vascular thrombosis in women followed for Behcet's disease.

Method: This is a retrospective, descriptive monocentric and analytical study spanning 20years from 2003 to 2022 including the records of patients followed for Behcet's disease associated with vascular thrombosis.

Discussion: We collected 136 cases of vascular thrombosis. Women were older than men with a mean female age of 44.5 years ± 12.7 compared to a male age of 39.9 ± 11.1 years and the overall 40.6 years ±11.4 years. Sex ratio was: 5.18 (114 H/22 F). Thrombosis revealed Behcet's disease in 6 women. Among the 127 cases of venous thrombosis, 1 woman/2 men (30%) presented with superficial venous thrombosis. 20 women/124 men (16%) had deep vein thrombosis (2 cases of thrombosis of the superior and inferior vena cava, 12 cases in the lower limb veins). Arterial thrombosis was present in 3F/28 men (11%). No woman presented with pulmonary artery thrombosis.. 21/22 Women presented severe forms of thrombosis i.e. 16% of the overall series (21F/133H). women had a tendency to present thrombosis at a single site (18.75% (14F/75H) and 14F/ 106 H (13.2%) cases of proximal thrombosis. Univariate and multivariate analyses did not objectify statistical links between female sex and risk factors (age, hypertension, diabetes and smoking), and between the clinical- anatomical presentations of thrombosis and the female sex (p>0.05).

Conclusion: Women followed for Behcet's disease tend to have less frequent, less severe vascular thrombosis and a single thrombosis site. Female sex does not show a statistically significant relationship with the different presentations of thrombosis in this study.

Unusual localization of Behcet's disease revealed by a false aneurysm

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Introduction: Behçet's disease is a multisystemic chronic inflammatory disease. Vascular involvement can include both arteries and veins, with lesions ranging from arterial occlusions and aneurysms to superficial thrombophlebitis. The majority of patient deaths due to vasculo-Behcet's disease are related to rupture of the aneurysms. We report an exceptional case of Behçet's disease revealed by false aneurysm of right subclavian artery.

Method: Case1 A 47-years-old patient, was admitted for a swelling in the right supraclavicular fossa. His medical history was notable for a recurrent orogenital aphthosis. Physical examination showed an hemodynamically stable patient with an expansive and pulsatile mass measuring 5cm located above the collarbone. CTA and arteriography revealed a false aneurysm of the right subclavian artery. The diagnosis of vasculo Behcet disease was retained. Before the operation, methylprednisolone was injected intravenously 3 days in a row associated with cyclophosphamide. The patient underwent surgery. We performed open surgery with aneurysm flattening, the damaged arterial segment was resected with restoration of vascular continuity

Discussion: Behcet disease has a peculiar geographic distribution. It is more common in Mediterranean regions, where an association with HLA B51 has been proved. Vascular involvement of behcet's disease is characterized by arterial and venous occlusions, and arterial aneurysms. The frequency of aneurysmal lesions is very rare. Various treatment modalities in artery aneurysm include immunosuppressive therapy, surgery, and endovascular surgery. Cyclophosphamide azathioprine and glucocorticoids constitute the basic treatment of aneurysms on Behcet. TNF a cures have been reported in refractory cases or with severe aneurysmal lesions

Conclusion: Surgery combined with immunosuppressive treatment in patients with vasculo Behcet disease appears to be a promising and effective management option. A longer follow-up is required to allow confidence of lasting success

Impact of Psychiatric Complications in Behçet's Disease: Case Study

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Introduction: Behçet's disease, a Mediterranean vasculitis, manifests with oral and genital ulcers and induces psychological disorders, profoundly affecting patients' lives, as detailed in two cases emphasizing the impact on patients and their families' well-being.

Method: files from hospitalizations.

Discussion: Observation 1: A 57-year-old man initially diagnosed with Behçet's disease experienced recurrent oral and genital ulcers, pseudo-folliculitis, and deep vein thrombosis, well managed with corticosteroids, Imuran, and anticoagulants. Subsequently, memory disturbances worsened post-stroke, leading to severe disability and major depression, necessitating antidepressant treatment. Observation 2: A 47-year-old patient with Behçet's disease, under neurology care since 2015, presented with bipolar ulcers and neurological symptoms, including intracranial hypertension and pyramidal syndrome. Treatment with cyclophosphamide, corticosteroids, and colchicine led to pseudobulbar syndrome, causing uncontrollable emotional outbursts. This significantly affected the patient's ability to express emotions, prompting initiation of antidepressant therapy to alleviate symptoms. Observation 3 A patient, initially managed in internal medicine at 27 years old, faced severe ophthalmological complications despite residing over 200 km from the hospital. Treatment included corticosteroids, 12 cycles of cyclophosphamide, and Imuran. Delayed access to anti-TNF therapy due to social coverage issues led to the patient's death after ten years of treatment marked by vision deterioration and severe complications. Severe depression, suicidal ideation, and frontal syndrome necessitated antidepressants, social support, and corticosteroid reduction.

Conclusion: This manuscript highlights significant cases of Behçet's disease, emphasizing its vascular nature and the substantial impact of both the disease and its treatment on patients' lives, underlining the importance of researching psychiatric symptoms for comprehensive patient care.

Behçet's Disease: Clinical Profile and Mortality Risk Factors

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Introduction: Behçet's disease is a systemic vasculitis affecting various-sized blood vessels, leading to recurrent oral and genital ulcers and uveitis. While historically prevalent along the Silk Road, its incidence is increasing in North America and Europe. The disease exhibits considerable variability in symptoms, progression, and treatment response, with potentially fatal complications in some cases. Our Internal Medicine and Clinical Hematology Department in Rabat documents cases where Behçet's disease patients experienced adverse outcomes, including death.

Method: This is a retrospective descriptive, analytical, and observational monocentric study conducted at the Internal Medicine Department of CHU Rabat over 10 years, from January 2012 to January 2022. We collected 531 patient records of those diagnosed with Behçet's disease.

Discussion: In our study, 14 Behçet's disease patients died, with a mean age of 40 years and a male predominance (13 males, 1 female). All had bipolar aphthosis, 11 had posterior uveitis, and 13 had vascular involvement, including carotid artery lesions, femoral vein thrombosis, and 6 with pulmonary artery aneurysms. Mortality was statistically linked to arterial vascular involvement, notably pulmonary artery aneurysms ($p = 0.042$) and carotid artery aneurysms ($p = 0.001$). Mortality in Behçet's disease patients mainly stems from arterial vascular complications, sometimes exacerbated by treatment, leading to severe infections. However, precise excess mortality calculation is hindered by the lack of comprehensive data.

Conclusion: Behçet's vasculitis is a severe disease with potentially fatal consequences, leading to patient mortality.

Peripheral arteriel involvement in Behçet's Disease: Multi-center retrospective case series

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Introduction: In Behçet's disease(BD), arterial involvement represents a significant clinical challenge due to its potential for serious complications. In this study, we aimed to investigate the characteristic features, affected vessels, types of lesions, treatment modalities, and mortality of patients with arterial involvement in BD.

Method: Fifty-four BD patients were included in this retrospective cohort study. The data is collected from medical records.

Discussion: Forty-nine patients(90,7%) were male. The median follow-up duration was 4(2-12) years. The most commonly affected vessels were the aorta(n=19,35,1%) and femoral artery(n=17,31,4%), respectively. Aneurysm (72,2%) and thrombosis (18,5%) were the predominant vascular lesions. Ten(18,5%) patients experienced arterial involvement under immunosuppressive(IS) treatment (azathioprine,n=9; infliximab,n=1). Out of 54 patients, 10(18,5%) had cardiac involvement (coronary artery aneurysms,n=7; intracardiac thrombi,n=3). The most commonly used ISs were cyclophosphamide (n=37,68,5%), azathioprine (n=34,63%), and anti-TNF drugs (adalimumab,n=3; infliximab,n=11;25,9%). Surgical and interventional procedures were performed on 26(48,1%) patients, with 19(73%) of these procedures being successful. During follow-up, a second arterial event occurred in 8 patients(14,8%), and a third arterial event occurred in 1(1,8%) patient. Despite treatment, 9(16,6%) patients still had active disease on their latest visits (evidenced by elevated acute phase reactants and the vascular events detected through imaging). Mortality was observed in 5(9,2%) patients (intracardiac thrombi and pulmonary artery aneurysm,n=1; acute coronary syndrome,n=2; infection,n=1; alveolar hemorrhage,n=1).

Conclusion: Arteriel involvement in BD mostly presents with aneurysm formation in especially young male patients. About half of the patients need surgical intervention in addition to IS treatments during follow up. Mortality was found 9,2% mostly in patients with both arteriel and cardiac involvement.

Aorta (n=19)	35,1 %
Femoral (n=17)	21,4 %
Coronary (n=7)	12,9 %
Popliteal (n=6)	11,1 %
SMA (n=3)	5,5 %
SCA (n=3)	5,5 %
CCA (n=1)	1,8 %
ICA (n=1)	1,8 %
Brachiocephalic (n=2)	3,7 %
Gastric and splenic (n=1)	1,8 %
Renal (n=3)	5,5 %
Iliac (n=2)	3,7 %
Tibial (n=3)	5,5 %
Dorsalis pedis (n=1)	1,8 %
Axillary (n=1)	1,8 %

Table 1: Distribution of arteriel involvement.

CCA: common carotid artery ICA: internal carotid artery SCA: subclavian artery SMA: superior mesenteric artery

The role of surgery in Behçet's disease.

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Introduction: Behçet's disease, primarily affecting young adults aged 20 to 40, is a vasculitis prevalent along the Silk Road. Among its severe complications are arterial aneurysms, prone to rupture with grave outcomes. Effective management depends on disease activity and aneurysm stage. This manuscript shares our department's experience with 532 patients and their surgical interventions.

Method: This study is a retrospective, descriptive, analytical, and observational investigation conducted in the internal medicine department in Rabat over ten years, from January 2012 to January 2022. We reviewed 531 patient records of individuals treated for Behçet's disease. Statistical analysis was performed using JAMOVI software version 1.3, utilizing chi-square, Fisher's exact, and Student's t-tests.

Discussion: Among the 531 patients with Behçet's disease, 48% showed vascular involvement, predominantly in males. Clinical symptoms included pain (90%), skin erythema (60%), and aneurysms primarily detected by vascular bruit (80%). Arterial aneurysms, comprising 35% of cases, were most commonly located in the pulmonary artery (66%), with favorable outcomes observed in 78% of patients. Surgery was required in only 15 cases (1.5%), mainly for arterial aneurysms, highlighting its crucial role in severe cases. However, significant associations were noted between surgically managed aneurysms and diabetes, which likely weakened vessels, as well as severe ophthalmologic involvement, relapses, and deaths. aneurysms in Behçet's disease are rare but serious. The EULAR recommends immunosuppressive agents and surgery, though some experts advise against surgery during active phases. Endovascular treatment with stent grafts is an alternative to open surgery, reducing recurrences.

Conclusion: Arterial aneurysms in Behçet's disease are effectively managed through a blend of immunosuppressants, corticosteroids, and endovascular procedures, enhancing prognosis and necessitating a multidisciplinary approach for optimal outcomes.

Predictors of Severity in Ocular Involvement of Behcet's Disease: A Monocentric Study of 92 Patients (163 Eyes)

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Introduction: To date, there is insufficient data on general predictors for vision-threatening disease in patients with Behcet's disease. Our study aimed to identify and highlight systemic findings commonly associated with severe ocular Behcet's disease.

Method: This study included 92 patients with ocular involvement of BD seen from January 2010 to December 2023 in the internal medicine department of the university hospital Mohammed VI of Marrakech. Patients were further sub-grouped as having a severe form of the disease or not. Patients with a best-corrected visual acuity less than 1/10, as well as those with documented pan-uveitis, retinal vasculitis, chorioretinitis, and/or papillitis or a combination of these findings, were classified as a severe form.

Discussion: Ninety-two patients (163 eyes) were enrolled, representing 23% of all patients with BD. The male/female ratio was 2,3. The most common ocular findings were retinal vasculitis (45,4%) and intermediate uveitis (39,9%). Mucocutaneous involvement was present in all patients, with bipolar aphthosis in 87 (94.6%), isolated oral involvement in 5 (5.4%), pseudo-folliculitis in 37 (40.2%), and erythema nodosum in 15 patients (16.3%). Thirty-two patients (34.8%) had joint involvement, 11 patients (11.9%) had vascular involvement. Neurological involvement was present in two patients (1%), and digestive involvement in one patient. No correlation was found between systemic manifestations and the severity of ocular involvement in Behçet's disease in our series. However an above average age (more than 34 years) was correlated with a severe ocular involvement in our series.

Conclusion: If the severity of ocular involvement in Behcet's disease has certain predictors, identifying them in an early stage can help in planning the frequency of follow-up visits and determining the aggressiveness of treatment.

Ocular Behcet's Disease: Evaluating Visual Prognosis and Influencing Factors in Patients: A Monocentric Study of 92 Patients (163 Eyes)

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Introduction: Behçet's disease (BD) is one of the most frequent causes of severe visual impairment. Understanding the visual prognosis and identifying factors that contribute to the development of severe and irreversible visual loss are crucial for optimizing management and improving outcomes. This study aims to investigate these factors.

Method: This is a retrospective study of patients with ocular involvement of BD. All patients met the International Criteria for Behçet's Disease 2014. Uveitis was classified according to the International Uveitis Study Group.

Discussion: Ninety-two patients (163 eyes) were enrolled. The most common ocular findings were retinal vasculitis (45,4%) and intermediate uveitis (39,9%). Initial visual acuity was "counting fingers" in 36 eyes, 1/10 in 21 eyes, and "hand motion" in 16 eyes. All patients were taking colchicine and steroids. The immunosuppressive drugs prescribed were cyclophosphamide, azathioprine, tacrolimus and methotrexate. Biologic agents prescribed included TNF inhibitors and tocilizumab. On follow-up, visual acuity exceeded 3/10 in 86 eyes, "counting fingers" in 17 eyes and 1/10 in 14 eyes. Complete recovery was observed in 52 eyes. The most important complications were synechiae, blindness and macular edema in 22 eyes. Key predictors of visual loss included male gender, an age superior to 34 years, presence of posterior uveitis, and delayed initiation of immunosuppressive therapy. Patients receiving early and aggressive treatment, including biologic agents, had a significantly better visual prognosis compared to those on conventional therapies alone.

Conclusion: Our findings support the use of early immunosuppressive and biologic therapies in patients with severe ocular BD to improve long-term visual prognosis and quality of life.

Specificities of Angio-Behçet: Experience of the Internal Medicine Department at Mohamed V Military Hospital, Rabat

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Introduction: Vascular involvement in Behçet's disease, known as Angio-Behçet, are the most frequently encountered manifestations. They can impact various types of vessels. Our research seeks to explore the epidemiological, clinical, and therapeutic aspects of Angio-Behçet

Method: This is a monocentric, descriptive, and retrospective study conducted within the Internal Medicine Department "B" of the Mohammed V Military Hospital in Rabat, spanning a 20-year period (2003 – 2023), collecting the medical records of patients treated for Behçet's disease. All patients met the international classification criteria for Behçet's disease of 2013

Discussion: One hundred seventy-seven cases were identified. The prevalence of vascular involvement was 35.6% (63 patients). There were 55 men and 8 women (male-to- female ratio = 6.8). The average age at the time of vascular involvement was 37 years. Vascular involvement revealed Behçet's disease in 18 patients. Venous thromboses were the most common manifestation, observed in 56 out of 63 patients (88.9%). These included deep vein thromboses of the lower limbs in 82.1%, superior vena cava thrombosis (12.5%), superficial vein thromboses (16.1%), Budd-Chiari syndrome (5.3%), and upper limb vein thrombosis (1.8%). Arterial involvement was found in 16 out of 63 patients (25.4%) who were male. Nine patients with arterial involvement had a history of thrombophlebitis in the same territory and presented cardiovascular risk factors. There were 6 thromboses and 11 aneurysms. Corticosteroid therapy was necessary in all cases in combination with other therapies: anticoagulants, colchicine, and immunosuppressants. Among patients with aneurysmal lesions, six underwent surgery. The outcome was favourable for the majority of patients. Two patients had pulmonary embolism and two others had postoperative complications. There were 2 reported deaths in our series

Conclusion: Our studied series underscores notable epidemiological, clinical, and treatment- related features identified in Angio-Behçet

Digestive involvement in Behçet disease in Behçet disease

Bott Zineb, Médecine interne
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Introduction: Behçet 's disease (BD) is a complex systemic vasculitis primarily manifesting as vascular,mucocutaneous, and ocular symptoms. However,digestive involvement is rare but can significantly impact patient.prognosis, often mimicking inflammatory bowel diseases (IBD).

Method: This retrospective study was conducted at Marrakech University Hospital's Department of Internal Medicine from January 2010 to December 2023. It included 399 patients who met the 2014 International Criteria for Behçet's Disease.

Discussion: Among the 399 patients 5 presented with digestive involvement (1.25%). The mean age was 42 years; the sex ratio was 1.5, with a male predominance. The main symptoms were abdominal.pain,diarrhea and melena in 3 patients, rectal bleeding in 2 patients, Additional workup showed peptic ulcer in one patient, colitis in 3 patients, intestinal ulcers and peritonitis in 2 patients, intestinal perforation and ulcerated esophagitis in one patient. Four patients experienced favorable outcomes, while one was lost to follow-up. Treatment regimens included colchicine and high-dose steroids for all patients. Additionally, azathioprine was administered to two patients, and

three received sulfasalazine.

Conclusion: Digestive symptoms in BD, while rare, pose a diagnostic challenge due to overlap with IBD symptoms. Accurate diagnosis relies on thorough histological examination and diagnostic scoring. Awareness of gastrointestinal manifestations in BD is crucial for better recognition and management.

Validation of the International Study Group Criteria (ISG) and the International Criteria (ICBD) for Diagnosing Adamantiades-Behçet Disease in German and Turkish Patients in Germany

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Introduction: The challenge of optimizing sensitivity and specificity for diagnostic and classification cirteria in rare diseases like Adamantiades-Behçet Disease (ABD) is still persisting and under debate. This study aims to validate the diagnostic sensitivity of the International Study Group Criteria (ISG, 1990) and the International Criteria (ICBD, 2014) in diagnosing ABD in both German and Turkish patient populations living in Germany, utilizing data from the German Registry of ABD. Both the ISG and the ICBD criteria were employed within these groups to assess diagnostic sensitivity.

Method: A retrospective cohort investigation utilized data from the German Registry of ABD, categorizing patients into three distinct groups: the entire cohort, all patients of German descent, and all patients of Turkish descent in the Registry.

Discussion: The ISG criteria exhibited a sensitivity of 72.2% for the entire cohort, 68.1% for German patients, and 75.6% for Turkish patients. Conversely, the ICBD criteria demonstrated superior sensitivity across all categories: 89.3% overall, 89.1% for German patients, and 89.8% for Turkish patients. Recurrent oral aphthosis emerged as the predominant symptom (97.6%), succeeded by cutaneous manifestations (72.3%), genital ulcerations (63.6%), and ocular lesions (54.3%).

Conclusion: The ICBD criteria appear to offer significantly higher diagnostic sensitivity compared to the ISG criteria among ABD patients residing in Germany. The accurate diagnosis of ABD remains a complex, interdisciplinary challenge, highlighting the importance of confirming the diagnosis by specialists in the field. At present, given the diagnostic uncertainty when ICBD or ISG criteria are not met, it is beneficial to consider other criteria for classifying and diagnosing challenging cases. These include those proposed by Hewitt (1969), Mason and Barnes (1969), O'Duffy (1974), Cheng and Zhang (1980), the Japanese criteria (1987), Dilsen (2000), the Korean criteria (2003), and others.

Eye And Behcet's Disease: A Monocentric Study of 92 Patients (163 Eyes)

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Introduction: Ocular manifestations of Behçet's disease (BD) have a prevalence varying between 50–70% and can lead to sight-threatening complications. The aim of our study was to describe the clinical features and outcome of ocular involvement in BD in our department.

Method: This is a retrospective study of patients with ocular involvement of BD, seen from January 2010 to December 2023 in the internal medicine department of the university hospital Mohammed VI of Marrakech. All patients met the International Criteria for Behçet's Disease 2014. Uveitis was classified according to the International Uveitis Study Group.

Discussion: Ninety-two patients (163 eyes) were enrolled, representing 23% of all patients with BD. The male/female ratio was 2,3. Ophthalmic manifestations were the initial presentation of the disease in 63% of patients. The most common ocular findings were retinal vasculitis (45,4%) and intermediate uveitis (39,9%). All patients were on colchicine and steroids. The immunosuppressive drugs prescribed were cyclophosphamide, azathioprine, tacrolimus and methotrexate. Biologic agents prescribed included TNF inhibitors and tocilizumab. The most important complications were synechiae (25,8%), blindness (16,6%) and macular edema (13,5%).

Conclusion: Ocular involvement in BD is a serious condition, and can lead to blindness. A close collaboration between the ophthalmologist and internist is required to improve the prognosis.

Digestive venous thrombosis in Behçet's disease: observations from 5 cases

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Introduction: Venous thrombosis of unusual sites is common in patients with Behçet's disease.The occurrence of digestive venous thrombosis is serious but exceptional.

Method: We collected 5 cases of Behçet's disease with at least one episode of digestive venous thrombosis documented by imaging, diagnosed in the internal medicine department A of the Mohamed V military hospital in Rabat.

Discussion: The mean age of the patients was 43.8 years, with four women and one man. The mode of onset of digestive thrombosis was acute in three patients. The territory affected concerned the supra hepatic veins in two cases, the superior mesenteric vein in one case and the cava vein in two cases. Thrombophilia tests were negative in all patients. Therapeutically, all our patients were treated with curative anticoagulants combined with immunosuppressive therapy.

Conclusion: Venous thrombosis is the main cause of vascular damage in angiobehçet and is primarily related to inflammatory phenomena. Digestive vascular involvement remains rare, dominated by Budd Chiari Syndrome and thrombosis of the cava vein, as highlighted by a French study which found that 2.4% of patients suffering from Behçet's disease had Budd Chiari Syndrome. Immunosuppressive treatment is the cornerstone of management, alongside anticoagulant therapy to limit thrombus extension.

A Study of Novel Inflammatory Markers in Behcet's Disease with Ocular Involvement

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Introduction: Only few studies have investigated the use of systemic inflammatory markers to reflect disease activity of Behçet's disease (BD) such as-reactive protein (CRP), CRP/albumin ratio (CAR), neutrophil to lymphocyte ratio (NLR), platelets to lymphocytes ratio (PLR), lymphocytes to monocytes ratio (LMR), and mean platelet volume (MPV). The purpose of our study was to explore these markers as indicators of disease activity in BD with ocular involvement.

Method: This study included 92 patients with ocular involvement of BD seen from January 2010 to December 2023 in the internal medicine department of the university hospital Mohammed VI of Marrakech. Laboratory tests were conducted to measure complete blood count, CRP, CAR, NLR, PLR, LMR, and MPV during the active phase and after resolution. The activity of ocular involvement was determined by BD ocular attack score 24 (BOS24). The results measured during the two phases were assessed and compared.

Discussion: Ninety-two patients (163 eyes) were enrolled with a male/female ratio of 2,3. The most common ocular findings were retinal vasculitis (45,4%) and intermediate uveitis (39,9%). Statistical analysis showed significant variations between active and inactive phases of the disease. MPV and NAR demonstrated highly significant changes with p-values less than 0.001. Similarly, LMR and CRP levels showed significant differences (p=0.023 and p=0.007, respectively). The CAR also had a statistically significant variation (p=0.014). Conversely, PLR and NLR did not show a significant change (p=0.910 and p=0,222, respectively).

Conclusion: The blood indices CRP, NAR, CAR, NLR, PLR, LMR, and MPV are potential inflammatory markers that can be used to evaluate disease activity in patients with ocular involvement of BD.

Depression is Still Frequent Among Male Patients with Behçet's Syndrome in the Biologic Era: Cross-Sectional Study

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Introduction: Behçet's syndrome (BS) is a chronic multisystem inflammatory disorder of unknown etiopathogenesis, and the incidence of depression is increased in BS patients. Also these patients usually have poor health-related quality of life (HRQoL). This study aimed to investigate the predictors of depression in male BS patients by adjusting for established risk factors for depression.

Method: Each patient fulfilled the Beck depression inventory (BDI), Short form (SF)-36, and hospital anxiety and depression (HADS) scales, and disease severity and disease activity were evaluated with the Krause score and BDCAF, respectively.

Discussion: The present study included 185 male BS patients (mean age: 39.6±9.2). The median disease duration was 13 years (min-max: 0.1-36), and 107 (58%) patients had ocular, 131 (70%) vascular, and 43 (23%) neurological involvement. Ninety (49%) patients were treated with biologics and 33 (18%) with pegylated-interferon-α, whereas the remaining were using cDMARDs. Fifty-four (29%) patients had depression, and 18 (10%) had suicidal ideation. The overall HRQoL (OR: 0.928) and the use of biologic agents (OR: 3.982) were identified as independent predictors of depression. Among the biologics, depression was more prominent with infliximab, possibly related to periodic intravenous infusions. Also, overall HRQoL (OR: 0.949) and duration of BS (OR: 0.877) were significantly associated with suicidal ideation. BDI was strongly and negatively correlated with the physical and mental components of SF-36. On the other hand, BDI was positively correlated with disease activity and disease severity. While Krause scores were substantially correlated with the mental component of SF- 36, there was no significant correlation between Krause scores and SF-36 physical component.

Conclusion: Just as we treat the physical disease in BS, patients' mental problems should also be addressed in order to cope with depression. Also, caution should be exercised, especially in BS patients receiving biologic treatment.

Clinical and Epidemiological Characteristics of Adamantiades-Behçet Disease in a Moroccan Cohort

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Introduction: This study aims to analyze the demographic data, frequency of clinical manifestations, familial occurrence, and potential prognostic signs in Moroccan patients diagnosed by the end of 2023.

Method: A total of 111 patients with Adamantiades-Behçet Disease (ABD), permanently residing in Morocco, were documented.

Discussion: The cohort consisted of 72 males and 39 females. 109 patients were of Moroccan descent, one was from Libya, and one had unknown origin. Family history was positive in 35.1%. Oral aphthae were present in 99.1% and were the most common initial symptom in 87.4%. Cutaneous lesions were prevalent overall in 90.1%, with folliculitis in 87.4%, erythema nodosum in 25.2%, and superficial thrombophlebitis in 11.7%. Ocular manifestations, incl. hypopyon and retinitis, were notably frequent affecting 66.7%. As second most common initial manifestations, retinitis occurred in 3.6% and hypopyon-iritis in 2.7%. Genital ulcers were observed in 59.5%, being the most common 2nd symptom in the course (32.4%). Other 2nd symptoms during the disease course included retinitis (12.6%), hypopyon-iritis (7.2%), and cutaneous manifestations such as folliculitis (13.5%) and erythema nodosum (7.2%). Arthritis was confirmed in 29.7%, though joint pain was reported in 75.7%. Neurological manifestations were documented in 73%, most frequently as headache, though 24.3% were confirmed as CNS manifestations (e.g., via EEG or as meningoencephalitis). Other manifestations included gastrointestinal in 19.8%, vascular manifestations in 27%, epididymitis in 15.3%, cardiac in 11.7%, pulmonary in 6.3% and renal involvement in 2.7%. Pathergy test was performed on 87 patients and was positive in 67.6% of the cohort (86.2% of those tested). Most severe complications included blindness in 10.8% (n=12), meningoencephalitis in 9.9%, lung bleeding in 1.8% (n=2), and 2 deaths (1.8%).

Conclusion: ABD in the Moroccan cohort exhibits a high prevalence of mucocutaneous and ocular manifestations, with significant familial occurrence.

Clinical and Epidemiological Characteristics of Adamantiades-Behçet Disease: A Comparative Study of German and Moroccan Patients

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Introduction: This study compares the clinical features and epidemiology of Adamantiades-Behçet Disease (ABD) in German and Moroccan cohorts. We analyzed demographic data, the frequency of clinical manifestations, familial occurrence, and potential prognostic indicators.

Method: The German cohort included 355 patients of German descent, with data collected via standardized forms from 32 clinics and practices. The Moroccan cohort consisted of 111 patients, documented by the end of 2023.

Discussion: In the German cohort, oral aphthae (OA) were the most common symptom (98.1%), followed by genital ulcers (GU, 64.8%) and cutaneous lesions (CL, 64.4%). Ocular manifestations were observed in 39.1% of patients. Arthritis (AR) was reported in 56.1%, and neurological involvement in 20.8%, with 11.9% having CNS involvement. Familial occurrence (FO) was low in 3.7%. In the Moroccan cohort, OA were the most common symptom (99.1%), followed by CL (90.1%), especially folliculitis (87.4%). Severe ocular manifestations were more frequent (66.7%), including hypopyon-iritis and retinitis. GU were observed in 59.5%. Typical AR was confirmed in 29.7%, though joint pain was reported in 75.7%. NI was suspected in 73%, primarily as headache; CNS involvement was confirmed in 24.3%.

Conclusion: The comparison of German and Moroccan ABD cohorts shows both similarities and differences in disease presentation. OA were nearly always present (98.1% in Germans, 99.1% in Moroccans). Moroccans had higher ocular manifestations (66.7% vs. 39.1%), FO (35.1% vs. 3.7%), CNS involvement (24.3% vs. 11.9%), and CL (90.1% vs. 64.4%), with statistic significance. These findings highlight the importance of ethnic and genetic factors in ABD management and suggest tailored treatment strategies based on patient demographics.

Splanchic Venous Thrombosis in Behcet's Disease: A Monocentric Study of 22 Patients

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Introduction: Splanchnic venous thrombosis is an unusual manifestation of Behçet's disease. The purpose of our study was to explore the clinical features and outcome of these patients.

Method: This is a retrospective study carried out in the internal medicine department of the university hospital Mohammed VI of Marrakech between 2010 and 2023. All patients met the International Criteria for Behçet's Disease 2014.

Discussion: Twenty-two cases were identified (5,5% of all behcet's patients). Among all etiologies considered, splanchnic vein thrombosis occurred in 42.3% of patients. Splanchnic venous thrombosis was inaugural in 71% of cases.Mucosal and cutaneous involvement was present in all patients. Articular involvement was present in 6 patients. Six patients had ocular involvement. One patient had renal involvement.Thrombosis of other vascular sites besides splanchnic thrombosis, was present in 13 patients. Splanchnic venous thrombosis was acute in 6 patients and chronic in 16 patients.Biological inflammatory syndrome was present in 59% of cases, 50% of patients had biological cholestasis, 31.8% had cytolysis, and 13.6% had low prothrombin time. Budd chiari syndrome was seen in 68% of cases, followed by superior mesenteric vein thrombosis (27,1%). All patients were on colchicine and steroids. Fifteen patients were on anticoagulation. Eighteen patients received an immunosuppressive agent (cyclophosphamide and azathioprine).Follow-up imaging was performed in 9 patients, and showed recanalization in 7 patients. Three patients died.

Conclusion: The prognosis of this patients may be favorable with early medical interventions, including steroids and immunosuppressive therapy.

Management of Severe Adamantiades-Behçet's Disease with Hughes-Stovin Syndrome, Cardiac Thromboses, and HIV Infection using Pegylated Interferon Alpha- 2a: A Case Report

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Introduction: We present a case of a 31-year-old German female of Rwandan descent with Adamantiades-Behçet's disease (ABD) complicated by Hughes-Stovin syndrome (HSS), cardiac thromboses, and concurrent HIV infection (since birth).

Method: We highlight the therapeutic challenges and successful management with pegylated interferon alpha-2a (Pegasys®).

Discussion: Clinical History: The patient was diagnosed with ABD in 2019 following deep vein thrombosis in the pelvis. She had multiple recurrent oral and genital ulcers, erythema nodosum, and superficial thrombophlebitis. Cardiac involvement included severe tricuspid insufficiency and polyserositis with pleural and pericardial effusions. In March 2019, the patient developed HSS with cardiac thromboses, leading to a biologic tricuspid valve replacement. Additional complications included a history of tuberculosis (1997) and heterozygous MTHFR mutations, further increasing her thrombotic risk. The patient underwent cataract surgery in November and December 2023. Previous and Current Treatment: Multiple immunomodulatory medications were ineffective or poorly tolerated, including methotrexate, cyclophosphamide, cyclosporine, azathioprine, colchicine, apremilast, anakinra, certolizumab, and infliximab. Peginterferon α-2a resulted in significant clinical improvement, with a reduction in disease activity and stabilization of symptoms. The initiation dose of 180 micrograms (µg) per week was reduced to 135 µg per week due to tolerability reasons. The patient is also on ongoing antiretroviral therapy for HIV. Discussion: The efficacy of Peginterferon α-2a in stabilizing ABD, suggests its therapeutic potential, especially in refractory cases with vascular involvement. Combination of Peginterferon α-2a with antiretroviral therapy for HIV was well tolerated. The postoperative courses under ongoing treatment with interferon alpha-2a were uneventful.

Conclusion: This case illustrates the complexity of managing ABD, particularly in the presence of HIV and other comorbidities.

Insights into Adamantiades-Behçet Disease in Germany: A Focus on Prevalence, Treatment Patterns, and Clinical Characteristics

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Introduction: In many European countries Adamantiades-Behçet Disease (ABD) poses diagnostic and therapeutic challenges due to its rarity and varied clinical presentations. This study aims to analyze ABD in Germany, focusing on prevalence, incidence, clinical manifestations, and treatment.

Method: Data from the German Registry of ABD (>900 patients) were compared with preliminary data from the Institute for Applied Health Research Berlin database (see Ref.). Prevalence, incidence, and treatment patterns were evaluated.

Discussion: ABD typically manifests in the third decade, with oral aphthae as the primary symptom. Patients of Turkish descent showed higher rates of positive familial history and uveitis prevalence. HLA-B5 positivity correlated with certain clinical manifestations, including uveitis and gastrointestinal involvement. A cohort study indicated a steady rise in ABD prevalence (estimated prevalence in Germany approximately 4.2: 100,000). Prednisolone, colchicine, and azathioprine were common treatments, with 15% receiving combination therapy.

Conclusion: In Germany, recent cohort studies have indicated a notable rise in ABD prevalence, aligning with the trend in our registry. The estimated prevalence of approximately 4.2 cases per 100,000 underscores its growing recognition in the healthcare system Source database: German Registry of Adamantiades-Behçet Disease Reference: Zouboulis, C.C., et al. (2021). Prevalence and Incidence of Adamantiades-Behçet's Disease: An Epidemiological Study from Germany. In: German Rheumatology Congress 2021, 49th Congress of the German Society for Rheumatology (DGRh), September 15th - 18th, 2021, virtual. [URL: https://www.egms.de/static/en/meetings/dgrh2021/21dgrh057.shtml]

Behcet's syndrome comorbid with bipolar disorder: A case report

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Introduction: Behcet's Syndrome is a chronic inflammatory disorder of unknown etiology, characterized by aphthous lesions and recurrent ulceration of the mouth, genitals and uveitis.The central nervous system is involved in about 20% of cases. Only few reports deal with affective symptoms associated with Behcet's syndrome.

Method: We report the case of a 19-year-old young man with Angio-Behcet's presenting with a psychotic and manic crisis. He developed Behçet's syndrome at the age of 14, with cerebral venous sinus thrombosis, oral aphthosis and genital aphthosis.

Discussion: At the age of 19, he presented to our department in a depressive episode with anxiety, asthenia with non-restorative hypersomnia, with suicidal ideation, associated with psychomotor agitation, talkative behavior. We referred him to the psychiatric ward, where he was diagnosed as bipolar II disorder with DSM-IV. Psychiatric examination revealed decreased psychomotor activity, hypomanic affect, quantity and speed of affect expression.

Conclusion: There are a few reports of bipolar disorder as an entity related to Behcet's syndrome.

An angio-behçet in an unusual location

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Introduction: Angio-Behçet refers to all vascular manifestations of Behçet's disease (BD), whether arterial or venous; it is one of the serious manifestations that could jeopardize the patients' prognosis. We report a case of angio-Behçet revealed by a superior vena cava syndrome (SVC)

Method: A 43-year-old, with no medical history, particularly no history of oral or genital aphthosis, who presented for management of SVC syndrome. Clinical examination revealed facial swelling, distended jugular veins, thoracic and abdominal collateral venous flow. Paraclinical assessment revealed inflammatory syndrome. The CT scan revealed a pulmonary embolism, thrombosis of the SVC, right internal jugular vein, and the azygos vein arch, with significant mediastinal collateral flow, Budd-Chiari syndrome, right portal vein thrombosis and thrombosis of the right and left iliac veins. The urine testing for proteinuria, serum albumin, thrombophilia testing, ANCA, APS, PNH clone search, JAK2 mutation, and bone marrow biopsy, all of which were négative. We started the patient on anticoagulants. Follow-up was marked by the appearance of recurrent oral and genital aphthosis, leaving genital scars that allowed us to retrospectively establish the diagnosis of BD, with HLA B 51 positive. The patient received azathioprine and corticosteroids. The evolution was favorable with regression of all clinical signs

Discussion: SVC thrombosis is a rare but serious manifestation of BD; its association with Budd- Chiari syndrome makes the prognosis even darker. The treatment objective is to quickly suppress inflammatory exacerbations and prevent recurrences to prevent irreversible organ damage. Treatment involves corticosteroids, immunosuppressants, as well as biologic therapy that should be maintained for an extended duration

Conclusion: Our case illustrates the need to consider BD in the etiological assessment of thromboembolic disease even in the absence of typical oral and genital aphthosis

Neuro-Behçet: 4 pseudo tumoral forms

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Introduction: Behçet's disease is a systemic disease that can affect all organs and progresses in outbreaks interspersed with remissions.The pseudo-tumor forms represent a rare entity in the neuro-Behçet leading to confusion with brain tumors

Method: Case1:18-year-old patient has consulted for intracranial hypertension associated with left hemiparesis.ARM has showed encephalitis with suspicion of a low-grade infiltrative right capsulothalamic lesion process.The management has consisted of the administration of 3boli MP and cyclophosphamide.Case2:the patient has presented right hemicranial headaches with left hemiparesis.ARM has revealed capsulothalamic and right mesencephalic lesions. The clinical evolution after medical treatment has been spectacular and the control imaging showed a clear regression of the right thalamic and mesencephalic involvement.Case3:The patient has presented a sudden onset of right temporal headaches with left hemiparesis.ARM has showed a pseudotumoral image evoking a low-grade paraventricular glioblastoma of malignancy. After receiving 2boli MP and 6cyclophosphamide,the control has showed a clear improvement.Case4:A 27-year-old patient has presented at the age of 23 a pseudo-bulbar syndrome and a pyramidal irritation syndrome.ARM has objectified punctiform hypersignals in T2 and FLAIR following the path of the pyramidal beam.The evolution was good after treatment

Discussion: The neuro-Behçet in its pseudotumoral form remains polymorphic posing the problem of differential diagnosis with classic tumors of CNS.Imagin's interpretation in isolation can be confusing,suggesting a tumor pathology requiring a surgical procedure.Such a hasty decision would then be harmful and regrettable when the solution was simply early medical treatment consisting of corticosteroid therapy and immunosuppressant

Conclusion: Neuro-Behçet varies between 4 and 59% depending on the series. An early diagnosis allows an adequate treatment avoiding the darkening of the prognosis.

Neurological manifestations of Behçet's disease

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Introduction: Behçet's disease is a rare systemic vasculitis of unknown etiology,which can present neurological manifestations in the form of neuro-Behçet syndrome,including parenchymal and extra-parenchymal manifestations.

Method: This was a retrospective single-center study involving 118cases of neurological involvement out of 563cases of Behçet's disease collected in the Internal Medicine Department of CHU Ibn Rochd in Casablanca, over a 17-year period from January2006 to December2023,meeting the diagnostic criteria defined in 1990 and the new criteria of 2013

Discussion: 118cases of neurological involvement were observed, representing a frequency of 21%. There were 88male patients (74.5%) and 30female patients (25%), with a male- to-female ratio of 2.93. All patients were of Moroccan origin except for one patient who was of Ivorian origin. They had an average age of 34.1years at the onset of neurological symptoms.Among the various neurological manifestations,we noted 65cases of parenchymal involvement represented by encephalitis,encephalomyelitis, and meningoencephalomyelitis,32cases of extra-parenchymal involvement represented by intracranial hypertension and involvement of cerebral arteries,and 6cases of mixed involvement.Pyramidal syndrome was found in 17patients (14%), pseudobulbar syndrome in 18cases (15%), and cerebellar syndrome in 3cases.The treatment received was corticosteroid therapy alone in 22 cases, corticosteroid therapy with immunosuppressive treatment in 66cases, 13cases benefited from biologic therapy,and anticoagulant treatment was initiated in 70cases. The outcome was favorable in 48cases, stable in 10cases, 3cases were resistant to treatment, and 3deaths were recorded

Conclusion: This study highlights the complexity and diversity of neurological manifestations of Behçet's syndrome and underscores the importance of imaging in determining and individualizing major clinical aspects.The prognosis remains poor, necessitating early and adequate management

Imaging in the neuro-Behçet: Basis of diagnosis and follow-up

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Introduction: Neuro-Behçet NB is known as nervous system involvement in Behçet's disease.It can be either intra-axial corresponding to parenchymal involvement or extra-axial affecting mainly the encephalic vessels.Magnetic resonance imaging MRI remains the best exploration method for detection and monitoring NB lesions

Method: We conducted a monocentric study within our department of internal medicine in Ibn Rochd University Hospital Center.200patients diagnosed with NB were included over a period of 20years extending from2004 to2023.All patients met the modified international criteria for Behçet's disease and medical records were processed by means of the operating sheets

Discussion: In our case series,22.9%of patients were affected by NB.A male predominance was noted with sex ratio of 2.39.MRI made it possible to objectify72.4%of parenchymal lesions,23.6%of vascular lesions and 4%of mixed lesions.The brain stem was the most affected (35%)represented mainly by pons.The periventricular white matter was affected in 15%of cases,the internal capsule in14.5%and the thalamus in11.5% of cases. MRI time-of-flight angiography showed ischemic vascular accidents in 13 cases.There were 6spinal cord injuries and 2pseudotumoral.The clinical and radiological evolution was good (64%) under treatment combining corticosteroid therapy, immunosuppressants and anticoagulants.Only16%relapsed and 2%died. MRI plays a crucial role in the process of diagnosis and monitoring the therapeutic response.For this fact,it provides detailed images that,combined with the clinical context and the course of the disease,can suggest a more specific diagnosis.It also allows differentiation between active and progressive lesions and those that are stable or linked to sequelae

Conclusion: NB is a rare disease but potentially life-threatening in the absence of appropriate diagnostic and therapeutic approaches.A prompt recognition is thus required to correctly treat.MRI plays a key role in diagnosis of central nervous system's lesions and their follow-up

Particularities of the Juvenile Form of Behçet's Disease

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Introduction: Juvenile Behçet's disease is a chronic vascular inflammatory condition diagnosed when at least two criteria of the disease are present in individuals under 15 affecting blood vessels of all sizes, impacting both veins and arteries. Its pathophysiology is poorly understood and heterogeneous involving autoinflammation and autoimmunity characterized by mucocutaneous, articular, digestive, neurological, and ocular manifestations.

Method: A retrospective, monocentric study conducted within the Department of Internal Medicine of U.H. Ibn Rochd over 42 years (1981-2023). 90 patients were diagnosed with juvenile Behçet's disease of a total of 1571 patients followed for Behçet during the same period.

Discussion: We identified 90 patients meeting the criteria for juvenile Behçet's disease, representing 5.73% of 1571 patients with 56 males and 34 females,sex ratio:1.6 vs 2.4 for the adults. The average age at diagnosis was 7 years in the juvenile group vs 12 years for adults. A family history of Behçet's disease was present in 12.5% of juveniles vs 5.1% of adults. Family history of aphthosis was found in 50% of juveniles, vs 13.3% of adults. Oral aphthosis was present in 92.5% of juvenile vs 99.6% of adults. Genital aphthosis was observed in 88.6% vs 87.9%, and erythema nodosum in 37.2% vs 15.43%. Ocular involvement occurred in 65.7% vs 60.8%. Articular involvement was observed in 55.3%. Vascular involvement was found in 6.67% vs 23.85%. Neurological involvement was present in 12.22% vs18.36%. Digestive involvement was seen in 7.8% vs to 9.18%.

Conclusion: Juvenile Behçet's disease presents specific features compared to adult Behçet's disease, as demonstrated by our study, including a stronger familial predisposition, earlier and more severe ocular involvement, yet paradoxically better treatment response and fewer relapses in juveniles. Articular involvement is equally frequent, while vascular, neurological, and digestive involvements are more common and severe in adults, aligning with literature data.

Clinical features and outcomes of vitreous hemorrhage associated with Behçet uveitis

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Introduction: Objective: To describe demographic and clinical features, and visual outcomes of patients with Behcet uveitis associated with vitreous hemorrhage (VH)

Method: Retrospective observational study, including patients diagnosed with Behcet uveitis associated with VH.

Discussion: Among 46 patients diagnosed with VH secondary to uveitis, 8 patients (17.4%) were diagnosed with Behçet disease. Seven patients were male adults, and one was a female child. The median age at presentation with VH was 25.5 years. VH was unilateral in 6 patients and bilateral in 2 patients. The median time between the diagnosis of uveitis and the occurrence of VH was 9.5 months, and VH was the presenting manifestation of uveitis in two patients. The initial median BCVA was 1.8 logMAR. Fundus examination after the VH clearance showed retinal hemorrhages in five eyes, diffuse retinal venous sheathing in six eyes, frosted branch angiitis in three eyes, branch retinal vein occlusion in four eyes, optic disc neovascularization in five eyes, and retinal neovascularization in three eyes. Fluorescein angiography showed occlusive retinal vasculitis with ischemia-induced newvessels in five eyes and non-occlusive retinal vasculitis with inflammatory optic disc neovascularization in three eyes. All patients were treated with systemic corticosteroids combined with conventional immunosuppressive agents. Laser photocoagulation of peripheral non- perfusion areas was performed in 4/5 eyes. Intravitreal injection of Bevacizumab was performed in 4 eyes, and pars plana vitrectomy in one eye. At the final follow-up visit, VH completely resolved in seven eyes, and the median final BCVA was 1 logMAR.

Conclusion: VH is a sight-threatening complication of Behçet uveitis. Fluorescein angiography is helpful to distinguish between ischemia- and inflammation-induced neovascularization. Besides aggressive medical treatment, peripheral scatter laser photocoagulation is recommended in those patients where ischemic retina is driving neovascularization.

Macular hole complicating Behçet's uveitis: a report of 5 cases

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Introduction: Objective: To report a series of macular hole complicating Behçet's uveitis.

Method: A retrospective case series including patients with macular holes associated with Behçet's uveitis.

Discussion: Four male and one female patient, diagnosed with Behçet's uveitis, presented with macular hole affecting a total of 6 eyes. The mean age at presentation was 28.8 years. Patients reported bilateral vision loss and eye redness in 3 cases, and unilateral symptoms in 2 cases. The mean best-corrected visual acuity in eyes with MH was 20/1000. Anterior segment examination revealed non-granulomatous uveitis in all cases. Macular hole was suspected during fundus examination and confirmed by Optical Coherence Tomography (OCT). OCT showed intraretinal cysts at the hole's edge in 4 eyes, elevated edges in 2 eyes, and flat edges in 2 eyes. Treatment modalities included systemic corticosteroids and immunosuppressive drugs (azathioprine). Pars plana vitrectomy with internal limiting membrane peeling and gas tamponade was performed in two patients (3 eyes) with closure of the macular hole in 2/3 eyes.

Conclusion: Macular hole is an unusual complication of Behçet's uveitis that can ultimately lead to severe vision loss. Visual outcomes after vitrectomy may be correlated to the severity of inflammation. Early diagnosis and aggressive treatment might prevent this invalid complication.

Frosted branch angiitis as a presenting feature of Behçet's uveitis

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Introduction: Objective: To describe demographic and ophthalmological characteristics of patients presenting with frosted branch angiitis associated with Behçet's disease.

Method: A retrospective case series including patients with Behçet's disease who presented with frosted branch angiitis.

Discussion: Results: Five male patients (7 eyes), with a mean age of 30 years, presented with frosted branch angiitis associated with Behçet's disease (BD). All patients reported sudden visual loss. Best corrected visual acuity (BCVA) ranged from negative light perception to counting fingers. Slit lamp examination revealed non-granulomatous uveitis and vitritis in all cases. Fundus examination showed occlusive retinal vasculitis with a characteristic frosted branch angiitis involving 2 to 4 quadrants of the retina. Macular optical coherence tomography (OCT) revealed macular edema in all cases, with serous retinal detachment in 4 eyes and epimacular membrane in 3 eyes. All patients received systemic steroids and conventional immunosuppressives. Scatter laser photocoagulation of peripheral non-perfusion areas was performed. Systemic work-up unveiled a coronary aneurysm in one patient, leading to his sudden demise, and multiple ischemic and hemorrhagic strokes detected on neuroimaging in another patient. Ocular complications included fellow eye involvement in one patient and vitreous hemorrhage in another. At the last follow-up, the mean BCVA was 20/200.

Conclusion: Frosted Branch Angiitis is a rare but sight-threatening ocular manifestation of Behçet's Disease which might be a presenting feature of the disease. Aggressive and appropriate therapy is mandatory to prevent vision loss.

Binocular diplopia revealing angioBehçet

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Introduction: AngioBehçet refers to the various vascular disorders that occur in Behçet's disease. They are frequent and can affect all types of vessel. Rapid diagnosis and intensive specific treatment are therefore essential to improve their prognosis. Arterial involvement remains rare, often characterized by thrombosis and/or aneurysm.

Method: This case report concerns a patient with exceptional arterial involvement revealing Behçet's disease. It involved a left carotid-cavernous aneurysm associated with bilateral carotid dissection.

Discussion: A 26-year-old patient, with no pathological history, presented with binocular horizontal diplopia accompanied by exophthalmos and headaches, with no visual acuity impairment or associated digestive symptoms. Ophthalmological examination revealed paralysis of the left abducens (VI) nerve. To investigate the diplopia, a cerebral arteriogram was performed, revealing a giant left carotid-cavernous aneurysm surgically treated with a stent in the left carotid artery. During post- operative follow-up, a bilateral carotid dissection was discovered. Questioning revealed a history of recurrent oral aphthosis and inflammatory joint pain of the gonalgia type, and a careful clinical examination revealed an achromic scar of an aphthosis on the scrotum. The diagnosis of angiobehcet's was retained given the association of bipolar aphthosis, inflammatory joint pain and this particular vascular involvement, and after ruling out other causes of carotid dissection such as connective tissue diseases of genetic origin (Marfan syndrome or Ehlers-Danlos syndrome), for which a search for specific mutations was negative. Treatment consisted of corticosteroid therapy combined with a cyclophosphamide-based immunosuppressant, resulting in clinical improvement and lesion stability.

Conclusion: The appearance of vascular involvement in Behçet's disease in young subjects with no vascular risk factors, combined with clinical symptoms that are sometimes misleading, can lead to a delay in diagnosis.

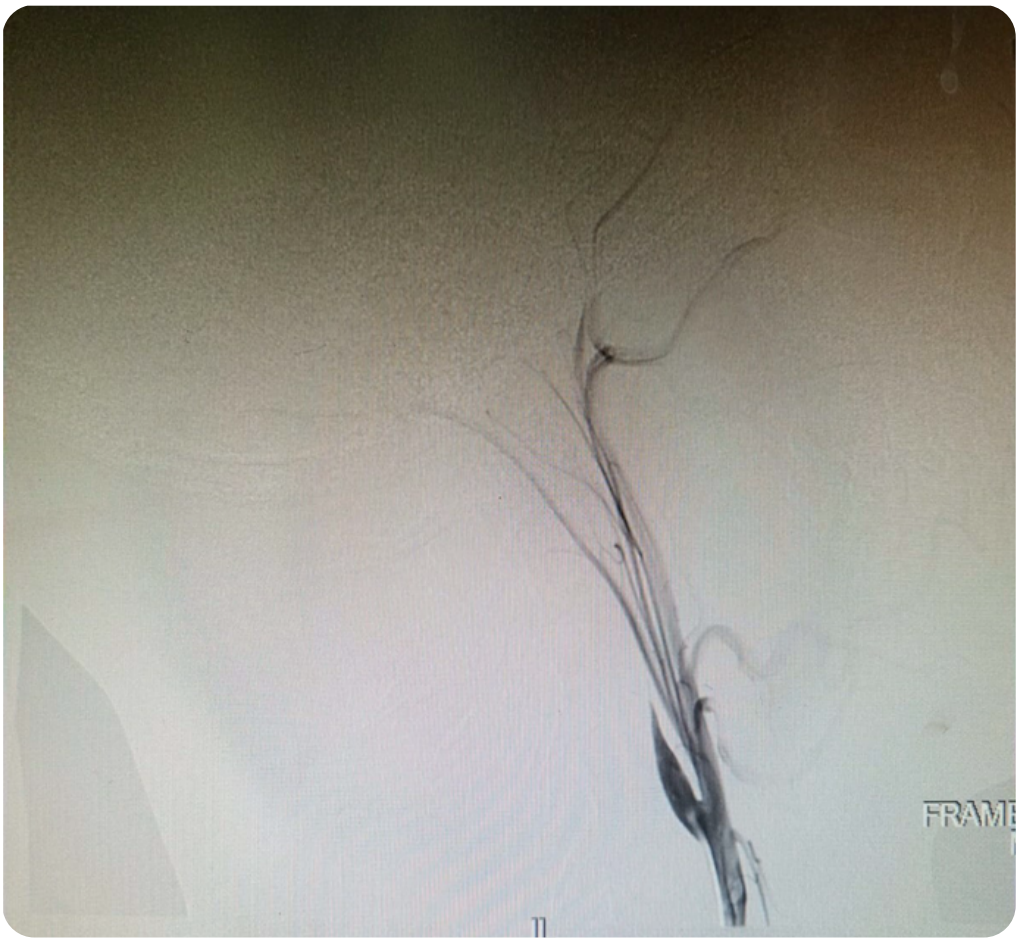

Figure 1. Cerebral arteriography showing absence of opacification of the right and left internal carotid arteries, with candle-flame arrest characteristic of carotid dissection.

Concomitant discovery of Behçet's disease, pulmonary tuberculosis and viral hepatitis C: Therapeutic strategy and management challenges

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Introduction: Behçet's disease (BD) is a systemic vasculitis of unknown origin that leads to a state of weakened immune system favoring the occurrence of infections. While pulmonary tuberculosis is a commonly encountered infection in Morocco, its association with viral hepatitis C in the context of Behçet's disease presents a challenging medical situation to handle.

Method: Mr E. M., aged 33, single, with a history of recurrent bipolar aphthosis, was admitted to the internal medicine department for weight loss and non-quantified fever, evolving for 3 months. Physical examination found genital aphthous scars. The lab results revealed a moderly elevated C-Reactive Protein (CRP) value of 64 mg/l, an acid-fast bacilli (AFB) smear-positive sputum, positive anti-HCV antibody. In addition to an aneurysmal dilatation of the thoracic aorta seen on computed tomography. The retained diagnosis was Angio-Behçet's associated with pulmonary tuberculosis and chronic viral hepatitis C without liver cirrhosis. The patient was treated with antibacterial drugs, colchicine, glucocorticoids and monthly boluses of cyclophosphamide. Treatment of the viral hepatitis C was deferred until the antibiotics were stopped.

Discussion: Pulmonary tuberculosis (TB) is a condition frequently observed in the Moroccan population. Its association with BD and viral hepatitis C makes managing each of the three conditions hard due to the prolonged necessary treatment and the underlying immunosuppression. This immunosuppression can be increased by the deficiency in cell-mediated immunity caused by both BD and TB, highlighting their close interconnection.

Conclusion: Knowing when to consider Behcet's disease in the presence of bipolar aphthosis is a valuable reflex. However, when general signs are at the upfront of clinical manifestations, infection must remain the physician's first concern, not only because of the underlying immunodepression, but also because of immunosuppressive treatments' effects.

What a simple headache can hide

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Introduction: Headaches are the first onset complaints in patients with Behcet disease (BD)affetcting the nervous system. It is a multisystem vascular-inflammatory disease of unknown etiology that can involve multiple systems in the body. Neurological affection is commonly misdiagnosed and is commonly taken as septic meningitis or chronic migraines

Method: We report the case of a 19-year-old female with no significant medical history, who presented with sudden onset severe migraine. She had a brain CT scan which showed a cerebral venous thrombosis, and a LP revealed a lymphocytic meningitis. She received an antimicrobial therapy, anticoagulants and prophylactic antiepileptic drugs. a brain MRI was done showing a cerebral venous sinus thrombosis of the lateral left sinus extensive to the jugular bulb and the right sigmoid sinus. Thoracoabdominopelvic CT scan was performed revealing a segmental pulmonary embolism. Than she developed oral ulcers, which allowed for a refinement of the diagnosis towards a very probable Behcet's disease.

Discussion: BD is an autoimmune vasculitis affecting all types of blood vessels. Studies have shown that neurological symptoms are present in less than 10% of cases and appear in about 5 to 6 years after the initial diagnosis. [1] This affection is called Neuro- Behcet's disease (NBD) and has a worse prognosis of BD, as 50% of NBD patients experience moderate to severe disability after 10 years of the disease evolution. [1] NBD is diagnosed based on clinical symptom presentation, CT scan and MRI findings, in addition to cerebrospinal fluid analysis with lymphocytosis as the prominent cell type in most cases. In the long term, studies suggest that NBD is associated to poorer prognosis and increased risk of relapse. [2]

Conclusion: Headaches are known to be among the common complaints managed daily by physicians. BD can masquerade as a simple headache leading to not consider serious nervous pathology such as vascular thrombosis and missing the correct diagnosis on multiple occasions.

Neuro-Behcet's disease

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Introduction: Neurological manifestations in Behçet's disease represent between 4 to 49% of systemic manifestations and constitute significant morbidity and mortality risks. Our study aims to review the epidemiological profile and various aspects of neurological involvement in Behçet's disease.

Method: A retrospective study of 399 Behçet's disease patients fulfilling the International Study Group criteria for Behçet's disease was conducted over 14 years (from January 2010 to December 2023) at the University Hospital Mohamed VI of Marrakech

Discussion: There were 59 patients (14.7%); 33M/26F with neurological manifestations. Isolated parenchymal involvement was the most frequent aspect (49.15%), followed by vascular involvement (23.72%), mainly cerebral thrombosis. The major clinical signs are: headaches (50.84%) and focal deficits (42.37%). Peripheral neurological involvement was present in 3 cases (5.08%). No significant correlation was found between neurological and extraeurological damage. Studies of cerebrospinal fluid revealed aseptic hypercytosis with lymphocytic predominance in 7 patients (11.86%). Brain MRI results were normal in seven cases (11.86%), and revealed abnormalities in 52 cases (88.13%), including white matter signal abnormalities (28.81%), cerebral venous thrombosis (11.86%), demyelinating lesions (10.16%), signs of increased intracranial pressure (5.01%), vasculitis (3.38%) and a pseudo-tumoral appearance (3.38%). Treatment included methylprednisolone pulses, oral steroids, colchicine, immunosuppressants, biologics (Tocilizumab) and anticoagulants, with improvement in 44.06% of cases, recurrence in 8.47% and mortality in 1, 69% of cases.

Conclusion: In our study, parenchymal involvement was the most frequent manifestation with a good clinical outcome

Infarct during Behçet's Disease: A Case Report

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Introduction: Behçet's disease is a systemic vasculitis with a polymorphic clinical presentation. Bone lesion has rarely been reported, particularly bone infarct.

Method: We present the case of a 50-year-old patient with a 20-year history of Behçet's disease, diagnosed based on cardinal signs, ocular, articular, vascular, and neurological involvement. He was admitted for left thigh pain radiating to the ipsilateral leg, with clinical examination revealing hypersensitivity of the leg. X-ray showed localized osteolysis with some calcifications. CT scans revealed a bone infarct. Therapeutic management consisted of medical treatment with corticosteroids and orthopedic treatment. The evolution wasmarked by a clinical and radiological improvement.

Discussion: Articular manifestationis common in Behçet's disease, usually benign. Bone infarct, particularly in the metaphyseal-diaphyseal region of long bones, is rarely reported. The role of long-term corticosteroid therapy is widely debated. However, the prognosis with treatment is generally favorable.

Conclusion: Bone involvement in Behçet's disease is exceptional. Clinicians should consider it in cases of unexplained bone pain in patients on long-term corticosteroid therapy.

Cluster analysis of phenotypes of patients with Behçet's syndrome from a Tunisian population

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Introduction: Behcet's syndrome (BS) is a chronic multisystem disease with an heterogenous clinical presentation which varies according to the ethnic background. The purpose of this study was to analyse cluster phenotypes of BS based on clinical manifestations.

Method: This was a cross-sectional study of BS patients from the Internal Medicine and Ophthalmology Departments of Fattouma Bourguiba University Hospital, Monastir, Tunisia, between January 2004 to December 2023. The TwoStep Cluster Analysis with clinical variables was performed to determine subgroups of patients

Discussion: A total of 449 BS patients were included:137 females (30.5%) and 312 males (69.5%) (Sex ratio M/F was 2.27). Mean age at diagnosis was 32 ±10.5 years. Six clusters (C1-C6) were observed. C1 (n = 132) showed the skin and mucosa type. In C2 (n = 56), ocular manifestations were associated to angio BS in 89.3% and cutaneous manifestations in 87.5%. In C3 (n = 103), ocular manifestations were associated to cutaneous manifestations only. In C4 (n = 39), cutaneous manifestations were associated with angio BS in 61.5% and articular manifestations in 97.4%. C5 (n = 44) was the neuro BS cluster with cutaneous manifestations. C6(n=75) consisted of patients with angio BS and cutaneous manifestations.

Conclusion: Cluster analysis of Tunisian patients with BS showed that ocular manifestations may occur with cutaneous involvement associated or not with angio BS. The latter may also present with cutaneous manifestations associated or not with articular symptoms. Neuro BS seems to occur in association with cutaneous manifestations only

Behçet's Disease: The Pitfall of Unrecognized Associations

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Introduction: Behçet's disease (BD) is a multisystemic vasculitis with an etiopathogenesis that remains incompletely elucidated. In the absence of a diagnostic test, diagnosing BD remains challenging and relies on a set of criteria with exclusion of potential differential diagnoses. We report 2 cases of associated or complicated Behçet's disease

Method: Case 1 A 42-year-old, was treated for portal thrombosis diagnosed 4 years ago and put on anticoagulant. The etiological work-up revealed antiphospholipid positivity
and the presence of the PNH clone. Anamnesis revealed the occurrence of recurrent oral aphthosis for the past 4 years. Clinical examination revealed oral aphthosis and genital scars with a positive pathergy test. The diagnosis of portal thrombosis in the context of BD, APS and paroxysmal nocturnal hemoglobinuria was established. The patient was treated with corticosteroids, cyclosporine, anticoagulants, and colchicine. The evolution was favorable Case 2 A.N aged 39, was admitted with a superior vena cava syndrome. He had a recurrent oral and genital aphthosis. Furthermore, a deficiency in protein C, hyperhomocysteinemia with vitamin B12 deficiency were identified. Anti-intrinsic factor and anti-parietal cell antibodies were positive. The diagnosis of BD associated with protein C deficiency, hyperhomocysteinemia, and pernicious anemia was established. The patient was treated with corticosteroids, immunosuppressants, and anticoagulants along with vitamin B12 supplementation. Unfortunately, the patient died as a result of a recurrence of thrombosis in the portal trunk, complicated by Acute Esophageal Variceal Bleeding

Discussion: We insist on the necessity of evoking BD in any young patient presenting with venous thromboembolic disease.However, the presence of authentic BD should not preclude conducting an exhaustive etiological investigation

Conclusion: The association of BD with other pathologies further complicates the diagnostic process, often resulting in polymorphous clinical presentations

Fistulization of a subclavian artery aneurysm in the left main bronchus in the setting of Behçet's disease.

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Introduction: Behçet's disease (BD) is systemic vasculitis. It is characterized by multiple manifestations,including cutaneous,ocular,articular,neurological and vascular involvement.Vascular involvement is one of the main causes of mortality in BD.This is a case of a patient in whom BD was revealed by hemoptysis

Method: A 39-year-old with a history of a miscarriage, aseptic lymphocytic clear meningitis treated as tuberculous complicated by paresthesia of both lower limbs, and recurrent oral ulcers. She presented with mild hemoptysis. Clinical examination revealed oral aphthosis, genital scarring, and an added murmur along the left subclavian artery. Bronchoscopy was normal, a CT scan identified 2 pseudoaneurysms one at the left subclavian artery the other at the left common iliac artery. We diagnosed BD with cutaneous and vascular tropism. The patient was treated with corticosteroids and cyclophosphamide. 2 months later, the oral aphthae had disappeared, but the hemoptysis persisted. A second bronchoscopy revealate a fistula between the left main bronchus and the aneurysm of the subclavian artery. Given the severity of the vascular lesions, surgical treatment of the aneurysms became imperative. Unfortunately, the patient died following hemorrhagic shock complicating rupture of the subclavian aneurysm in the left main bronchus

Discussion: Patients with symptoms such as chest or abdominal pain or signs of internal bleeding the context of BD require prompt evaluation to exclude the presence of aneurysm. Managing aneurysms in patients with BD can be complex and requires an approach including corticosteroids,immunosuppressants, or biologic therapy; surgical or interventional radiology are considered after inflammation subsides, except in cases where the patient's prognosis is threatened

Conclusion: Aneurysms represent a serious vascular complication of BD, requiring clinical vigilance and appropriate management. A thorough understanding of these vascular risks is essential for effective patient care in this disease.

Dermatological manifestations during Behçet's disease: a series of 60 cases

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Introduction: Dermatological manifestations are the cornerstone of the diagnosis of Behçet's disease (BD), a systemic vasculitis characterized by a variety of clinical manifestations, in which mucocutaneous involvement is the most frequent. The aim of our study was to describe the dermatological manifestations of BD in a cohort of Moroccan patients.

Method: Retrospective and descriptive study including 60 patients with BD, collected in an internal medicine department over a 5-year period between 2019 and 2024. Data were entered and analyzed using JAMOVI software version 2.3.

Discussion: Our study included 60 patients with Behçet's disease, 48 men and 12 women, for a M/F ratio of 4. The mean age at diagnosis was 30.4 ± 8.5 years, ranging from 12 to 50 years. Clinical manifestations were dominated by mucocutaneous involvement, present in all patients (100%) and inaugural in 75% of cases. The cumulative frequencies of dermatological manifestations were as follows: oral aphthosis (100%), genital aphthosis (53.3%), pseudofolliculitis (26.7%), erythema nodosum (10%), and hypersensitivity to sampling points with a positive pathergic test in 25% of patients (test performed in all patients). No cases of cutaneous aphthosis were observed. Perianal aphthosis (n=1) and vascular purpura (n=1) were noted. The mean time between the first mucocutaneous manifestations and diagnosis was 4.2 years. The type of dermatological involvement was not influenced by age of onset or gender. The main associated manifestations were joint involvement in 56.7% of cases, ocular involvement in 45% of cases, angio-Behçet in 40% of cases, neuro-Behçet in 18.3% of cases, entero-Behçet in 15% of cases and psychiatric involvement in 11.6% of cases.

Conclusion: Our study reveals the clinical diversity of mucocutaneous involvement in BD and confirms the high frequency of dermatological manifestations in this disease.

Late-onset Adamantiades-Behçet's disease

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Introduction: Adamantiades-Behçet's disease (ABD) usually manifests during the 2nd to 3rd decade of life. Late-onset (lo) ABD, namely manifestation after the age of 40 years, is not well defined. The purpose of the present study was to evaluate the prevalence and the clinical features of patients with lo-ABD and compare them with early-onset ABD patients.

Method: A systematic review and meta-analysis was conducted in accordance with the MOOSE. PubMed and Scopus databases, bibliographies of selected articles and abstract books of ISBD conferences published 1990-2022 were searched using relevant MeSH terms and free text keywords. Moreovee, results in the MEDLINE database using age filters have been registered. Statistical analysis was performed by using the Chi-Square Test Calculator ($p<0.05$, Bonferroni correction).

Discussion: Twelve original investigations related to the lo-ABD from Algeria, China, Greece, Iran, Israel, Korea, Lebanon, Morocco, Taiwan, Tunisia and Turkey (3 studies) were included. Males represented 57% of the lo-ABD patients (n=361/631). Among the 8,524 patients reported in comparative lo vs. early-onset (eo) studies, only 596 patients exhibited a lo-ABD (7%). Oral ulcers were present in 98% of patients, followed by genital ulcers (72%), ocular lesions (47%) and a positive pathergy test (45%). In the meta-analysis, lo-ABD was associated with significantly lower rates of vessel involvement, such as erythema nodosum, ocular and vascular lesions (all $p<0.00001$). No sex-associated statistical differences of clinical signs were calculated. Recurrent oral and genital ulcers, ocular lesions and a positive pathergy test were clinical signs with a prevalence of ≥45%. It is associated with significantly lower rates of vessel involvement, such as erythema nodosum, ocular and vascular lesions than in eo-ABD.

Conclusion: In conclusion, lo-ABD might occur predominantly in males with a milder course in comparison to eo-ABD, especially regarding a vessel involvement.

Adamantiades-Behçet's disease and COVID-19

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Introduction: The course of Adamantiates-Behçet's disease (ABD) in patients infected with COVID- 19 has not been widely studied diuring and after the pandemic period.

Method: Patients registered in the German Registry of Adamantiades-Behçet's disease (GR- ABD) who were reported infected with SARS-CoV-2 during the period 1.2.20-31.10.22 were prospectively assessed.

Discussion: 14/900 (1.6%) registered patients have been infected with SARS-CoV-2. They were 45 year-old (IQR 42-56) with equal sex distribution and of different nationalities. The duration since ABD diagnosis ranged from 1-22 years. 9 patients (64.3%) had systemic involvement. 13 patients received systemic treatment. 7/12 patients were HLA-B51+. 9/9 patients had low vit. D levels (5.41–59.9 nmol/l; NL >75). 13 patients (92.9%) were asymptomatic or reported moderate fatigue and transient myalgia. Loss of taste was reported in the 2 patients (14.3%) infected in 2021. No hospitalisation/changes of ABD medication was required in 13 patients (92.9%) and no COVID-19-specific treatment has been introduced in 10 patients (71.4%). All patients recovered completely, although 2 patients (14.3%) had long-term sequelae.

Conclusion: The prevalence of COVID-19 in GR-ABD patients was substantially lower than the median seroprevalence in Germany (2.93%). The ISBD has also reported in January 2021 lower COVID-19 prevalence in ABD patients compared to the general population. This might be due to extra precautions advised in risk groups. Lower hospitalization, intensive care and death rates of COVID-19-infected ABD patients were also observed in 2021/2022 in other cases series, when compared with 2020. Our only hospitalized patient has been infected in 2020. Our data suggest that systemic ABD involvement is not necessarily associated with a severe COVID-19 course and that the medication-induced immunosuppressed/immunomodulated ABD state does not predispose to SARS-CoV-2 infection. Vitamin-D levels but not HLA-B51 appeared to bear on the severity of COVID-19 in ABD patients.

Prepapillary vitreous exudate: a rare, but typical feature of Behçet disease

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Introduction: Objective: To describe a series of prepapillary inflammatory vitreous exudate (PIVE) associated with Behçet's disease.

Method: A retrospective case series including patients with Behçet's disease who presented with PIVE. Demographic, ophthalmological data at presentation and during follow-up were collected and analyzed.

Discussion: Results: Six patients (7 eyes) were included in this study. Mean age at presentation was 35.5 years old (range, 19-52). All patients were male. At presentation, PIVE was unilateral in 5 patients and bilateral in 1 patient. Fundus examination revealed a
localized inflammatory vitreous exudate overlying an infiltrated optic disc and peripapillary retina in all eyes. Associated ocular findings included retinal vasculitis in 2 eyes of 2 patients, multifocal retinitis in one eye and macular edema in 2 eyes of 2 patients. There were associated 1 to 2 + vitreous cells in all eyes. Optical coherence tomography of the optic nerve head showed a "mushroom-shaped" hyperreflectivity of the PIVE seen clinically in all cases. The mean time to resolution of the PIVE was 7 days. All patients received systemic steroids and conventional immunosuppressives. After a mean follow-up of 46 months (range, 6-132 months), a recurrence was noted in the same eye in one patient and in the fellow eye in another patient.

Conclusion: PIVE, also called prepapillary vitreous opacity, has been rarely reported in the literature. This finding is considered as a typical feature of Behçet's disease uveitis. It may be the presenting feature of the disease, and OCT can play an essential role in its accurate detection and in follow-up after early and appropriate treatment.

A 21-year-old male with intracardiac thrombus

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Introduction: Behçet's syndrome is a systemic inflammatory disease generally presented with the triad ofpseudofolliculitis, oral and genital ulcers.. Intracardiac thrombus (ICT) formation is an uncommon but important complication of BD.

Method: A 21-year-old man with Behçet's syndrome was hospitalizedwith a history of cough, fever, chest pain, hemoptysis, and weight loss. The patient was placed under antibiotic treatment, but the evolution was marked by the non-clinical improvement and persistence of hemoptysis. Chest scan Transthoracic and echocardiography revealed a right ventricular thrombus. The patient was initially received colchicine, anticoagulation, corticosteroids,azathioprine, monoclonal TNF-alpha antagonists and heart surgery. The clinical evolution was favorable.

Discussion: Intracardiac thrombus is an uncommon complication of Behçet's disease. Classic cardiac manifestations, like pericarditis and endocarditis, are exceptional. A genetic predisposition is incriminated since it occurs predominantly in patients from the Mediterranean basin. Corticosteroids, azathioprine, cyclosporine A, and cyclophosphamide are recommended in the management of acute deep vein thrombosis. In resistant cases, anti-tumor necrosis factor agents could be also effective. The addition of anticoagulants to prevent relapses is discussed. Surgical treatment is not recommended in the active phase of the disease.

Conclusion: We present a case of intracardiac thrombus reveals Behçet's disease successfully treated with immunosuppressant, corticoids, anticoagulant and surgery. Early echocardiography seems advisable to detect the presence of cardiac involvement.

Cyclophosphamide and Neuro-Behçet's Disease: An Old Therapy Still Effective

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Introduction: Behçet's disease (BD) is a chronic systemic vasculitis of as yet unidentified etiology. Cutaneous-mucosal manifestations, often represent the initial symptoms of the disease; its severity consists of cardiovascular, ocular and neurological involvement. The classic therapeutic approach to BD is based on glucocorticoids, immunosuppressants and colchicine; currently, the use of biologics is increasingly considered for severe or refractory forms. We report a case of refractory neuro-BD to biologic therapy

Method: A 29-year-old patient, followed for cutaneous and neurological BD. Neurological involvement was suspected due to the recurrence of meningitis episodes requiring
an average of 3 hospitalizations per year. Clinical examination revealed a febrile meningeal syndrome, scar from genital aphthae, and concomitant oral aphthae with the meningeal syndrome. Paraclinical assessment revealed an inflammatory syndrome and aseptic lymphocytic clear fluid meningitis. Brain imaging was normal. The patient was treated with corticosteroids, colchicine, and azathioprine, achieving
a response lost by the 4th year of treatment. Cyclophosphamide was rejected by the patient due to the risk of infertility. Subsequently, methotrexate was initiated without improvement, followed by tocilizumab. Initial improvement was noted, but a loss of response was observed by the 2nd year. In light of the failure of previous treatments, cyclophosphamide was considered, followed by azathioprine, with a favorable outcome as no relapse occurred after a follow-up of 4 years.

Discussion: Neuro-BD is a severe and potentially serious condition, requiring the use of immunosuppressants such as Cyclophosphamide, Azathioprine, and Methotrexate in the therapeutic arsenal; the current approach leans more towards biologic therapies

Conclusion: Despite various studies showing the effectiveness of biologic therapies in BD our observation argues that even in the era of biologic therapies, conventional treatments still retain their indication and efficacy

Arterial involvement in Behçet's disease: Aneurysms and thrombosis

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Introduction: Behçet's disease (BD) is a multisystem vasculitis. Arterial involvement is an important facet of the disease. Various forms can be described including arterial thrombosis and aneurysms. The study aimed to describe the different arterial involvements in BD patients, the therapeutic arsenal and the outcomes.

Method: We designed a retrospective study. The medical records of adult BD patients who had arterial involvement were collected.

Discussion: The study included 44 patients. A male predominance was observed (91%). The mean age at disease onset was 34 years [18-61]. The Coexistence of aneurysms with arterial and/or venous thrombosis was observed in 31 patients (70%): 10% had arterial thrombosis, 71% had venous thrombosis, and 19% had both arterial and venous thrombosis. Hughes-Stovin syndrome was noted in eight patients (18%). The topographic distribution of aneurysms was: infrarenal aorta (29%), pulmonary arteries (20%), femoral arteries (9%), cerebral arteries and primitive iliac arteries (7%), 5% in each one of subclavian arteries, cardiac circumflex arteries, the ascending aorta, and popliteal arteries and 2% in coronary arteries, splenic arteries, renal arteries, the celiac trunk, and the superior mesenteric artery. All patients received colchicine, intravenous methylprednisolone and/or oral corticosteroid therapy and immunosuppressors such as cyclophosphamide (for induction), and azathioprine (for maintenance). Twenty-three patients (52%) received medical treatment, only. In these patients, the imaging control showed aneurysm stabilization (35%), vanishment (26%) size reduction (26%), and new aneurysms (13%). Surgical interventions were needed in 21 patients (48%). The outcome was favourable in 44% of cases. However, 38% experienced prosthesis thrombosis a few years after the surgery and 18% underwent limb amputation.

Conclusion: This study showed the different forms of arterial involvement in Behçet's disease and outlined the poor surgery outcomes.

Prevalence of Behçet's disease in 218 patients with uveitis, internal medicine consultant in a private outpatient setting, Settat, Morocco

Said El Kettani, Liberal

Introduction: Behçet's disease is a systemic vasculitis. Ocular damage represents one of the major diagnostic and prognostic criteria. The published series concern almost exclusively patients followed in hospitals. This work was undertaken to determine the clinical, diagnostic and prognostic features of Behçet's disease in patients with uveitis treated in a private internal medicine practice in a small Moroccan town, in this case Settat.

Method: This is a prospective study conducted from September 2009 to December 2023 that involved 218 patients with uveitis. They are on average 42.3 13.5 years old. Behçet's disease was selected on the International Criteria for Classification of Behçet's disease, revised in 2013. To assess the evolution, we contacted patients who had lost sight by telephone. The statistical analysis was carried out under SPSS version 20.

Discussion: The prevalence of Behçet's disease in patients with uveitis is 36.4%. This prevalence is not influenced by age or sex (p = 0.783 and p = 0.165). Patients with uveitis and Behçet disease have significantly more family history of Behçet disease (p = 0.003), more oral Aphtosis (p = 0.000), more genital Aphtosis (p = 0.000), more skin Involvement (p = 0.000), Joint Manifestations (p=0.002) and neurological manifestations (p=0.021). Only 34% of patients were contacted by phone. The evolution was considered indeterminate in 144 patients (66.1%) satisfactory in 51 patients (23.4%) and not favorable in 23 patients (10.6%). Adverse outcomes are less common in patients with Behçet disease than others (p = 0.056).

Conclusion: The prevalence, in outpatient internal medicine, of Behçet's disease in patients with uveitis is 36.4%. The adverse course of uveitis is encountered more frequently in patients without Behçet's disease (p = 0.056).

Hughes Stovin syndrome in Behçet's disease

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Introduction: Hughes Stovin syndrome is a rare form of angio-Behçet's disease (BD) defined by the coexistence of pulmonary aneurysms and venous thrombosis. This study aimed to report the clinical features and the outcome of Hughes stovin syndrome in BD patients.

Method: A descriptive, retrospective study was conducted by the internal medicine team of the Rabta University Hospital. The medical records of adult patients with angioBehçet's disease from 2002 to 2024 were collected.

Discussion: The study included eight patients, seven were men. The mean age at the disease onset was 29.5 years [18-47]. In five cases pulmonary aneurysms were segmental. Multiple pulmonary artery aneurysms were found simultaneously in five patients. In five cases, pulmonary embolism and pulmonary aneurysms coexisted. Cerebral venous thrombosis and intraventricular thrombi were found in three and two cases, respectively. Associated peripheral venous thrombosis included: deep vein thrombosis of the lower limbs (N=5) and the inferior vena cava (N=3). The coexistence of pulmonary aneurysms, pulmonary embolism, cerebral venous thrombosis and inferior vena cava thrombosis was observed in two patients. No case of coexistent pulmonary aneurysms and other arterial aneurysms can be reported. Extravascular manifestations included: bipolar aphtosis (N=7), neuro Behçet's (N=2), joint involvement (N=1), and panuveitis (N=1). Hemoptysis revealed the disease in seven cases and chest pain in one. All patients received medical treatment with colchicine, curative anticoagulation, intravenous and oral route corticosteroids associated with cyclophosphamide (six perfusions: N=6, four perfusions N=1, twelve perfusions N=1) followed by maintenance therapy with Azathioprine. No patient underwent surgery. After treatment, pulmonary aneurysms disappeared in four cases, persisted in three cases and new pulmonary aneurysms appeared in one case.

Conclusion: Hughes Stovin syndrome is rare. Its association with intracardiac thrombi is a variant of the disease.

Interest in the PNN/lymphocyte ratio: new marker of inflammation during vascular thrombosis in Behcet's disease?

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Introduction: Behcet's disease is an inflammatory vasculitis characterized by stimulation of neutrophils and T lymphocytes and inflammatory sites. The PNN/Lymphocyte ratio is considered an inflammation marker of the systemic inflammatory response. We evaluated the diagnostic performance of the PNN/Lymphocytes ratio during the inflammation accompanying the vascular thrombosis of Behcet's disease through specificity and sensitivity tests.

Method: This study is a monocentric retrospective descriptive and analytical study spanning from 2003 to 2022 including the files of patients followed for Behcet's disease associated with vascular thrombosis.

Discussion: We collected 136 cases. The average age of our patients was 40.6 years +/- 11.4. Sex ratio was: 5.18 (114 M/22 F). deep venous thrombosis was present in 108 cases (79%) and arterial thrombosis in 28 cases (21%). The median CRP was 36 mg/l (10-76) and the erythrocyte sedimentation rate was 22 (6-30). The median PNN was estimated at 7320 ele/mm 3 (4825-8811), and the median of lymphocytes was 1895 ele/mm3 (1100-2560). The median PNN/LYM ratio was 4.51 (2.39 - 6.31). The statistical analysis did not identify significant statistical links between this ratio of PNN/LYM and sex; CRP and ESR as well as the different clinical presentations of thrombosis (venous – arterial), (superficial – deep), (proximal – distal). The sensitivity of the PNN/lymphocyte ratio was estimated at 62.2% and its specificity was estimated at 66.7%. Its positive predictive value was estimated at 92.2% and its negative predictive value was estimated at 22.2%. the positive likelihood ratio of this marker was low and estimated at 1.86. the negative likelihood ratio was low at 0.56, suggesting that the diagnostic gain when the test is positive or negative is low. The ratio of air under the curve was low at 0.5.

Conclusion: These results imply that the PNN/Lymphocyte ratio is a useless test in the diagnosis of inflammation during vascular thrombosis in patients followed for Behcet's disease.

Benefit and risk: the anti TNF alpha dilemma in ocular Behçet's treatment

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Introduction: Anti-TNF alpha drugs have proved effective in treating severe ocular damage in Behçet's disease, but sometimes at the cost of side effects.

Method: We describe the case of a patient with retinal vasculitis during Behçet's disease, who developed paradoxical psoriasis in close chronological relationship with infliximab treatment.

Discussion: A 23-year-old patient with panuveitis and bilateral retinal vasculitis in Behçet's disease, who had failed to respond to conventional treatment, was started on infliximab 5mg/kg/cure at So S2 S6 S8 and then every 2 months with good ocular improvement. 36 months after treatment, the patient developed a psoriasiform erythemato -squamous rash covering 30% of the body surface area, with an estimated pasi of 11.7. Biotherapy was stopped and the patient received 10 phototherapy sessions with disappearance of the skin lesions. However, his ocular involvement reappeared, forcing us to reintroduce infliximab with spaced treatments.

Conclusion: Paradoxical skin lesions induced by anti-TNF alpha drugs are increasingly being described. In paradoxical psoriasis, discontinuation of anti-TNF therapy should be reserved for patients whose lesions cover more than 5% of their body surface area, and whose quality of life is impaired.

Assessing Treatment Adherence in Tunisian Behçet's Disease Patients

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Introduction: Behçet's disease (BD) is a chronic, inflammatory disease with multisystem involvement. Treatment adherence is an important parameter in the follow-up of patients. The purpose of this study was to assess treatment adherence in BD patients and its associated factors.

Method: We conducted a transversal monocentric study in the Rabta University Hospital Center enrolling BD patients aged over 18 years. The 4-item Morisky Green Levine Medication Adherence Scale was used.

Discussion: Our study included 41 participants, with a mean age of 43.1 ± 11.5 years and a male-to-female ratio of 3.2. The mean duration of the disease was 8.9 ± 9.1 years. Vascular involvement was observed in 31.7% of patients: lower limb deep vein thrombosis (19.5%), aneurysms (9.8%), pulmonary embolism (2.4%) and Hughes- Stovin syndrome (4.9%). Ocular involvement was observed in 28.7% of patients: retinal vasculitis (16.7%) and panuveitis (11.9) were the most common manifestations, followed by anterior (7.9%) and posterior uveitis (7.1%). Neuro- Behçet was observed in 19.5% of patients. Treatments included: Colchicine (100%) Corticosteroids (59.5%) cyclophosphamide (26.2%), azathioprine (21.1%), and Infliximab (2.4%). Most patients had moderate adherence to treatment (51.3%), 36.6% had low adherence and 12.1% showed high adherence, respectively. There was no statistically significant association between age and treatment adherence (p=0.24). Neither the duration of the disease (p=0.19) nor the treatment type showed an association with the treatment adherence: (p=0.9) for both corticosteroids and infliximab, (p=0.32), (p=0.14), and (p=0.6) for colchicine, azathioprine, and cyclophosphamide, respectively.

Conclusion: Non-adherence is common in BD patients. Tailored interventions may be needed to enhance adherence and raise awareness among BD patients.

Prevalence of bundle retinal nerve fiber layer defects in patients with new-onset Behçet Uveitis

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Introduction: Bundle retinal nerve fiber layer (RNFL) defects in Behcet Uveitis are detected commonly after resolution of retinal infiltrates (superficial retinitis). Thus, their presence can serve as a clue of previous overlooked retinal infiltrates. The aim of our study was to assess the prevalence of bundle RNFL defects in newly diagnosed Behçet uveitis during the decade of 2013-2023 in a referral center in Tunisia, North Africa.

Method: The charts of 101 patients newly diagnosed with Behçet uveitis were retrospectively reviewed to screen for a bundle RNFL defects on color and red free fundus photographs.

Discussion: Of 101 patients, 10 (9.9%) presented with a bundle RNFL defect unilaterally (7 patients) or bilaterally (3 patients). The mean age was 25 years (range, 11–39). There were 8/10 males and 2/10 females. Of 13 eyes with RNFL defect, 6 (46.15%) had a papillomacular bundle defect, 5 (38.46%) had a superior or inferior arcuate defects and 2 (15.38%) had co-occurrence of both defect types. Associated ocular findings included mild to moderate vitritis in 12 eyes (92.31%), inferior pearl-like vitreous precipitates in 4 eyes (30.77%), active retinal infiltrates in 4 eyes (30.77%), retinal vasculitis in 9 eyes (69.23%), serous retinal detachment in 3 eyes (23.08%), and optic disc edema in 7 eyes (53.85%).

Conclusion: RNFL defect in Behcet's disease is considered as a retrospective indicator of a regressed retinal infiltrate and posterior pole involvement. In this single center cross- sectional study, we found bundle RNFL defects in 9.9 % of patients with new-onset Behçet uveitis.

Relapse in Behçet's Disease: prevalence and associated factors

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Introduction: Behçet's disease (BD) is a multisystem inflammatory vasculitis with an important heterogeneity among patients regarding demographic and clinical features, treatment response, frequency, and severity of relapses. This study aimed to determine the most relapsing forms of the disease and their associated factors.

Method: We conducted a retrospective monocentric study in the internal medicine ward of the Rabta University Hospital Center. BD patients aged over 18 years were included.

Discussion: The study included 41 patients. The mean age was 43.1±11.5 years. Male to female ratio was 3.2. The mean duration of the disease was 8.9±9.1 years. Oral and genital ulcers were observed respectively in 83,3% and 64.3% of cases. Articular and vascular involvements were noted respectively in 33.3% and 31.7% of patients. Ocular involvement was found in 28.7% of patients with bilaterality observed in 21.4% of cases. Neurological manifestations were noticed in 19.5% of patients, half of whom displayed parenchymal involvement, while the remaining half presented non-parenchymal manifestations. All patients were treated with colchicine. Corticosteroids were administered to 59.5% of patients, while 26.2%, 21.1%, and 2.4% of patients received cyclophosphamide, azathioprine, and infliximab, respectively. The highest relapse rate was observed in patients with ocular involvement (38%), followed by those with vascular involvement (29.8%), especially deep vein thrombosis (20%). Low treatment adherence was associated with the recurrence rate (p=0.032), as well as smoking (p=0.025) and male gender (p=0.046). There were no statistically significant associations with the disease duration (p=0.71).

Conclusion: Our study highlighted the frequency of relapse in BD, particularly in patients with ocular and vascular involvement. Factors such as low treatment adherence, smoking, and male gender contribute to this increased risk. Hence, the importance of tailored treatment approaches, and patients'-based approach to improve outcomes.

Anxiety and Depression Prevalence in Tunisian Behcet's Disease Patients

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Introduction: Behçet's disease (BD) is a rare type of vasculitis affecting both small and large blood vessels. Psychological impairment appears to be frequent, yet few data are available. The aim of our study is to evaluate the depression and anxiety levels in BD patients.

Method: We conducted a transversal monocentric study in our internal medicine department enrolling BD patients. The Hospital Anxiety and Depression Scale was employed to assess the symptoms of anxiety and depression

Discussion: Our study enrolled 41 patients with an average age of 43.1±11.5 years and an average disease duration of 8.9±9.1 years with a male to female ratio of 3.2. Oral and genital ulcers were observed respectively in 83,3% and 64.3% of the patients. Vascular involvement was observed in 31.7% of the patients with 19.5% presenting with lower limb deep vein thrombosis. Vascular aneurysms were diagnosed in 9.8% of cases, with half of them located in the abdominal aorta and the rest equally distributed between the femoral and coronary arteries. Moreover, pulmonary embolism and Hughes-Stovin syndrome were identified in 2.4% and 4.9% of patients, respectively. Ocular involvement was detected in 28.7% of patients with bilaterality observed in 21.4% of cases. Neurological manifestations were detected in 19.5% of the patients, half of whom displayed parenchymal involvement, while the remaining half presented non-parenchymal manifestations. All patients were treated with colchicine. Corticosteroids were administered to 59.5% of patients, while 26.2% and 21.1% of patients received cyclophosphamide and azathioprine, respectively. Depression and anxiety were respectively absent in 32% and 53.7% of the patients, likely present in 39% and 24.4% of the patients, and confirmed in 29% and 22% of the patients, respectively.

Conclusion: Our study reveals that anxiety and depression are common among BD patients. Further investigation is needed to evaluate the relationship between BD and the psychological wellbeing of the patients.

Questionnaire on the Use of Biologic Therapies in Managing Behçet's Disease by Internal Medicine Residents in National Hospitals in Morocco.

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Introduction: Behçet's disease (BD) is a multisystemic disorder characterized by vascular inflammation affecting arteries, veins, and capillaries. Pro-inflammatory cytokines and T lymphocytes play a crucial role in its pathophysiology, paving the way for therapeutic approaches such as inhibition of TNF, IL-1, or IL-6 signaling pathways, as well as T lymphocyte depletion, which can be targeted by biologic therapies. This study aims to evaluate the use of biologic therapies in the management of BD by internal medicine residents in Moroccan hospitals.

Method: Our study is based on the analysis of an online survey on the use of biologic therapies for managing Behçet's disease by internal medicine residents in national hospitals in Morocco.

Discussion: Our study included 31 responses from 35 residents, mainly aged 25 to 30 (66%) and mostly female (70%). Participants were from various medical faculties in Morocco, predominantly in the second and fifth years (25%). Most had no overseas training (70%). Findings revealed that 83% identified young age and the HLAB51 gene as Behçet's disease onset factors. All recognized recurrent genital aphthae's importance for diagnosis, while 95% deemed pathergy testing unnecessary. Regarding therapy, all emphasized biologic therapies, immunosuppressants, corticosteroids, and colchicine. Most (71%) felt informed about biologic therapy management, while 29% felt less informed. All recommended biologic therapy for severe or resistant cases. Additionally, 75% had prescribed infliximab and/or adalimumab, all affirming their efficacy. Finally, 91% felt capable of future biologic therapy prescribing.

Conclusion: This study emphasizes the growing role of biologic therapies in Behçet's disease treatment. Internal medicine residents exhibit solid understanding and confidence in prescribing these treatments, highlighting the evolving treatment options and the need for proper education. Biologic therapies offer promise for improving patients' lives and outcomes.

Behcet's Disease In Women Versus Men

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Introduction: Behçet's disease is a systemic vasculitis common in the Mediterranean basin, affecting mainly men; it is uncommon in women. The main objective of our work is to describe the epidemiological, clinical and therapeutic profile of this disease in women, in comparison with men.

Method: A retrospective and descriptive study, including all patients followed for Behçet's disease over a 5-year period [January 2019- January 2024], at the Internal Medicine Department B of the Military Hospital in Rabat. Patients were compared by gender.

Discussion: We included a total of 60 patients, 48 men (80%) and 12 women (20%), with a sex ratio of 4. The mean age was 30 ± 8 years for men and 33 ± 10 years for women. Our study shows the influence of gender on certain clinical manifestations of Behçet's disease. Articular, digestive and neuropsychic manifestations such as headaches and depressive syndrome are more frequent in women than in men, who present a more severe disease, with a higher frequency of ophthalmological and vascular involvement. In terms of treatment, almost all our patients were treated with colchicine (97%), while 80% required corticosteroid therapy. Anticoagulants were used in patients with thromboembolic manifestations (31% of patients). The use of immunosuppressants, mainly azathioprine, followed by methotrexate and cyclophosphamide, was noted in 40%, 5% and 5% of patients respectively. Biotherapy, notably anti-TNF, was prescribed in 10% of cases. In our series, the evolution was favorable, with lesion stabilization in 70% of cases, whereas it was unfavorable, marked by frequent relapses in 18% of cases. Two patients (men) with severe vascular damage died during the course of the disease. Five patients were lost to follow-up.

Conclusion: Behcet's disease remains a more serious vasculitis in men; The lower frequency of female Behcet's could be explained by the benignity of its forms.

Value of the pathergy test in Behcet's disease

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Introduction: The pathergy test is a non-specific hypersensitivity skin reaction induced by a needle stick, which is performed to look for evidence of this phenomenon.In addition to Behcet's disease BD, pathergy is also widely reported in a variety of other conditions, including neutrophilic dermatoses such as pyoderma gangrenosum PG and Sweet's syndrome. The pathergy test remains one of the most important diagnostic criteria for BD. Its performance therefore requires standardization of methodology and improved conditions to achieve the best possible sensitivity and sensibility.

Method: Our study was based on a questionnaire on the diagnostic value of the pathergy test in Behcet's disease in national hospitals in Morocco aimed at internal medicine residents in Morocco in order to gather perceptions and practices concerning this test.

Discussion: This study contained 48 responses from various internal medicine residents at different national university hospitals, mostly in the 25-30 age bracket. 50% of residents have been able to do it, and 75% have at least seen someone do it. All responders selected the doctor as the person to perform the procedure and assess the result after 48 hours in 68%. The test is performed intradermally on the forearm using a single 21 G blunt disposable needle, and in the majority of cases physiological saline is used. In 64%, the test is only performed if there is diagnostic doubt. 66.7% approve of the clinical correlation with test positivity. 48% find the test non-specific for BD. It may also be positive in PG,Wegener, Sweet syndrome.Interpretation of results may be affected by the use of corticosteroids and immunosuppressants.

Conclusion: BD is the only disease whose diagnostic criteria include a positive pathergy test.Our study revealed the diversity of ways in which the pathergy test can be used, which can influence the interpretation results. We note that, despite its ultimate interest in the positive diagnosis and evolution of BD, it is still little used in this context.

Neuro-Behcet's syndrome and cognitive impairment

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Introduction: Several earlier studies have reported that Neuro-Behcet's syndrome (NBS) patients, and even Behcet patients who do not meet NBS diagnostic criteria can suffer from cognitive impairment, which has a detrimental effect on quality of life and cannot be accurately predicted by disease duration. To evaluate the microstructural integrity of brain white matter tracts in patients with NBS and BS without neurological manifestations using diffusion tensor imaging (DTI) and to investigate potential utility of DTI as a surrogate biomarker of neurocognitive functioning by exploring possible correlations between DTI findings and neurocognitive function tests.

Method: This cross-sectional study comprised 34 NBS patients and 32 BS patients without neurological involvement. Cognitive functions, including attention, memory, verbal fluency, abstraction, executive control, visuospatial skills, and sensorimotor performance were assessed using standardized questionnaires. DTI data were analyzed using tract-based spatial statistics (TBSS) and automated probabilistic tractography to investigate inter-group differences. Subsequently, correlations between tensor-derived parameters of white matter tracts and neurocognitive test scores were examined.

Discussion: DTI revealed decreased fractional anisotropy and increased radial diffusivity, mean diffusivity, and axial diffusivity in both supratentorial and infratentorial white matter in NBS patients, indicating widespread loss of microstructural integrity. Moreover, this loss of integrity was also observed in BS patients without neurological manifestations, albeit to a more limited extent. In NBS patients, certain white matter tracts, including cingulum bundle and fornix, were associated with poor cognitive performance across multiple domains.

Conclusion: DTI findings might have a potential utility as a neuroimaging marker to predict the extent of neurocognitive impairment associated with central nervous system involvement in BS.

Symptom clusters in Behçet's disease: A case series

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Introduction: Herein, we report cases of central nervous system involvement of Behçet's disease (BD) and vascular involvement of BD respectively, underlying gastrointestinal involved state.

Method: -

Discussion: First, a 44-year-old man was diagnosed with BD (incomplete type according to Japanese criteria; oral ulcers, genital ulcers, and skin lesions) in 1998 at the age of 19. Intestinal involvement was first confirmed through colonoscopy in 2005, and subsequently he was administered Humira (adalimumab), however he got right colectomy including ileal resection in 2020, as the disease was uncontrollable. Later, in 2023, the patient complained of headaches accompanied by slurred speech, fecal and urinary incontinence, and even suffered seizure. Brain magnetic resonance imaging (MRI) revealed 5.5 x 4 cm-sized brain abscess in the Lt. frontal lobe, which was drained by catheter insertion. At last, the abscess found to be sterile. Second, a 40-year-old man was diagnosed with BD (incomplete type according to Japanese criteria; oral ulcers, genital ulcers, and skin lesions) in 2009 at the age of 26. And he got right hemicolectomy in 2015 due to the aggravation since intestinal involvement confirmation in 2009. Then in 2016, aortic aneurysm had been found, and graft interposition was performed. After that, he started psychiatric consultation because of the adjustment disorder with neurotic depression.

Conclusion: Previously, symptoms of BD were considered independent. However, there have been some reports that demographic and genetic features may be related to different clinical charteristics. And recently, research have been conducted grouping the symptoms of BD and approach them as a cluster. With this case reports, neurologic and vascular symptoms developed in gastrointestinal BD patients, we conclude that clustering symptoms could be an important clinical feature of BD and may have pathogenetic and therapeutic implications.

Two cases of exceptional association between Behçet's disease and Takayasu vasculitis

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Introduction: Behçet's disease is characterized by ophthalmic,cutaneous, and vascular involvement,while Takayasu's arteritis is a chronic inflammatory disease of the large elastic arteries.The association of these two pathologies is exceptionally rare.

Method: Association between Behçet's arteritis and Takayasu's was determined due to the presence of criteria for both

Discussion: case 1:A 53-year-old male, Behçet's disease since 2018 with bipolar aphthosis,pseudofolliculitis,and a right carotid aneurysm.Treated by carotid-carotid bypass,corticosteroid,cyclophosphamide followed by azathioprine. In 2022,he presented intermittent claudication of the left limb, blood pressure asymmetry, a left carotid murmur,and absence of radial pulse.sedimentation rate of 53.Imaging revealed left carotid stenosis,total thrombosis of the carotid-carotid bypass,occlusion of the left subclavian artery, and pre-occlusive stenosis of the left internal carotid artery. The diagnosis of Behçet's disease associated with Takayasu's disease was confirmed. He received corticosteroid and anti-TNF. Case2:A 42-year-old female with oral aphthosis, followed since 2015 for Takayasu's disease with intermittent claudication,decreased radial pulse of the left arm,blood pressure asymmetry,tight stenosis at the left subclavian and vertebral arteries, partially thrombosed aneurysm of the right renal artery and of the abdominal aorta,In 2021 she experienced an acute decrease in visual acuity due to sectorial papilledema,an abdominal aortic murmur and absent of left arm pulses.imagery revealed same lesions and thrombosis of the left subclavian artery, Due to the oral aphthosis,the topography of the arterial lesions and ocular involvement,Takayasu's disease associated with Behçet's disease was confirmed.She received corticosteroid,cyclophosphamide followed by methotrexate.

Conclusion: Vascular involvement in patients with Behçet's disease is not necessarily vasculo- behçet's disease, but may be Takayasu's arteritis, hence the need for clinical analysis.

Clinical features of Behçet's disease in Jordan

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Introduction: Behçet's disease has a high prevalence in countries along the ancient Silk Road, and the number of healthy individuals carrying the disease susceptibility gene is also high in that region. In this study, we investigated the clinical features of Behçet's disease and HLA-B51 gene frequencies in Jordan.

Method: Clinical presentation and treatment were examined retrospectively based on medical records. Saliva samples were collected from the subjects and analyzed for HLA-B51- specific SNP rs1050502 C allele frequency.

Discussion: Forty-seven patients with Behçet's disease (33 men, 14 women, average 41.9 years old) who visited King Abdullah University Hospital, and its affiliated hospital from 2019 to 2023, and forty-five healthy controls were enrolled. Oral aphthous ulcers were reported in 100%, skin lesions in 88.9%, genital ulcers in 84.4%, and ocular lesions in 71.1%. Arthritis, gastrointestinal lesions, vascular lesions, CNS (central nervous system) involvement, and epididymitis were observed in 17.8%, 9.3%, 46.2%, 29.5%, and 40.7% of patients, respectively. Anterior uveitis was present in 66.7% and posterior segment inflammation in 64.4%. The corrected visual acuity was 0.8 or better in 69.4% of patients, but 0.1 or lower in 5.6%. Treatment consisted of corticosteroid periocular injections in 26.7%, systemic corticosteroids in 91.5%, colchicine in 88.9%, immunosuppressants in 83.0%, and TNF (tumor necrosis factor) inhibitors in 29.7%. The HLA-B51 prevalence in homozygotes or heterozygotes was 70.2% in the patient group and 15.6% in the healthy group, and the rs1050502 C allele frequency was 39.4% in the patient group and 7.8% in the healthy group, all of which were significantly more frequent in the patient group.

Conclusion: The clinical features of Behçet's disease in Jordan were similar to those in Japan and other Silk Road countries, with many HLA-B51 carriers.

Long-term Results of Interferon-alpha Therapy in Patients with Behçet's Uveitis at a Tertiary Referral Center in Turkey

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Introduction: Behçet's uveitis (BU) treatment aims to swiftly suppress inflammation, prevent recurrent vasculitis, and avoid vision-threatening complications. Conventional therapies may fall short. 2018 EULAR guidelines recommend high-dose steroids, infliximab, or IFN-α for ocular attacks. IFN-α offers rapid action and sustained remission but faces discontinuation in Turkey post-2020. This study aims to present the long-term outcomes of IFN-α therapy recipients, covering complete or partial remission and non-responders.

Method: We analyzed the medical records of 45 BU patients treated with IFN-α (Jan 2014–Mar 2020). Initial IFN-α dose: 6 MU/day in 34 (75.5%) and 3 MU/day in 11 (24.5%) patients. Patients are categorized by treatment response. Demographics, BCVA, remission status, and long-term outcomes were assessed.

Discussion: In 37 patients (82.2%), ocular involvement was bilateral; in eight patients (17.8%), unilateral. Sixty-six patients (80%) were male, nine (20%) female. Mean age: 33.24 ± 7.51 years, BU duration: 56.27 ± 43.39 months, mean uveitis attacks: 3.03 ± 1.24. Fellow eye BCVA improved significantly from 1.38 ± 0.98 to 0.98 ± 1.10 logMAR ($p<0.001$). Similarly, better-seeing eye BCVA improved from 0.35 ± 0.68 to 0.16 ± 0.32 logMAR ($p=0.002$). 10 patients (22.2%) started IFN-α therapy without prior immunosuppressive treatment. 21 patients (46.7%) required anti-TNF therapy during treatment. Switching reasons: side effects (3), inadequate remission (9), new attacks (9). Among those transitioning, 52.4% received Adalimumab, 28.6% Infliximab, and 19.0% alternated. Additionally, 26.7% on Adalimumab needed weekly dosing. 2 non- responders transitioned to tocilizumab. Long-term complete remission rate with IFN- α: 53.3%, partial remission: 33.3%. No significant post-treatment BCVA difference among groups (ANOVA, $p=0.523$ and $p=0.809$).

Conclusion: 86.6% of Turkish patients with BU had a partial or complete response to IFN-α therapy. This shows that IFN-α is still safe and effective for treating BU.

Uveitis associated with Behçet's disease in children: a case series

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Introduction: To report a series of uveitis associated with Behçet's disease in children.

Method: A retrospective case series including children with uveitis associated with Behçet's disease. Demographic, ophthalmological data at presentation and during follow-up were collected and analyzed.

Discussion: Six patients (11 eyes) were included in this study. The rate of pediatric Behçet disease with ocular involvement among all cases of Behçet's disease with ocular involvement was 6%. Mean age at diagnosis was 11.5 years (range, 7-16). Four patients were male and 2 patients were female. Ocular involvement was bilateral in 5 patients and unilateral in one patient. Mean best corrected visual acuity (BCVA) at presentation was 1 logMar (range, 2.6-0 logMar). Ocular findings included non- granulomatous anterior uveitis with vitritis in all eyes, hypopyon in one eye, non- occlusive retinal vasculitis in 6 eyes, occlusive vasculitis in one eye, retinal infiltrates lesions in one eye, papillitis in 6 eyes, and macular edema in 3 eyes. Systemic manifestations included oral aphthosis in 4 patients, genital aphthosis in one patient, and pseudofolliculitis in one patient. The ocular involvement was the presenting feature of Behçet disease in all cases. Treatment modalities included systemic corticosteroids, conventional immunosupressive drugs and biotherapy in all patients. Mean follow up was 4.5 years (range, 3-6 years). Final BCVA was 0.6 logMar (range, 2.6 and 0 logMar).

Conclusion: Behçet's disease is a rare cause of uveitis in children. Ocular manifestations and systemic associations are comparable to those encountered in adult patients with Behçet disease. Aggressive and long-term treatment is mandatory to prevent vision loss.

The Thrombosis Chronicles: A journey through 10 cases of Behçet disease

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Introduction: Behçet's disease is characterized by its vascular involvement, which may include venous thrombosis, aneurysms, and other complications related to vessel inflammation.

Method: A retrospective descriptive study was conducted on 10 patient records presenting thrombosis within the context of Behçet's disease among 184 patient records followed up in over a 12-month period from 03/2023 to 03/2024,

Discussion: We examined the records of 184 patients. We identified 10 patients followed for thrombosis due to Behçet's disease: 6 women and 4 men with a sex ratio of 0.67 (6F/4M). The average age was 32.6 years, ranging from 27 to 49 years. One patient (12.5%) was hypertensive, 2 patients (25%) had fractures, 2 female patients (25%) were on oral contraception, 1 female patient (12.5%) had a history of miscarriage, 1 patient (12.5%) was a chronic smoker, and 1 female patient (12.5%) had a family history of thromboembolic disease. The clinical manifestations at diagnosis were painful swelling of the leg in 5 cases (50.4%), chest pain in 2 cases (20%), and acute abdominal pain in 2 cases (20%). One case (10%) presented dyspnea. Concerning the localization of thrombosis: 6 patients (60%) had unusual thrombosis including cerebral thrombosis, 2 portal thromboses, 1 internal jugular thrombosis, 1 external jugular thrombosis, and 1 splenomesenteric thrombosis. Two cases involved superficial venous thrombosis (internal and external saphenous veins) and 2 cases involved deep venous thrombosis (iliofemoral thrombosis). The diagnosis of Behçet's disease revealed by venous thrombosis was established based on the presence of bipolar aphthosis in 10 patients, pseudofolliculitis in 8 patients, episcleritis in 4 patients, and uveitis with synechiae in 6 patients. One patient had a seizure episode.

Conclusion: Early recognition of venous thrombosis revealing behcet's disease can allow for appropriate management

Regional Register of Behçet Disease in northern Morocco

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Introduction: Behçet's disease is a chronic multisystem inflammatory vasculitis, which affects arteries and veins of all sizes. The epidemiology of this disease varies greatly depending on the geographical location and the ethnic group and registers are mandatory.

Method: The creation of our registry was made by including all patients followed for Behçet's disease in Tangier University Hospital for a period of 5 years extending from April 2019 to April 2024

Discussion: The total number of patients was 108, with a median age of 34 years and a male predominance. Oral aphthosis was present in 86% of cases, genital aphthosis in 54% of cases. Ocular damage was present in 53% of cases, mostly in the form of bilateral panuveitis. Blindness as a complication was found in 13 patients. Vascular damage was present in 53% of cases, predominated by deep vein damage in 72% of cases. Pulmonary embolism was present in 4 patients. Arterial damage was present in 29% of cases mostly an aneurysm of the pulmonary arteries. Cardiac involvement was present in 06 patients, with an intracardiac thrombus in 4 patients and myocarditis in 2 patients. Neurological damage was present in 20.4%. The manifestations found are a pyramidal syndrome in 59%, a damage to the cranial pairs in 45%, lymphocytic meningitis in 41% and encephalitis in 14%. For therapeutic management, 71% were on colchicine, 72% on oral corticosteroids and 50% on immunosuppressive treatments. 41% received curative anticoagulation. For the evolution of the patients, 42% had a partial remission, 24% were in relapse and15% had complete remission.There were 3 deaths, 2 from myocarditis and 1 from ruptured pulmonary aneurysm.

Conclusion: Our experience during data collection for the Regional Behcet Disease Registry encouraged us to seek a better collaboration between the different services that support Behçet's disease, which will in turn help us to peserve patients vital and functional prognosis.

Eye and Behçet's Disease: Clinical and Prognostic Features Regarding 291 Cases

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Introduction: Behçet's Disease (BD) is a vasculitis of unknown origin affecting vessels of all sizes. It features recurrent oral and genital ulcers, with ocular involvement being the most common and critical for visual prognosis. This study aims to determine the clinical and prognostic characteristics of ocular involvement in BD.

Method: This monocentric retrospective study, conducted from January 2006 to December 2023 in our internal medicine department, involved 563 BD patients. All patients met the 2013 international classification criteria for BD and underwent systematic ophthalmological examinations.

Discussion: Results: Ocular involvement in Behçet's Disease was observed in 291 cases (51.6%). The average age of affected patients was 29.5 years, with a strong male predominance (70.8%). Ocular involvement was unilateral in 55.6% of cases, revealed BD in 62.42%, and was inaugural in 89 patients (30.58%). The average diagnostic delay was 18 months. Anterior uveitis was observed in 48 patients (16.49%), with hypopyon uveitis in 4 patients (8.33%). Episcleritis was noted in 1% of cases. Intermediate uveitis occurred in 43 patients (14.78%). Posterior uveitis was the most frequent, affecting 109 patients (37.46%), with retinal vasculitis in 43.12%, macular involvement in 28.44%, retinal detachment in 15.6%, papillitis in 6.42%, NORB in 2.75%, and intra-vitreous hemorrhage in 3.67%. Panuveitis was present in 88 patients (30.24%). Therapeutically, 64.5% of patients received corticosteroids with immunosuppressants, and biological treatment was initiated in 6% of cases. Outcomes were good in 52.9% of cases, 12.7% relapsed, and 16.8% experienced blindness.

Conclusion: The ocular involvement of BD is polymorphic, affecting different segments of the eye with a predominance of posterior uveitis and panuveitis. It is severe and can be a cause of irreversible blindness in cases of diagnostic and therapeutic delay, hence the importance of systematic screening by an ophthalmological examination in all patients with BD.

Identification of Oral Ulcer Patterns in Behçet's Syndrome through K-Means Cluster Analysis and Correspondence Analysis

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Introduction: The aim of the study was to identify oral ulcer activity (OUA) patterns using K-Means Cluster Analysis and Correspondence Analysis to develop disease management strategies for Behçet's Syndrome (BS).

Method: In this cross-sectional study, 526 patients with BS were included from two tertiary centres in Turkey and United Kingdom. Disease severity scores reflecting organ involvement were calculated. Patients were then grouped according to organ involvement (mucocutaneous and musculoskeletal vs. major organ involvement). Patients with active oral ulcers (n=306) were classified into four clusters based on disease severity score and the number of oral ulcers through K-Means Analysis. Correspondence Analyses (CA) were then used to visualize associations described by categorical variables regarding these clusters, disease course, medications (non- immunosuppresive (Non-IS) and IS), gender and regular or irregular tooth brushing habits (RTB vs. ITB).

Discussion: K-Means Analysis identified four clusters with homogeneous clinical profiles in the group. The numbers of oral ulcers and severity scores were: • 2.18±1.13 and 4.28±1,27 in the Low OUA (65,03%), • 2.19±1.37 and 7.98±1.38 in the Low OUA with Major Organ Involvement (19,25%), • 7.60±1.88 and 4.77±1.47 in the Moderate OUA Group (9,8%) • 14.91±2.34 and 5.27±1.73 in the High OUA Group (3,59%) ($p<0.001$). In CA, the Low OUA (19,3 % for ocular involvement, 20,8% for vascular involvement), the Low OUA with Major Organ Involvement (87,5%, 39,7%, respectively) and the Moderate OUA (43,3%, 10%) represented male patients treated with IS and having ITB habits.

Conclusion: Since RTB decreases the bacterial load in oral environment, ITB could be considered a risk factor for disease management in males, especially treated with ISs. Therefore, patient empowerment strategies covering oral hygiene could be helpful for the disease management in BS.

Behçet without oral aphthosis: interest of new criteria

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Introduction: Behçet's disease (BD) is a vasculitis of vessels of all calibers, affecting both arterial and venous territories. Diagnosis is essentially clinical. Mucosal lesions are thought to appear at an early stage of the disease. Oral aphthosis has been reported as the most frequent manifestation in the majority of cases. However, studies of large series of BD patients indicate that 1-8% of patients do not develop oral aphthosis throughout the course of the disease. This study was designed to investigate the prevalence and demographic and clinical parameters of BD cases without oral aphthosis.

Method: Monocentric and retrospective study carried out in the internal medicine department of the CHU ibn rochd in casablanca over a period of 18years involving 563 patients with BD retained according to the new criteria proposed in 2006 and revised in 2013.

Discussion: Results: Of the 563 patients with BD 1.6% had no oral aphthosis. The sex ratio was 3.5 M/F. The mean age of onset was 28.3 years. Initial manifestations included genital aphthosis in 77.7%, ocular involvement 11.1%, and vascular involvement 11.1%. During the course of the disease, 66.6% of patients had pseudofolliculitis lesions, and 22.2% had a positive pathergy test. Joint involvement was present in 1/3 of patients. 6 patients had ocular involvement, and vascular involvement was present in 66.6% of cases. Pleuropulmonary and central neurological parenchymal involvement were observed in only 1 patient. Therapeutic measures included bolus and oral corticosteroids, azathioprine and cyclophosphamide, in addition to colchicine. Progression was good in 66.66% of patients, with therapeutic resistance in 11.1%, and the onset of AA amyloidosis in 1 patient.

Conclusion: BD patients without oral aphthosis constitute a subgroup in whom the 1990 international criteria posed a diagnostic problem by requiring the presence of oral aphthosis. The new criteria have a sensitivity clearly superior to the old ones, while retaining a reasonable specificity.

Epidemiological and clinical characteristics of Moroccan patients with Behçet's disease: A retrospective study.

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Introduction: Behcet's disease (BD) is a systemic vasculitis characterized by oral and/or genital aphthosis with or without any systemic manifestations. The prognosis depends on the clinical form of the disease. The aim of our work is to describe the epidemiological and clinical characteristics of Moroccan patients with Behçet's disease.

Method: This monocentric retrospective study conducted in the internal medicine department of CHU Ibn Rochd of Casablanca covered 1571 cases hospitalized for MB over 42 years (1981-2023), based on patients' medical records.

Discussion: One thousand five hundred and seventy-one patients meeting international criteria were selected, divided into 1108 men and 463 women, with a sex ratio of 2.39. The mean age at onset was 26.29 years and 33.46 years at diagnosis, with the most affected age group being between 30 and 50 years. The frequencies of clinical manifestations were as follows: oral aphthosis (99.61%), genital aphthosis (83.92%), pseudofolliculitis (59.37%), dermohypodermal nodules (15.43%), pathergic test positivity (40.24%), joint involvement (42.72%), eye involvement (60.39%), vascular involvement (23.85%), neurological involvement (18.36%), digestive involvement (9.18%), and juvenile Behçet (5.73%).

Conclusion: In this retrospective study, we examined the epidemiological and clinical characteristics of Behçet's disease in Moroccan patients. Our findings confirm the predominance of the disease in males and highlight a varied clinical presentation with a predominance of mucocutaneous manifestations. The data also suggest that the average age of disease onset is consistent with the literature. However, vascular, neurological, and digestive involvement were less frequent in our series.

The particularities of angio-behçet: a review of 160 cases

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Introduction: Vascular involvement in Behcet's disease is severe, occurring in almost 40% of cases, and all vessels, whatever their type (arterial or venous), size or location, may be affected. It is considered to be the main form of life-threatening disease, requiring rapid and aggressive treatment.

Method: out of 563 cases of MB,160 patients with vascular involvement were included and analysed between January 2006 and December 2023, in the internal medicine department of the ibn Rochd university hospital in Casablanca, excluding cardiovascular involvement.

Discussion: results:Of the 563 patients with Behçet's disease, 160 had vascular involvement (28.4%). Vascular involvement was the presenting feature of the disease in 70% of cases. The average age at onset of vascular manifestations was 35.5 years, with a male predilection. Venous manifestations were observed in 95% of cases. Deep vein thrombosis of the lower limbs was the most frequent in 66% of cases. Arterial involvement was present in 28.1% of patients, dominated by pulmonary artery aneurysms (11.25%). Arterial thrombosis was less common (1.25%). Treatment: All patients received oral corticosteroids/ bolus methylprednisolone 500mg-1g/d for three days, immunosuppressive therapy, anticoagulation in 87.5% of cases, and biotherapy (anti-TNF alpha) in 5% of cases. Progression was favourable in 38.7% of cases. Recurrence in 7.5% of cases, and 2 deaths complicating pulmonary aneurysms.

Conclusion: Vascular involvement in BD is frequent. Their inclusion in the criteria for classification of the disease may be proposed in view of the particular clinical aspects they present. They are serious and potentially life-threatening, and therefore require rapid diagnosis and intensive specific treatment, the only way to improve their prognosis. A better understanding of patients at risk will also enable them to be better cared for.

Neuroleptic malignant syndrome simulating a neurological relapse of behcet's disease: a case report

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Introduction: Neuroleptic malignant syndrome is a rare neuropsychiatric syndrome associated with the administration of an antipsychotic drug, characterised by hyperthermia, muscular rigidity, autonomic dysfunction and altered consciousness

Method: We report the observation of a patient treated for neurobehcet in whom neuroleptic malignant syndrome simulated a neurological relapse of behcet

Discussion: results: This 42-year-old patient had been treated for 7 months for Behçet's disease with cutaneous-mucosal, parenchymatous and psychiatric neurological involvement. His neurological involvement consisted of febrile meningoencephalitis, bilateral pyramidal syndrome, pseudobulbar syndrome, cerebellar involvement and sphincter disorders evolving in a depressive context. Treatment: the patient received 4 boluses of methylprednisolone 1g IV monthly, 6 boluses of cyclophosphamide IV monthly, switched to azathioprine and neuroleptic (Olanzapine) with very good clinical improvement. 1 month after introduction of olanzapine, the patient was readmitted for febrile confusion, motor deficit in the lower limbs, muscular rigidity, laboratory work-up: CPK 4267 IU/L,negative infectious work-up. Fearing a neurological relapse of the behcet's, a cerebro-medullary MRI was performed, which revealed only sequelae. Biological check-up two days later: CPK 11322 IU/L,LDH 1128 IU/L, ASAT: 164 IU/L:L ALAT normal, and K+= 8.2 mmol/L suggestive of severe rhabdomyolysis. The diagnosis of neuroleptic malignant syndrome on olanzapine was accepted. Treatment was stopped and a very good clinico-biological improvement was observed over a few days: (CPK at 200IU/L vs. 11322, LDH at 380 vs. 1128u/L, and kalaemia at 3.8mmol/L vs 8.2)

Conclusion: The incidence of neuroleptic malignant syndrome is currently estimated at 0.02% of patients undergoing neuroleptic treatment. Despite a better understanding of this syndrome, the mortality rate remains between 10 and 20%, and is probably higher if it occurs in patients with vasculitis such as Behcet's disease.

The articular manifestations of Behçet's disease

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Introduction: Behçet's disease is a systemic vasculitis described in 1937 by Behçet. The etiology of Behçet's disease remains unknown, and it can take on various manifestations, including cutaneous and mucosal, articular, ocular, neurological and vascular. Diagnosis is based on a range of clinical arguments. Our work analyses the articular manifestations of Behçet's disease.

Method: A Monocentric retrospective study conducted over 17 years, from 2006 to 2023, in the internal medicine department of the Ibn Rochd University Hospital,135 patients with articular manifestations were found in a series of 563 cases.

Discussion: The mean age of patients was 32.4 years, with a M/F ratio of 2. Inflammatory arthralgias were the most frequent (72.3%), followed respectively by oligoarthritis, polyarthritis and monoarthritis (15.1%; 13.9%; 7.8%). According to our results, the preferred site of joint involvement was the lower limb, and more specifically the knees and ankles (61%). The course of the disease was intermittent in 71% of cases, acute in 17% and chronic in 12%. We noted the following particular forms: 9 cases of deforming polyarthritis, 6 of which had progressed to joint destruction, 5 cases of pseudogout, 3 cases of popliteal cyst, 15 cases of spondyloarthropathy, 6 cases of isolated sacroiliitis and 1 case of arthritis simulating phlebitis. Treatment was based on colchicine, NSAIDs, low-dose corticosteroids and azathioprine in certain forms resistant to the usual treatment. Progression was favourable in the majority of cases.

Conclusion: According tothe literature, articlular manifestations in Behçet's disease is frequent. It is not uncommon for it to inaugurate the disease and precede aphthosis by several years. Peripheral joint involvement is the most common, ranging from simple inflammatory arthralgias to full-blown arthritis; however, involvement of the hands and feet is much less common.Progression under symptomatic treatment is generally favourable, and joint deformity; destruction are very rare.

Mucocutaneous manifestations of Behçet's disease

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Introduction: Behçet's disease is a complex multi-system disorder of unknown etiology characteristically presenting with recurrent oral ulcers. It is presumed to be a vasculitis and includes involvement of the ocular, cardiovascular, renal, gastrointestinal, pulmonary, musculoskeletal, and central nervous systems. Mucocutaneous lesions are the hallmarks of Behçet's disease.This study describes the main mucosal skin manifestations in a North African population.

Method: This is a retrospective study of cases of BD identified according to the international diagnostic criteria of the International Study Group, conducted over a 17-year period from 2006 to 2023 in an internal medicine department.

Discussion: Our study included 563 patients with Behcet's disease, including 399 men (70,9%) and 164 women (29,1%) (Sex ratio M/F: 2,4). The age of appearance was different; patients aged between 15-24 years old were affected the most. The clinical manifestations were dominated by mucosal skin disease with oral aphtosis in 98,4% and genital aphtosis in 80,3% of cases. Mucosal giant aphthae was present in 15 patients (4,8%), meanwhile genital giant aphthae was present in 33 patients (14,2%). Pseudofolliculitis was observed in 291 (75,8) of cases, erythema nodosum in 86 patients (22,4%), and pathergy test was positive in 91 of cases (23,7%). Other skin manifestations were observed in our patients: skin aphthae in 30 patients (7,8%), anal aphthae in 8 cases (2,1%). Minor ulcers healed without scarring in 5-15 days whereas major ulcers are more painful and healed within 2-8 weeks. Most of our patients responded well to colchicine and short-term oral corticosteroids. Penicillin was introduced in 4,2% cases. Immunosuppressive therapy was required in 0,2% of cases.

Conclusion: Behçet's disease presents diverse mucocutaneous symptoms preceding severe complications. Diagnostic criteria rely heavily on these manifestations, affirming the need for prompt recognition to prevent organ damage through appropriate management strategies.

Digestive Manifestations in Behçet's Disease

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Introduction: Behçet's disease is a vasculitis affecting vessels of all calibers, impacting both arterial and venous territories. It predominantly affects young adults and is a systemic vasculitis with a wide range of clinical manifestations, including rare but significant gastric involvement that can greatly impact patients' quality of life and prognosis. The most frequent gastrointestinal localization is the colon and ileocecal region.

Method: This is a retrospective monocentric study conducted in the internal medicine department over 17 years, from January 2006 to December 2023. We collected 563 patient files who were followed for Behçet's disease, diagnosed according to the International Study Group for Behçet's Disease criteria.

Discussion: Digestive involvement was diagnosed in 33 patients, representing 17% of cases. This included 13 women and 20 men with an average age at the time of digestive involvement diagnosis of 32.9 years. Minor digestive manifestations were observed in 23 cases, encompassing abdominal pain in all patients, transit disorders such as diarrhea and constipation, and nausea and vomiting in 12 cases. Major manifestations were dominated by rectal bleeding in 4 patients and pseudo-surgical abdomen in 3 patients. All patients underwent endoscopic fibro-colonoscopy with biopsies, echo-doppler, and abdominal CT angiography. Two patients had pseudomembranous colitis (5.9%), one had ulcerative colitis, and another had diffuse erythematous colitis. All patients received high-dose corticosteroid therapy, cyclophosphamide was used in 6 patients (35.3%), and anti-TNF therapy was used in one patient. Most patients had a favorable outcome.

Conclusion: Gastrointestinal manifestations macroscopically resemble lesions of Crohn's disease and, ulcerative colitis, posing diagnostic challenges.They occur in 40% of cases. It can lead to hemorrhages or perforations. Its presence should prompt the search for another disease besides Behçet's disease, particularly chronic inflammatory bowel disease.

Pulmonary Artery Aneurysms in Behcet Syndrome: A Case Series

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Introduction: Although pulmonary artery aneurysms (PAA) are considered rare (less than 5%), there are quasi-pathognomonic of Behçet syndrome (BS) and play a major role in patient's morbidity and long-term mortality. Our work aims to depict the clinical profile, imaging features, management, and outcomes of BS patients presenting with PAA.

Method: We conducted a retrospective, descriptive and monocentric study, between 2000 and 2022, including all BS patients followed in our department with documented PAA.

Discussion: 51/550 patients had PAA (10%). The mean age was 36 ± 11 [20-69] and the sex ratio (M/F) was 4.1:1. Cardiovascular risk factors comprised smoking (13%) and high blood pressure (10%). PAA occurred after a median of 6 years following BS onset. Patients were mostly asymptomatic (68%) or presented with hemoptysis (8%), cough (8%), chest pain (5%), and dyspnea (5%). PAA were often multiple (78%), proximal (46%), and bilateral (29%). Concomitant arterial manifestations were associated in 25% of the patients, consisting of peripheral artery aneurysms (17%), pulmonary artery thrombosis (10%), cerebral artery thrombosis (4%), and aortic aneurysms (2%). Deep venous thrombosis was found in 21% of the cases, in the superior vena cava (10%), inferior vena cava (9.6%), cerebral veins (7.7%), and lower extremities (6%). Right intracardiac thrombosis was visualized (17%). Extravascular involvements were mainly mucocutaneous (73%), ocular (46%), articular (29%), and neurological (10%). Patients were treated with glucocorticoids (100%), cyclophosphamide (89%), azathioprine (89%), Infliximab (3%), and colchicine (100%). Only one patient underwent surgery. Relapse and death were recorded in respectively 15% and 10% of the cases.

Conclusion: BS should be evoked in presence of PAA, especially in young male patients presenting with other vascular lesions and intracardiac thrombosis. Prompt initiation of immunosuppressants is of paramount importance to reduce the risk of massive hemorrhage from PAA's rupture.

Drug Survival of Adalimumab in 335 Patients with Behçet Syndrome

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Introduction: There are no controlled trials with adalimumab (ADA) in patients with Behçet syndrome (BS). Drug survival is an important outcome that indicates efficacy and safety in observational studies.

Method: We conducted a chart review of BS patients who received ADA between May 2008 and January 2023. Demographic features, reasons for ADA use, ADA duration, and reasons for discontinuation were recorded.

Discussion: 335 patients (211 men (63%); mean age: 29.2±9 years) were treated with ADA. ADA was prescribed for uveitis in 129 patients (38%), vascular involvement in 81 (24%), arthritis in 60 (18%), mucocutaneous involvement in 47 (14%), gastrointestinal (GI) involvement in 10, and parenchymal central nervous system (NBS) in 8 patients. In 30 patients, ADA treatment had to be intensified by shortening the dose interval to once a week in order to achieve remission. The main reason for this was uveitis in 21 patients. 198 (59%) were still using ADA during a mean (SD) follow-up of 40 ± 23 moths. The drug survival rate was 56% for uveitis, 65% for vascular involvement, 55% for arthritis, 64% for mucocutaneous involvement, 50% for GI involvement, and 62% for NBS. 137 (41%) patients had discontinued ADA after a median follow-up of 20.5 months (IQR: 6-26.5). Reasons for discontinuation were inefficacy in 59 (43%) (primary inefficacy in 9 and secondary inefficacy in 50), adverse events in 23 (18%), remission in 19 (14%) patients, and lack of patient compliance in 23 (17%). Other reasons were preparation for surgical operation (n=2), pregnancy/willing to get pregnancy (n=5), lack of health insurance (n=2), work-up for malignancy (n=2) and death (n=2). Among the 30 patients who received ADA at a weekly dose, 12 discontinued the treatment due to inefficacy and 1 due to tuberculosis.

Conclusion: ADA survival rate was suboptimal. Loss of efficacy over time and adverse events were the main reasons for ADA discontinuation.

Thoracic manifestations of Behçet's disease excluding cardiac involvement

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Introduction: Behçet disease is a systemic vasculitis with venous tropism.Thoracic involvement is relatively uncommon and includes pulmonary aneurysms,pulmonary embolism,superior vena cava thrombosis,alveolar haemorrhage,mediastinal fibrosis, parenchymal involvement and pleurisy.

Method: A monocentric retrospective study over 17 years, from 2006 to 2023, in the internal medicine department of the Ibn Rochd University Hospital, 58 patients with thoracic manifestations excluding cardiac involvement were found in a series of 563 cases.

Discussion: Our series included 39 men and 19 women, with a M/F ratio of 2. The average age of patients was 35.3 years.Respiratory symptoms were dominated by haemoptysis (44.8%), followed respectively by exertional dyspnoea associated with chest pain, superior cava syndrome and at last dry cough (22.4%; 18.9%; 13.7%). Thoracic angioscanner revealed superior vena cava thrombosis in 9 cases, ; aneurysms in 21 cases and 6 of them were thrombosed; pulmonary embolism in 22 cases; lung parenchymal involvement in 3 cases; 3 cases of pulmonary vasculitis. Patients were treated with corticosteroids and immunosuppressants, with the addition of anticoagulants for patients with superior venous thrombosis and those with pulmonary embolism and with surgical treatment for certain aneurysms. The disease progressed well with appropriate medical treatment, while the main cause of death was massive haemoptysis secondary to ruptured pulmonary aneurysms.

Conclusion: Thoracic manifestations of Behçet's disease are dominated by vascular involvement,on which the vital prognosis depends. However, other less common conditions may occur during the course of the disease, notably pleuropulmonary involvement and mediastinal fibrosis. The treatment of the thoracic manifestations is not well codified. Corticosteroids and/or immunosuppressants are the treatment currently used.

Cardiac Involvement in Behçet Syndrome: A Case Series

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Introduction: Cardiac involvement (CI) in Behçet syndrome (BS) is rarely described and associated with high morbidity and mortality. Our study aims to describe the profile and outcomes of BS patients presenting with cardiac lesions.

Method: This descriptive and single-center study was retrospectively carried out between 2000 and 2022, including all BS patients admitted to our department and presenting with CI.

Discussion: 35/550 cases were enrolled (6%). The sex ratio (M/F) was 3:1 and the mean age was 30 ± 7 years (21-48). Cardiovascular risk factors comprised smoking (21%), high blood pressure (15%), and diabetes (11%). CI was mostly asymptomatic (71%), or revealed by chest pain (21%), dyspnea (19%), dry cough (18%), and hemoptysis (17%). Inflammatory parameters were elevated with a median ESR of 46 mm/h and CRP of 55 mg/l, and normal hemostasis parameters. The lesions visualized were right intracardiac thrombosis (78%), pericarditis (38%), coronary thrombosis (13%), myocarditis (7%), and endomyocardial fibrosis (3%). Concomitant vascular involvement was frequent (75%), especially thrombosis in the inferior vena cava (23%), pulmonary arteries (21%), lower extremities (15%), cerebral veins (18%), and superior vena cava (9%). Arterial aneurysms were also common, mostly of the pulmonary arteries (25%). Noncardiovascular involvements were mucocutaneous (78%), articular (28%), ocular (43%), neurological (18%), and digestive (3%). Patients were treated with colchicine (96%), glucocorticoids (96%), anticoagulants (54%), cyclophosphamide (57%), azathioprine (46%), and infliximab (3.6%). Surgery was performed in 6% of the cases. One relapse of intracardiac thrombosis was recorded. Death was documented in five cases.

Conclusion: CI is a serious manifestation of BS. Although rare, it is also fairly underdiagnosed due to its nonspecific features and discrete evolution. Hence, better knowledge of the disease is crucial for early diagnosis and prompt initiation of immunosuppressive therapy.

Withdrawal of Colchicine in Behçet Syndrome

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Introduction: Previous randomized controlled studies reported conflicting results regarding the effectiveness of colchicine on oral ulcers in Behçet syndrome (BS). A good study scheme for assessment of efficacy is a randomized drug discontinuation design. We aimed to assess mucocutaneous disease activity after 12 weeks in BS patients who discontinued colchicine, in a randomized, controlled, single-blind study (NCT06146192).

Method: BS patients with mucocutaneous and joint disease but no major organ involvement who fulfilled ISG criteria and were using colchicine 1-1.5 mg/day were included. An independent observer blinded to group allocations performed all assessments. At the end of a 12-week observational period during which all continued to use colchicine, patients were randomized to continue or discontinue colchicine. Randomization was stratified according to disease activity, disease duration, and sex. At the end of the 12-week controlled period, groups will be compared for the number of oral ulcers, genital ulcers, erythema nodosum, swollen joints, tender joints, papulopustular lesions, pain of oral ulcers and genital ulcers, overall disease activity assessed using Behçet's disease current activity form (BDCAF) and Behçet's syndrome activity scale (BSAS), and quality of life assessed using Behçet disease quality of life index (BDQoL) and SF-36. Primary endpoint is the mean number of oral ulcers at week 12.

Discussion: A total of 141 patients have been randomized to continue (70 patients, 45 F/25 M, age; 45±10 years) or discontinue (71 patients, 46 F/25 M, age; 46±11 years) colchicine. At randomization, the mean number of oral ulcers was 1.11±1.14, mean BDCAF score was 1.75±1.17, mean BSAS score was 8.73±7.55 and mean BDQoL score was 1.71±2.41.

Conclusion: Study recruitment has ended and the randomized period is continuing. The 12-week results will be presented.

Frequency of Tuberculosis and Associated Risk Factors in Patients with Behçet Syndrome

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Introduction: The incidence of tuberculosis (TB) during TNF inhibitor (TNFi) therapy is higher in pts with Behçet syndrome (BS) compared to other diseases. However, the reasons for this and whether BS pts are more prone to develop TB were not well known.

Method: Among the pts recorded between 2006-2011, 300 consecutive BS pts who used only colchicine, 300 consecutive BS pts who used conventional immunosuppressives, 300 ankylosing spondylitis (AS) pts who used conventional treatment and 300 AS pts who
used TNFi as well as all BS pts who started TNFi between 2001-2022, were questioned for a history of TB and their charts were examined. The frequency of latent TB was compared in BS and AS pts who had a tuberculin skin test and/or quantiferon test before starting TNFi. The rate of having a TB ICD code among all BS and AS pts registered in the social security institution (SGK) database of Turkey was determined.

Discussion: History of active TB was similar across the groups who did not use TNFi (3%, 2%, and 3%; p=0.68). Latent TB during screening before starting a TNFi was higher in AS pts compared with BS pts (76% vs 63.8%, p=0.004). Active TB rate during TNFi use was higher in BS pts than in AS pts (HR 3.05, 95% CI 1.2-7.9). The frequency of TB in Turkey was 677/166,668 (0.4%) in BS and 978/286.973 (0.34%) in AS pts. This significant difference was attributed to the large sample size (OR 1.19; 95% CI 1.1- 1.3). Use of prednisolone >15 mg for at least one month, mycophenolate mofetil, age and smoking were associated with TB during TNFi therapy.

Conclusion: The frequency of a history of active TB was similar among BS and AS pts who did not use TNFi. According to SGK data, the incidence of TB in BS and AS pts was also similar and higher than that in the general population. The incidence of TB during TNFi use was 3 times higher in BS pts compared to AS pts. This higher frequency may be associated with concomitant use of corticosteroids and immunosuppressives in

BS.

Mimickers of Nervous System Involvement Among Patients with Behçet Syndrome

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Introduction: We aimed to identify conditions mimicking nervous system involvement among patients with Behçet syndrome (BS) and to determine clinical, laboratory, and imaging findings that may help in the differential diagnosis.

Method: We screened the charts of 500 consecutive BS patients to identify those who were referred to neurology at any time during their follow-up. The final diagnoses, presenting signs and symptoms, laboratory, and imaging results were retrieved from patient charts. Patients who did not have a follow-up visit during the last 3 months were invited to the clinic.

Discussion: Among the 500 BS patients, 116 (23%) had been referred to neurology. Among these, 29 (5.8%) were diagnosed with typical central nervous system involvement of BS (neuroBS). The type of neuroBS was parenchymal involvement in 21 patients, cerebral venous sinus thrombosis in 7 patients, and both in 1 patient. 30 patients (6%) had other conditions related to the nervous system, 46 (9.2%) did not have a nervous system disorder, and their symptoms recovered spontaneously, and 11 (2.2%) were lost to follow-up without a definite diagnosis. Of the 30 BS patients who were diagnosed with another nervous system condition, 14 (46%) had primary headache syndromes, 6 (20%) had psychiatric disorders, 2 had entrapment neuropathy, and 1 each had epilepsy, glial tumor, multiple sclerosis, Meniere's disease, optic neuritis, neuroretinitis, steroid myopathy and polyneuropathy.

Conclusion: Nervous system conditions other than neuroBS are frequent in BS patients referred to Neurology. Caution is required to avoid misdiagnosis of these patients as NeuroBS.

Neuro-Behçet revealed by an hemiface edema

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Introduction: Behçet's disease is a heterogeneous and multisystem inflammatory disorder of unknown etiology. Neuro-Behçet's disease, involving neurological symptoms, is not uncommon. Patients with neuro-Behçet's disease typically present with a diverse array of symptoms, most commonly in the brainstem and diencephalic regions. In this case, we report an unusual case of neuro-Behçet's disease in a patient who presented with right hemifacial edema.

Method: Here, we present a case report about a Moroccan patient, who revealed her Neurobehcet with an usual feature.

Discussion: This is a 32-year-old female patient with a history of familial Behcet's, presenting with bipolar aphthosis that had been recurrent for 4 months. This picture was complicated by headaches, vertigo and memory impairment. This symptomatology was revealed by right hemifacial edema. Clinical examination revealed a pyramidal syndrome. Biological tests showed the positivity of HLAB51.Cerebral MRI angiography and thoracic angiography scan revealed no abnormalities.The diagnosis of an infra-radiological neuro-Behcet ≥4 points was retained. The patient was put on corticosteroids 1 mg/kg/j and colchicine. Her clinical course was good with a follow up of six mounths.

Conclusion: Neuro-Behçet's disease is a medical emergency requiring appropriate therapeutic management and should be systematically investigated as it affects the functional and sometimes even vital prognosis of the patient. The phenotypic heterogeneity of this disease could lead to diagnostic delays, highlighting the importance of a better understanding of its clinical polymorphism and various modes of presentation. The normality of imaging results in our case reinforces the importance of clinical evaluation and underscores the limitations of imaging in diagnosing Behçet's disease.

Efficacy and Tolerability of Switching Colchicine Preparations in Patients with Behçet Syndrome

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Introduction: Beneficial results have been reported among FMF pts who switched from one colchicine (COL) preparation to another due to inefficacy or intolerability. We thought that this may also be true for Behçet syndrome (BS) pts with mucocutaneous and/or joint involvement, who are refractory or intolerant to a COL preparation and would otherwise be prescribed more costly and/or immunosuppressive treatment modalities.

Method: We conducted a retrospective chart review of 47 BS pts who switched to another COL preparation between 2017 and 2023.

Discussion: Among the 47 pts (29 F (60%), mean age: 38±14), the reason for switch was lack of efficacy in 34 and adverse events in 13. Overall, 32 (68%) were still on the switched COL preparation during a mean follow-up of 32±26 months. Seven had discontinued the drug due to adverse events (GI symptoms in 5 and elevated liver function tests in 2), and 6 had stopped the drug due to inefficacy. Mean pts' VAS (5.6±2.5 vs 3.6±3.4; p=0.013) and physicians' VAS (4.0±2.2 vs 2.2±2.7; p=0.003) were significantly improved at the final visit, while mean BDCAI scores and CRP levels were not different between the first and last visit. Among the 34 pts who switched due to inefficacy, the primary reason for switching was mucocutaneous manifestations in 27 and arthritis in 7. 25 (73%) were still using the switched COL preparation for a mean duration of 29±23 months, 5 stopped the drug due to inefficacy, and 4 discontinued the drug due to GI symptoms that were not present with the previous COL preparation. Among the 13 pts who switched due to intolerance, 7 were still using the drug for a mean duration of 42 ± 34 months, 3 had to discontinue the drug due to the same adverse event, 1 stopped the drug due to inefficacy, and 2 were unable to continue the drug due to insurance problems

Conclusion: Switching between COL preparations seems to be a reasonable option for BS pts, providing good drug survival rates, patient and physician global scores, and fewer adverse events.

Abdominal Surgical Interventions Among Patients with Gastrointestinal Involvement of Behçet Syndrome

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Introduction: We aimed to investigate the clinical characteristics, treatments, and long-term prognosis of gastrointestinal involvement of Behçet syndrome (GIBS) pts who underwent abdominal surgical interventions.

Method: Data were collected regarding demographics, type of intervention, treatment, recurrence, and outcome in GIBS pts. Relapse was defined as the presence of endoscopic or clinical activity with a positive fecal calprotectin test following bowel resection.

Discussion: Among our 11,200 pts registered between 1978 and 2022, 119 (1%) had GIBS, and 27 (24%) of these pts (19 M, mean age: 49±11 yrs) had undergone abdominal surgery. All except 1 patient were diagnosed with GIBS following surgery. Perforation (62%) and massive hematochezia (21%) were the main reasons for surgery. The type of surgery was bowel resection in 25 pts, primary closure in 2 and diagnostic laparotomy in 2 pts. 2 pts with refractory GIBS had died due to extensive vascular involvement (n=1) and secondary amyloidosis (n=1). Among the 25 pts who underwent bowel resection, 14 (56%) experienced a relapse during a median follow- up of 24 (IQR: 1.75-36) months, and 12 had only one relapse. 4/12 (29%) required a reoperation. The remaining 11 did not experience any relapses during a median follow-up of 13 (IQR: 10-22) years. Postoperative immunosuppressive treatment was associated with a reduced risk of relapse (3/11 (27%) vs 11/14 (79%); p=0.01; OR: 0.10; 95% CI: 0.02-0.64). Moreover, time to relapse was significantly longer in pts who received postoperative immunosuppressive treatment compared to those who did not (p=0.013)

Conclusion: GIBS may presently acutely with perforation or severe bleeding requiring surgery, as the first manifestation. 24% required abdominal surgery and all except 1 were diagnosed with GIBS following surgery. 50% experienced a relapse, with most relapses occurring within 3 years and 30% of them required reoperation. Immunosuppressive treatment after surgery reduced the risk of relapse by 90%.

Predictive Modeling of Behçet's Disease Using Machine Learning: Insights from Clinical Data Analysis

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Introduction: Behçet's disease (BD) is a chronic, multisystemic inflammatory disorder characterized by recurrent oral and genital ulcers, uveitis, and various systemic manifestations. The complexity of BD needs advanced methods like machine learning to better understand and predict disease patterns and outcomes.

Method: A cohort of Moroccan BDpatients was analysed using machine learning techniques. Data included demographic and clinical informations, and disease progression. Statistical models were applied to identify keypredictors of disease manifestations/outcomes

Discussion: The cohort consisted of 561 patients (398 males and 163 females) with a mean age of diagnosis at 30 years±12.5. Machine learning analysis identified significant predictors of disease severity and organ involvement. For instance, the presence of familial history (OR = 2.3, 95% CI: 1.1-4.5) and cardiovascular risk factors (OR = 1.8, 95% CI: 1.0-3.2) were associated with increased systemic involvement. The analysis also highlighted the correlation between early onset of symptoms and more severe ocular involvement (AUC = 0.78).

Conclusion: Machine learning models can effectively predict disease patterns in Behçet's disease, aiding in early diagnosis and personalized treatment plans. Further research with larger cohorts is needed to validate these findings and enhance predictive accuracy.

Crohn's Disease and Intestinal Behcet: Two Sides of The Same Coin

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Introduction: Crohn's disease is a chronic inflammatory disorder that may affect the same organs as Behcet's disease, Both disorders have fluctuating courses and when gastrointestinal symptoms are prevalent, differential diagnosis can be difficult, as current diagnostic criteria are not clear between both conditions.

Method: we report the case of two patients whose diagnosis of behcet's disease was not easy.

Discussion: Case 1: This 54-year-old woman was admitted to our department to assess the etiology of a 6-year history of oral ulceration of the inner cheek, associated with inflammatory polyarthralgia. Her pathological history included glutinous diarrhea with recurrent episodes of decreased visual acuity, and ophthalmological examination revealed unilateral anterior uveitis. Serologies for HIV and hepatitis A, B and C, cytomegalovirus, herpes virus and toxoplasmosis were negative, and the laboratory work-up revealed a moderate inflammatory syndrome.The endoscopic and pathologic findings were highly unspecific, which precluded an early diagnosis of BS and diverted the clinician towards the more frequent diagnosis of CD Case 2: A 53-year- old man treated in our department for behcet's disease was admitted with pseudofolliculitis and recurrent bipolar aphthosis with budd chiari syndrome, which had been dormant for 10 years, who consulted us for a recurrence of oral aphthosis with episodes of glairy diarrhoea. Biological work-up showed an inflammatory syndrome, and further colonoscopy revealed colonic aphthous ulcerations, with histological evidence of an inflammatory infiltrate and fibrosis, which are not specific to the disease. Both patients were treated with immunosuppressants, with clinical improvement but persistent aphthoid lesions on endoscopic examination, which did not indicate the need for biotherapy.

Conclusion: Differential diagnosis between intestinal BD and CD remains a challenge for clinicians, and both conditions have significant clinical, diagnostic and therapeutic overlaps.

Unusual Manifestations Revealing Behçet's Disease: A Study of 43 Cases.

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Introduction: Behçet's disease (BD) is a vasculitis with venous tropism affecting young individuals in countries bordering the eastern Mediterranean.Unusual manifestations may reveal this disease. In this study, we will investigate these manifestations in patients with BD.

Method: This is a retrospective monocentric study of BD cases selected according to the international diagnostic criteria of the International Study Group conducted over an 18-year period from 2006 to 2023.

Discussion: Results: Among a series of 563 patients followed for BD, 44 cases (7.8%), had an unusual mode of presentation. Acute fever was indicative in 21 patients (47.7%), BD presented in its benign cutaneous-mucosal form in 2 cases, in an articular form in one patient, and in the severe form involving the eyes, neurological system, vessels, and digestive system in the other 18 cases. It included panuveitis in 1 patient, thrombotic event in 7 cases such as lower limb venous thrombosis (3 cases) and pulmonary embolism (2 cases), Budd-Chiari syndrome (3 cases), cerebral venous thrombosis in one case, Behçet's enteritis complicated by severe digestive hemorrhage in one case, and central parenchymal neurological involvement in 7 patients. Prolonged fever was indicative in 15 patients, accounting for 34% of cases, and was correlated with severe vascular involvement in 8 cases. Central parenchymal neurological involvement was found in 6 patients, and benign articular involvement in 1 case. Orchiepididymitis was indicative in 6 patients (13.6%). Parotidomegaly associated with dry mouth syndrome revealed BD in only 1 case (2.3%). It was correlated with severe vascular involvement, and lip biopsy concluded to AA-type amyloidosis. Stage III papilledema associated with signs of intracranial hypertension revealed BD in only one patient (2.3%).

Conclusion: Although Behçet's disease is primarily revealed by classical symptoms, it is crucial to recognize the existence of unusual manifestations that may reveal this pathology.

Behçet's Disease and Cancer: a study of 6 cases.

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Introduction: Behçet's disease (BD) sits at the crossroads of autoinflammatory and autoimmune diseases, with its etiopathogenesis still poorly understood. The association between cancer and BD is rarely reported. It poses a significant diagnostic challenge, dominates the prognosis, and suggests an intrinsic carcinogenic potential of the disease. We report six cases of this association.

Method: This is a retrospective monocentric study of BD cases diagnosed according to the international diagnostic criteria of the International Study Group, conducted over a 17-year period from January 2006 to December 2023.

Discussion: Results: We collected six cases of cancer among 563 patients followed for BD (8%). These included lung cancer (n=1), breast cancer (n=2), multiple myeloma (n=2), and kidney cancer (clear cell adenocarcinoma) (n=1). These cancers occurred in 4 men and 2 women, with a mean age of 51.75 years and an average interval from BD diagnosis to cancer onset of 13.25 years. Our patients did not have particular risk factors. All patients had mucocutaneous involvement; two had vascular involvement, and one female patient had ocular involvement in the form of retrobulbar optic neuritis. Two patients received six boluses of cyclophosphamide (CYC), followed by azathioprine (AZT) 150mg/day. A nephrectomy was indicated for the patient with kidney cancer, and both breast cancer patients underwent chemotherapy and radiotherapy.

Conclusion: Data on the incidence of malignant tumors in patients with BD are limited. The autoimmune component of this disease is significant in its correlation with cancers, and this risk is increased by prolonged exposure to immunosuppressants, including AZT and CYC. In our study, only two patients received IS, so their implication cannot be proven. Further studies are necessary to better understand this causal link or to identify predictive factors for neoplasia in BD, in order to prevent and detect them early.

Behçet's disease and pregnancy: 23 pregnancies in 10 women

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Introduction: Behçet's disease is particularly common in Mediterranean countries and the Far East. Because it is predominantly male, the association with pregnancy is rarely reported in the literature.

Method: Retrospective descriptive study of 10 women with one or more pregnancies in a series of 25 cases of known female Behçet's disease followed for approximately 9 years in an internal medicine department, all meeting the classification criteria of the International Study Group of Behçet's Disease.

Discussion: During the study period, we observed 23 pregnancies in 10 women, an average of 1.4 ± 0.72 pregnancies per woman (extremes 1 and 3 pregnancies/woman-median 1). In all cases, Behçet's disease was already known and treated, the mean time between diagnosis of the disease and the first pregnancy observed being 4 years ± 2.3 extremes of 6 months and 5 years. The mean age at the time of pregnancy was 31.9 ± 5.34 (extremes 22 and 39, median 32). Among these women, 8 presented a minor or moderate form of the disease, cutaneous-mucosal, ocular and/or articular, and 2 a severe form of the disease, with vascular involvement such as thrombosis of the central retinal vein, and one presented a neuro-behcet's disease. In 8 patients, the disease remained stable during 16 pregnancies. In 2 patients, pregnancy was marked by the occurrence of one or more flare-ups, sometimes disabling, of oral, cutaneous and/or genital aphthosis. All 23 pregnancies were carried to term, and all newborns showed no clinical symptoms reminiscent of the maternal disease.

Conclusion: In line with the results of other series reported in the literature, pregnancy had no adverse effect on the disease, and the disease did not appear to interfere with the natural course of pregnancy. A close collaboration between internists and obstetricians and gynaecologists, so that pregnancies can be brought to term in the best possible conditions.

Intracardiac Thrombosis in Behcet Syndrome: A Case-Control Study

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Introduction: Intracardiac thrombosis (ICT) in Behcet syndrome (BS) is rarely documented, limiting our understanding of this condition. This study aims to provide a comprehensive description of patients with ICT and investigate the factors associated with its occurence in BS.

Method: This retrospective case-control study included 15 BS patients with ICT and 75 control BS patients. Six controls per case, matched on age and sex, were randomly selected. Categorical variables were compared using Chi-squared and Fisher's exact tests. Continuous variables were compared with t-Student test.

Discussion: Patients were predominantly men (87%), aged 30 ± 7 years old [18-47]. The main cardiovascular risk factors were smoking (28%) and high blood pressure (12%). ICT was the revealing manifestation of BS (50%) or occurred within the first 5 years of the disease. Patients with ICT were asymptomatic (45%) or presented with dyspnea (50%), chest pain (43%), hemoptysis (36%), and cough (36%). ICT was always visualized in the right cavities (n=15), with multiple thrombi observed in 6 cases. All patient had increased inflammtory markers with normal hemostasis paremeters and negative thrombophilia work-up. Compared to the control group, the factors associated with ICT were pericarditis (57% vs 1%, p<0.001), vascular involvement (91% vs 53%, p=0.01), particularly pulmonary artery aneurysms (62% vs 4%, p<0.001), pulmonary artery thrombosis (54% vs 1.5%, p<0.001) and vena cava thrombosis (43% vs 8%, p=0.005). There was no statistical difference in lower extremity DVT between the two groups (37% vs 16%, p=0.16). Cyclophosphamide was far more often administered in patients with ICT (87% vs 18%, p<0.001).

Conclusion: To our knowledge, this is the first case-control study on ICT in BS. Our findings reveal a distinct clinical phenotype with right-sided ICT, vena cava thrombosis and pulmonary artery involvement. This supports that ICT in BS is an in-situ phenomenon and a continuum of the vascular thrombosis due to the underlying vasculitis.

Vascular Thrombosis in Behçet's Syndrome: A Comprehensive Analysis of 146 Cases

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Introduction: Vascular thrombosis is common in Behçet syndrome (BS), associated with high morbidity and mortality. We aimed to investigate BS patients with thrombotic events (TE) and identify the factors associated with relapse.

Method: We retrospectively conducted a descriptive and analytic monocentric study, from 2000 to 2022, enrolling all BS patients with TE, including venous thrombosis (VT) and arterial thrombosis (AT).

Discussion: TE was observed in 146/550 patients (26%), mostly men (82%), aged 30 ± 8 years (18-50). TE was the initial presentation of BS (31%) or occurred after 5 years [0;15]. TE comprised (93%), AT (19%), and both (12%). VT mostly consisted of DVT in the lower extremities (60%), vena cava (30%), and cerebral (15%) veins, involving one (55%), two (27%), and multiple (16%) veins. AT mainly affected the pulmonary (44%), femoral (22%), coronary (16%), cerebral (9%) arteries, and aorta (9%). Arterial aneurysms were common (18%), especially in the pulmonary arteries (11%). Intracardiac thrombosis was also visualized (12%). Noncardiovascular manifestations were mainly ulcers (97%), pseudofolliculitis (37%), ocular lesions (40%), and arthralgias (20%). Hemostasis parameters were normal. Patients were treated with colchicine (98%), corticoids (98%), and immunosuppressants (80%). Anticoagulants and anti-platelets were administered in 90% and 15% of the cases, respectively. Six patients underwent surgery (4%). Post-thrombotic syndrome was common (15%). Relapse was observed in 25% of the cases, mainly in men (81%), of younger age (20 ± 6) at first TE. Relapse consisted of one (56%), two (30%), or multiple episodes (15%), occurring in the same vascular system (70%), a different one (11%), or both concurrently (20%). Multivariable analysis revealed TE at onset, arterial thrombosis and aneurysms as factors associated with relapse ($p<0.001$).

Conclusion: Our study pinpoints distincts aspects of TE in BS, with TE at onset, arterial thrombosis, and aneurysms emerging as factors associated with relapse.

Pulmonary artery thrombosis in Behcet Syndrome: A Case-Series

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Introduction: Arterial disease in Behçet's syndrome (BS) is not common, manifesting primarily in the form of pulmonary artery aneurysms (PAA), and pulmonary artery thrombosis (PAT) in the 1/3 of cases. This study aims to depict the profile and outcomes of BS patients with PAT.

Method: We conducted a retrospective, descriptive and monocentric study, between 2000 and 2022, enrolled all BS patients presenting with PAT.

Discussion: 18/550 cases were enrolled (3%). The mean age of the patients was 32 ± 7 years (19-58), and the sex ratio (M/F) was 4.8:1. The main cardiovascular risk factors were smoking (12%) and high blood pressure (6%). PAT occurred after a median of 4 years following BS onset. Patients were mostly asymptomatic (70%), or presented with dyspnea (35%), hemoptysis (31%), and thoracic pain (25%). On chest CTA, PAT was often bilateral (78%) and proximal (46%). PAA were frequently associated (25%). DVT was found in the lower extremities (19%), superior vena cava (27%), and inferior vena cava (6%). Cardiac involvement was frequent (40%), mostly consisting of right intracardiac thrombosis (35%). Concomitant noncardiovascular involvements included mucocutaneous (87%), ocular (26%), articular (25%), and digestive (6%) lesions. Inflammatory parameters were increased (97%) with normal hemostasis parameters and negative screening for inherited and acquired thrombophilia. Patients were treated with colchicine (97%), glucocorticoids (93%), cyclophosphamide (77%), azathioprine (77%), and TNF-a-inhibitor (6%). Curative anticoagulation with LMWH and VKA was frequently prescribed (60%). No surgery or endovascular procedure was performed. Relapse and death were recorded in respectively 15% and 29% of the cases.

Conclusion: Our study underscores particular features of PAT in BS, supporting that it arises from in-situ immunothrombosis due to the vasculitis rather than hypercoagulability. The diagnosis should be quickly considered in young men presenting with vascular lesions to ensure prompt and adequate mamanagement.

Clinical and Ultrasonographic Correlates of Postthrombotic Venous Remodeling in Behçet's Syndrome: A Cross-Sectional Study

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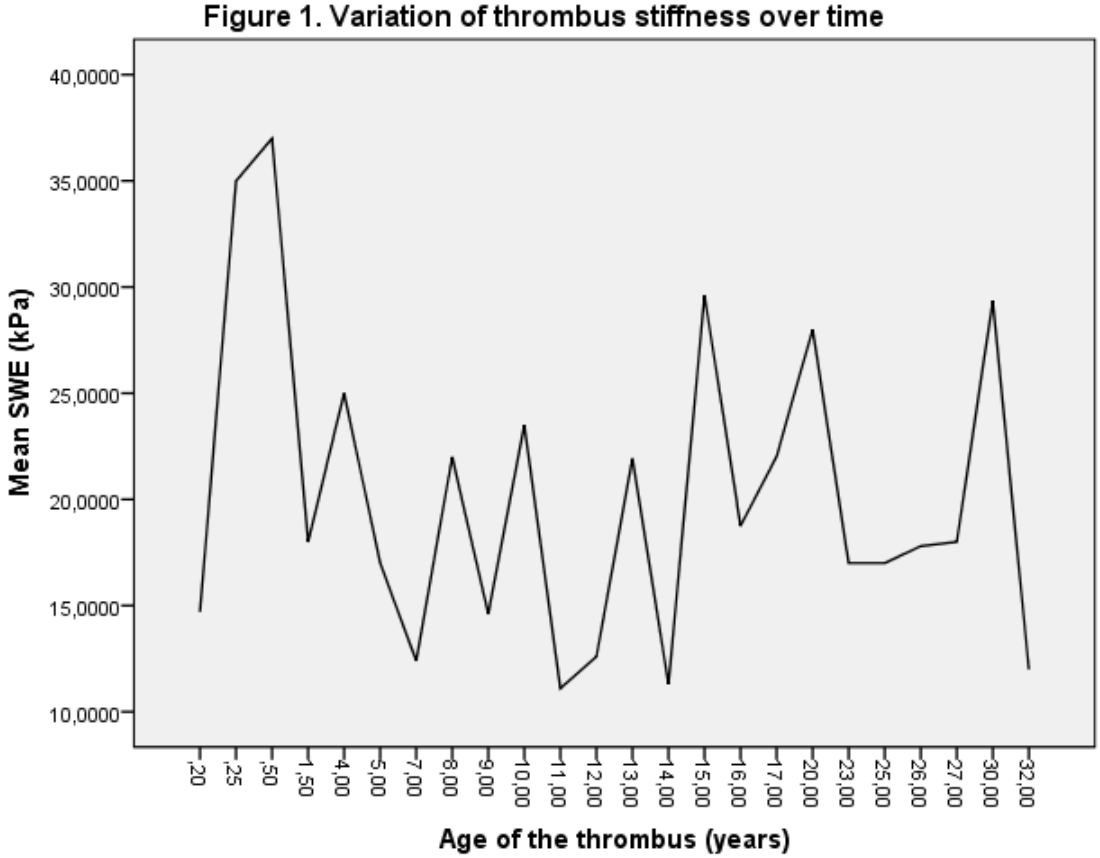
Introduction: Vascular involvement can occur in around half of patients and is characterized by frequent relapses. Superficial vein and deep vein thrombosis are the most common vascular involvement type, and severe postthrombotic syndrome (PTS) develops in up to 20% of patients. The aim of this study was to determine the clinical and ultrasonographic parameters affecting venous remodeling and risk factors for severe PTS.

Method: Two independent radiologists blinded to the clinical features of patients performed the examinations. Venous reflux, vein wall thickness and diameter were measured in saphenofemoral junction. Thrombus and CFV wall elastography were also performed.

Discussion: 79 BS patients (71M/8F) were included in this study. A total of 35 obvious thrombi were observed in 32 patients. Although no significant difference was detected in thrombus stiffness over time, the stiffness of thrombi tended to be higher in the first six months (Fig 1). While there was a trend towards an increase in the probability of severe PTS as the stiffness of the CFV wall increased (Fig 2), we found a negative relationship between CFV wall thickness and severe PTS (Fig 3). Moreover, as venous reflux time (sc) increased, the probability of severe PTS (Fig 4) and stasis ulceration (Fig 5) increased.

Conclusion: Our study shows venous reflux and fibrotic bands within the veins are the most critical parameters for severe PTS. Also, thrombus material in the veins probably causes venous reflux by disrupting the structure of the venous valves. Fibrosis in the vein wall, possibly due to chronic thrombus, contributes to venous insufficiency by making the vein wall stiffer and thinner. Age, disease duration, relapses, and thrombotic events in both legs stand out as clinical factors that trigger adverse venous remodeling and predispose to severe PTS. Achieving complete recanalization of the thrombus and preventing recurrent thrombotic attacks are the ultimate goals of treating BS thrombus.

Figure 1. Variation of thrombus stiffness over time



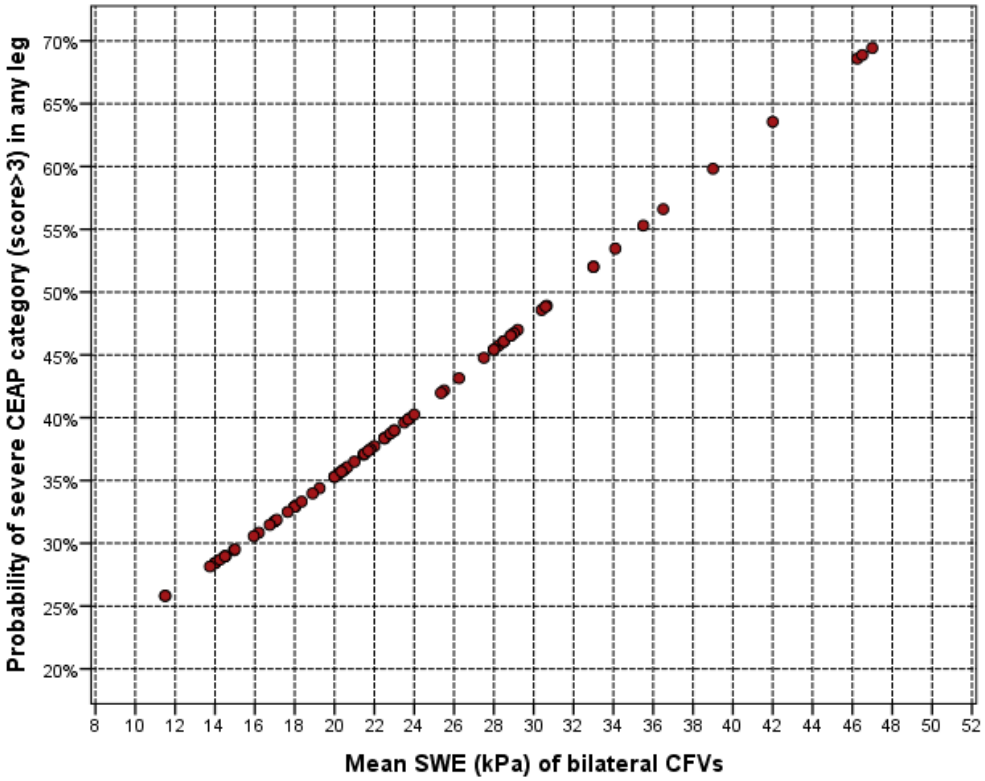


Figure 2. Probability of severe postthrombotic syndrome according to CFV wall stiffness.

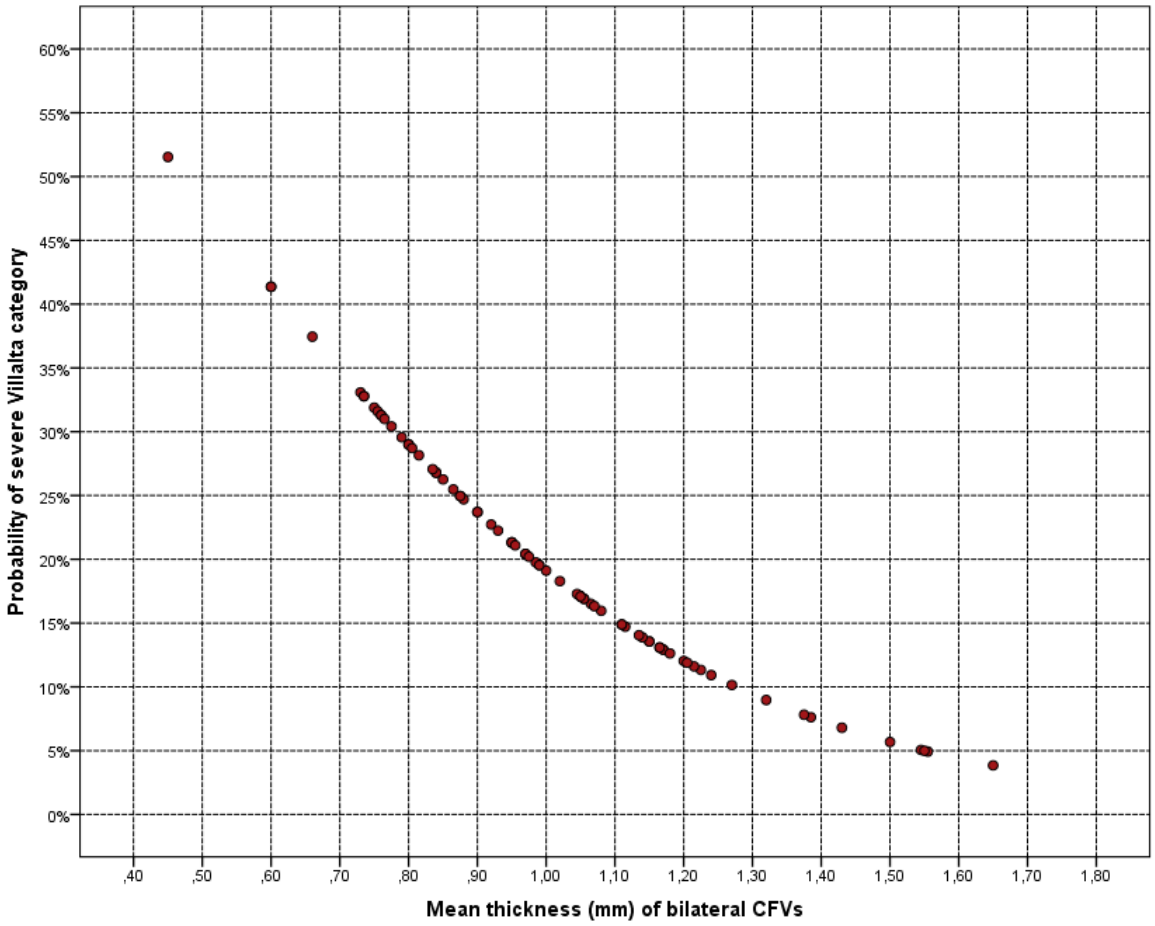


Figure 3. Probability of severe postthrombotic syndrome according to CFV thickness.

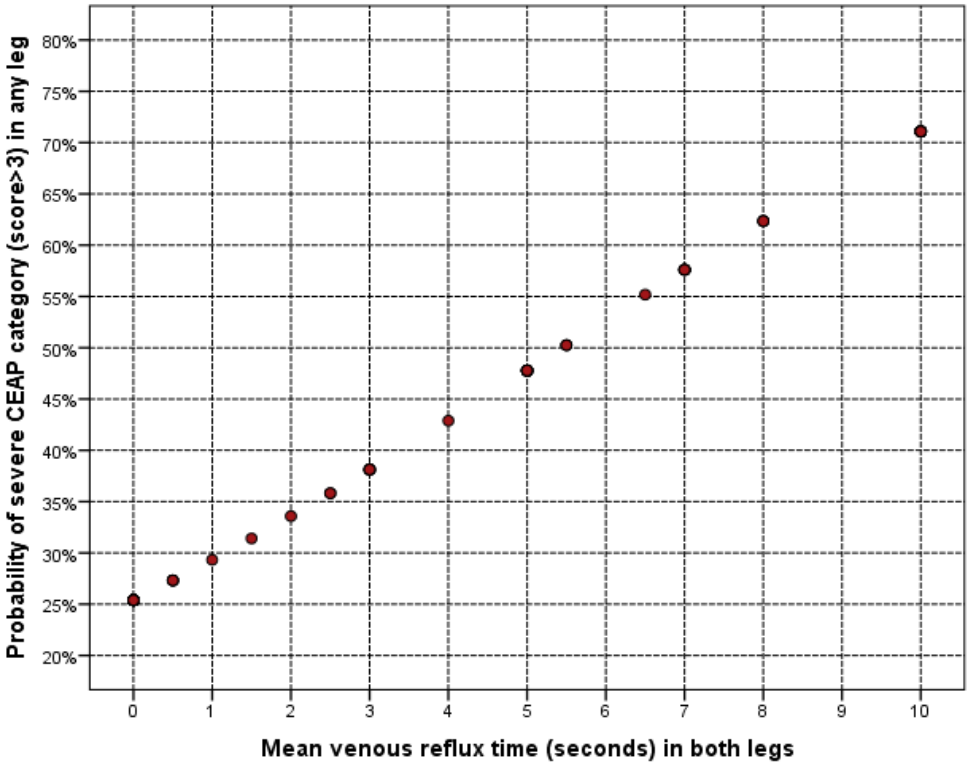


Figure 4. Probability of severe postthrombotic syndrome according to venous reflux.

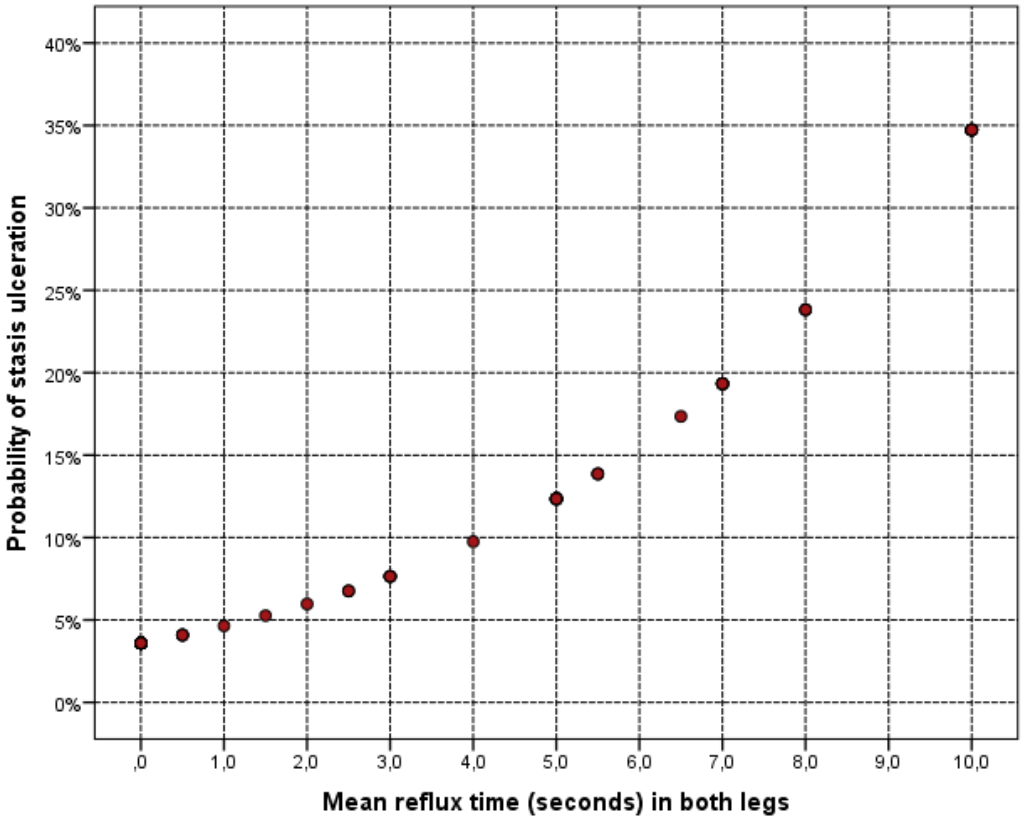


Figure 5. Probability of stasis ulceration according to venous reflux time.

Acute myocarditis revealing Behçet's disease: a case report.

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Introduction: Myocarditis in Behçet's disease is rarely documented and is one of the rarest cardiac disorders compared with more common manifestations such as pericarditis and vascular anomalies.

Method: We report a rare case of acute myocarditis revealing behcet's disease documented by cardiac magnetic resonance imaging (MRI).

Discussion: A 26-year-old patient presented to the emergency department on Day 3 with chest pain associated with fever and deterioration in general condition, followed by a picture of heaviness in the right hemisphere with facial involvement. He was admitted immediately to hospital for an aetiological assessment. The patient's medical history included recurrent episodes of mouth ulcers and genital ulcers. Clinical examination revealed fever, pseudofolliculitis lesions, right hemiplegia, aphasia and right facial paralysis. The ECG showed rhythm disturbances, and the cardiac echocardiography identified an alteration in ejection fraction to 45% with antero- and septo-apical hypokinesia; cardiac MRI: late non-transmural enhancement suggestive of myocarditis. Cerebral CT scan revealed a giant stroke in the territory of the sylvian artery. Aetiological work-up, in particular infectious, virological and autoimmune, and a thrombophilia work-up were negative. A biological inflammatory syndrome with a C-reactive protein of 150mg/L. The diagnosis of acute myocarditis complicating Behçet's disease with associated neurological damage was accepted, and treatment combining, systemic corticosteroid therapy with a bolus of methylprednisolone 1g/day for 3 days and IV bolus of cyclophosphamide 1g was started in addition to the stroke treatment. The clinical, biological and iconographic evolution was rapidly favourable.

Conclusion: Although myocarditis is a rare manifestation of Behçet's disease, its presence is significant and can have serious consequences. Understanding and recognising the potential of these rare complications is crucial to the management of these patients.

CMV Associated Uveitis in Immunocompetent Patient and Ocular Behçet's Syndrome: A Diagnostic Challenge

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Introduction: Ophthalmological manifestations in Behçet's disease are dominated by uveitis,retinal vasculitis and retinal vein thrombosis. It is a serious manifestation because it can lead to the most dreaded complication: blindness. This case is interesting because it discusses the differential diagnosis of behçet's disease and CMV associated uveitis.It also highlights the diqgnostic challenges of ocular manifestations of behçet's disease.

Method: A 34 years old patient with a 4-year history of behçet's disease was admitted. The main complaint was a bilateral blurry vision, eye redness and pain.

Discussion: The ophthalmological examination revealed bilateral panuveitis with retinal vasculitis and a visual acuity of 1/10. The patient was started on corticosteroids pulse therapy 1g during 3 days followed by oral corticosteroids without clinical response. The patient was started on immunosuppressive drugs: cyclophosphamide for 8 months with no improvement. He then received anti TNF alpha adalimumab but still without any improvement. Another visual evaluation was performed revealing a chorioretinal lesion and pizza-pie appearance suggestive of CMV associated uveitis. The patient was then started on antiviral therapy: ganciclovir followed by maintenance therapy with valganciclovir. The evolution was marked by complete remission. Making a differential diagnosis between CMV associated uveitis and ocular behçet's disease is fundamental for an accurate and early management. But this can be difficult in some cases, especially in immunocompetent patients. The classical form is associated with the presence of focal areas with retinal necrosis and retinal hemorrhages. The diagnosis is mainly clinical. Polymerase chain reaction of an aqueous or vitreous sample is very sensitive. The treatment consists of antiviral therapy.

Conclusion: Ocular manifestations of behçet's disease are frequent. CMV associated uveitis in behçet's patients is rare but can be severe. Hence, the importance of an early management of the disease.

Unusual arterial localization revealing Behçet's disease: Splenic artery aneurysm

Ichrak Hajaj, N. Mouatassim; H. Khibri, M. Maamar; H. Harmouche, Z. Tazi Mezalak, M. Adnaoui
Co-Authors: Chhih Yasnima, Khibri Hajar, Chadli Sara, Mouatassim Naima, Ammouri Wafaa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida

Introduction: Behçet's disease is a multisystemic vasculitis most often responsible for venous involvement: superficial or deep thrombosis. - Arterial involvement is rarely encountered: arterial aneurysms (aorta and pulmonary arteries). - We report a rare case of a patient in whom Behçet's disease was considered due to an aneurysm of the splenic artery.

Method: Patient aged 40 years. Medical history: 2 episodes of deep vein thrombosis and an episode of undocumented ocular redness. Disease history: 2 years, recurrent oral and genital aphthosis accompanied by inflammatory low back pain. Clinical examination: murmur at the mitral and aortic foci. Laboratory tests: no inflammatory syndrome. Negative immunological workup. Cervico-thoraco-abdominopelvic angioscanner: splenic artery aneurysm measuring 33*35mm. Diagnosis of Behçet's disease established. Endovascular treatment with good exclusion of the aneurysm on control arteriography. Medical treatment: corticosteroid therapy 0.5 mg/kg/day, colchicine 1 mg/day, and azathioprine 2mg/kg/day with resumption of anticoagulation 8 weeks after the intervention. Asymptomatic patient at 4 months of follow-up.

Discussion: Arterial involvement in Behçet's disease: 1.5 to 2.2% of cases. Involvement of arteries of various sizes. Splenic involvement rarely reported: 5 cases found in the literature. Discovery circumstances: incidental or severe presentations (hemoperitoneum). Positive diagnosis: computed tomography. Treatment: medical with immunosuppressants and surgical in case of rupture risk. Endovascular interventional radiology: a new therapeutic alternative, with decreased morbidity and mortality rates.

Conclusion: Aneurysms of visceral arteries in Behçet's disease: rare with a major risk of rupture. Prognosis: conditioned by an initial multidisciplinary management tailored to the situation (surgical treatment or emergency intervention).

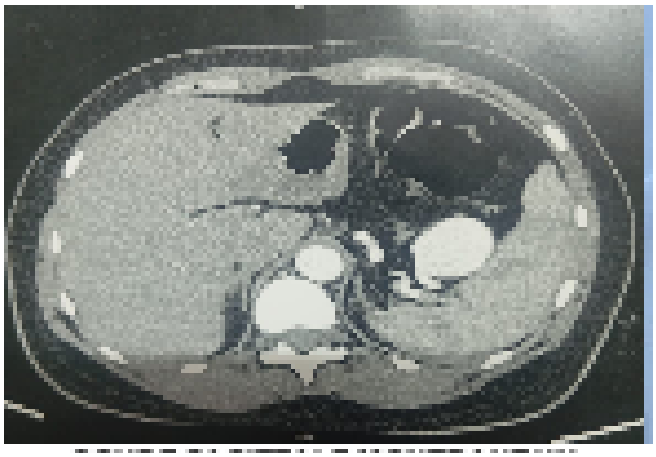

Figure 1. Sagittal section showing a splenic artery aneurysm measuring 33*35mm

Figure 2. Image of arteriography showing aneurysm splenic.

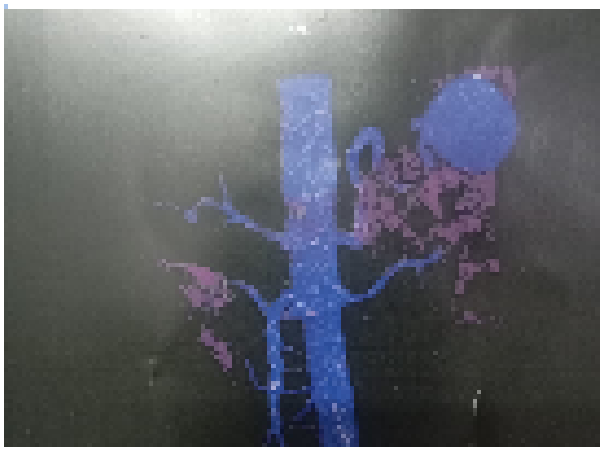

Figure 3. 3D reconstruction of the splenic aneurysm.

Unusual Stenosis in Vasculo-Behcet Disease (BD)

Ichrak Hajaj
Co-Authors: Chhih Yasmina, Khibri Hajar, Chadli Sara, Mouatassim Naima, Ammouri Wafa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida

Introduction: BD: chronic systemic vasculitis + vascular tropism Arterial involvement 4 to 17%: mostly aorta. We report a case of a patient whose vascular involvement was expressed by an aneurysm and a stenosis.

Method: Patient: 53-year-old Followed since 2018 for BD with cutaneous-mucosal involvement, ocular involvement, and vascular involvement (right carotid artery aneurysm). Treatment: carotid-carotid bypass surgery, methylprednisolone bolus, prednisone 1mg/kg/day, cyclophosphamide CYC (vasculitis protocol) and azathioprine AZA 2mg/kg/day for 2 years then lost to follow-up. 2023: upper limb paresthesia. Physical examination was unremarkable. Angio-CT scans of the upper limbs and supra-aortic trunk: total thrombosis of the carotid-carotid bypass, occlusion of the left subclavian artery ostium with segmental resumption, pre-occlusive stenosis of the left internal carotid arteryt. The rest of the vascular mapping: unremarkable. Laboratory tests: no inflammatory syndrome. Treatment: emergency methylprednisolone bolus,oral prednisone 1mg/kg/day, and infliximab with good evolution over a one-year follow- up.

Discussion: Arterial involvement: occlusions (38% to 80%), aneurysms (45% to 70%), pseudoaneurysms, stenoses (13%), aortitis (3%). Doppler ultrasound, contrast- enhanced computed tomography, magnetic resonance and Fluorodeoxyglucose-PET- CT angiography: preferred imaging methods. The Arterial involvement's prognosis: severe. Isolated occlusive or stenotic peripheral arterial involvement: medical treatment often sufficient (corticosteroids+ immunosuppressants: AZA). Treatment of potentially life-threatening conditions (aneurysms): CYC and glucocorticoids. Tumor necrosis factor blockers (infliximab and adalimumab):spectacular efficacy with severe refractory involvement to CYC. Surgery delicate+ frequent rate of complications. Endovascular treatment of aneurysms: increasingly recommended.

Conclusion: Arterial involvement in BD: rare Management: a major concern (high recurrence rate).

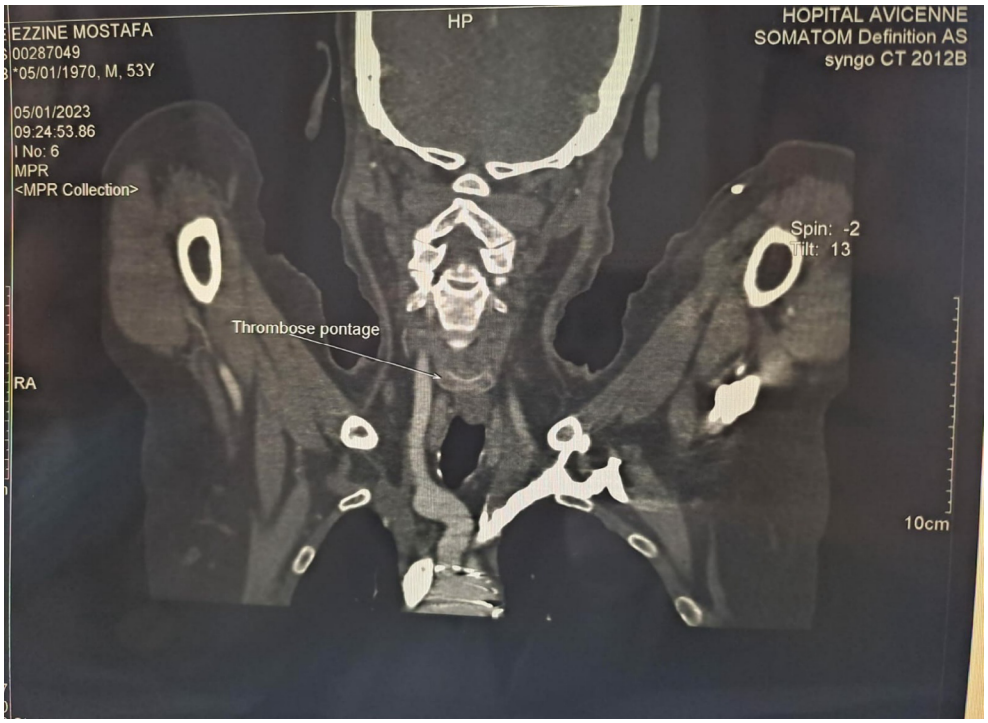

Figure 1. AngioCT showing bypass thrombosis.

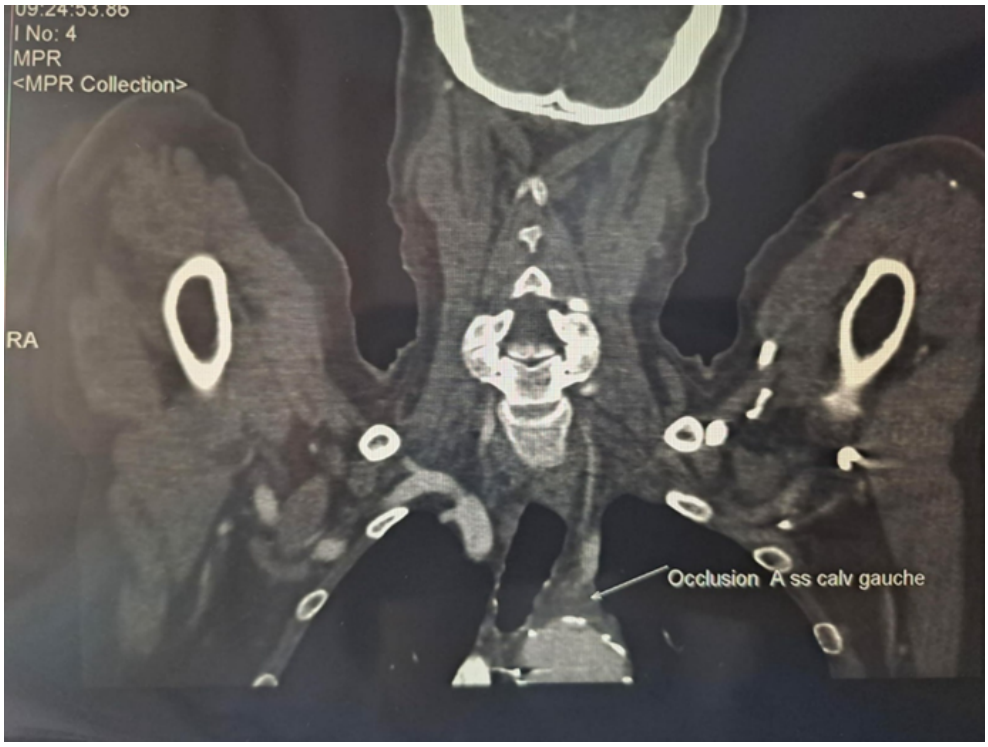

Figure 2. AngioCT showing the occlusion of the subclavian artery.

Clinical characteristics of pediatric Behçet's disease in different geographies

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Co-Authors: Farhad Shahram, Erdem Karabulut, Massoomeh Akhlaghi, Ozlem Akgun, Mustafa Cakan, Esra Esen, Caterina Matucci Cerinic, Seyede Tahereh Faezi, Ruya Torun, Erbil Unsal, Marco Gattorno, Nuray Aktay Ayaz, Aysenur Pac Kısaarslan, Betul Sozeri, Ezgi Deniz Batu, Isabelle Koné-Paut, Seza Ozen

Introduction: We aimed to describe the clinical features and patterns of phenotype aggregation in a large cohort in pediatric BD and to compare the clinical characteristics of BD in three patient groups from three different geographical regions: Turkey, Europe, and Iran.

Method: Retrospective data of pediatric patients with BD in 8 centers from 4 countries were evaluated. Patients with pediatric-onset (<18 years) BD who had a follow-up period of at least 6 months were included. Patients from Italy and France were grouped together as a single category, representing the European countries.

Discussion: A total of 600 patients (297 females, 49.5%) were included in the study. Of these, 231 were from Turkey, 306 from Iran, 44 from France, and 19 from Italy. The median age at diagnosis and the median follow-up time were 14.4 and 3.0 years, respectively. The most common presentations were mucocutaneous (97.5%) and ocular (48.0%), followed by musculoskeletal (43.2%), neurological (11.8%), and vascular (11.5%) involvement. Pathergy test and HLAB51 were positive in 224 (37.3%) and 296 (49.3%) patients, respectively. The frequency of ocular involvement was more prevalent in Iran, gastrointestinal involvement in Europe, and musculoskeletal and vascular involvement in Turkey compared to the other two geographic regions. In pediatric BD, we were not able to define clear clusters as defined in adults. For treatment, the most commonly prescribed drug was colchicine (73.2%) while azathioprine was used in 33.9%, methotrexate in 27.3%, and biologic treatments in 12.5% of patients. Remission on drugs was observed in 47.8% of the patients, remission off drugs in 11.4%, and partial remission in 46.0%.

Conclusion: There are notable variations in the prevalence of organ involvement in BD across geographical regions. However, clear clusters were not defined. BD mainly presents with mucocutaneous and ocular manifestations in the pediatric population but has wide phenotypic clinical variability.

Cardiac involvement in Behcet disease

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Co-Authors: Ines Naceur, Tayssir ben achour, mayssem jridi, Imed Ben ghorbel, Mounir Lamloum, Monia Smiti, Fatma Said

Introduction: Cardiac involvement (CI) in is an uncommon complication during Behcet Disease (BD). This feature is referred to as cardio Behcet and may occur in many forms. The aim of our study is to describe epidemiological, clinical, paraclinical, therapeutic features and outcomes of cardiac involvement in a Tunisian cohort.

Method: We conducted a retrospective, monocentric study enrolling data of BD patients (fulfilling International Criteria of Behcet Disease=ICBD) admitted in our internal medicine department between January 2011 and March 2024.

Discussion: Among 355 BD patients, 10 had cardiac involvement. The mean age at BD diagnosis was 50 years [30-48]. Gender ratio M/F was 9/1. Cardiac involvement revealed the disease in 2 patients. In the other cases, it was diagnosed after a mean delay of 6 years [4-13]. Revealing clinical features were chest pain (n=6), dyspnea (n=2). Two patients were asymptomatic. Biological findings showed inflammatory syndrome (n=2), anemia (n=1) and elevated troponin levels (n=1). Pericarditis was the most common cardiac involvement, reported in 6 patients. Myocardial involvement was reported in 3 patients with myocardial fibrosis, myocarditis, and myocardial ischemia each in one case. Coronary artery aneurysm was reported in 3 patients. Valvular insufficiency was reported in 3 cases. Five patients had heart failure with conserved normal left ventricular ejection fraction (LVEC) in 3 patients and decreased LVEC in two patients (25 and 20%). All patients were treated with corticosteroids which was associated with immunosuppressive therapy in 6 cases: Cyclophosphamide (n=5) and Azathioprine (n=1). Cardiac involvement was associated with extra-cardiac vascular involvement (n=7), neurological disorder (n=4), ocular involvement (n= 1) and IgA nephropathy (n=1).

Conclusion: Cardiac involvement in BD is uncommon but is associated with pejorative prognosis. Hence the importance of systematic and regular screening.

Exceptional association between Behcet's Disease and Overlap Syndrome

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Co-Authors: Yasmina Chhih

Introduction: Overlap syndrome is a rare disease characterized by the overlap of primary biliary cholangitis and /or primary sclerosing cholangitis with autoimmune hepatitis, Behçet's disease is an inflammatory thrombotic systemic vasculitis

Method: A 34 –year- old female patient, with no significant personal or family medical history, has been followed since 2022 for Overlap Syndrome initially revealed by hepatic colic accompanied by profound asthenia.Clinical examination revealed hepatomegaly and generalized jaundice.Laboratory tests showed hepatic cytolysis, as well as the presence of positive antimitochondrial antibodies (greater than 1/80)and a significant titer of anti-smooth muscle antibodies, accompanied by moderate periportal porcellar necrosis on liver biopsy. The patient was treated with corticosteroid therapy as prednisolone at a dose of 1mg/kg/day and azathioprine at a dose of 3 mg/kg /day (150mg/day) with favorable evolution. In January2024, abdominal imaging for surveillance incidentally revealed portal thrombosis.The patient's history was revisited,describing bipolar aphthosis with 4 episodes per year,along with episodes of visual fog.Clinical examination revealed genital scar,pseudofolliculitis,and red right eye. Ophtalmological examination revealed episcleritis without signs of active uveitis or sequelae synechiae.Given the strong suspicion of Beçet's disease, vascular mapping was performed,revealing thrombosis of the left cubital artery.cardiac evaluation was normal. The patient was treated with colchicine, azathioprine and anticoagulation as rivaroxaban20mg with significant clinical and biological improuvement over 5 month follow-up period

Discussion: Our case highlights the crucial importance of thorough history taking and meticulous clinical examination in the managment of patients presenting with thrombosis and concurrent inflammatory diseases.

Conclusion: Our case illustrates the mutifactorial caracter of venous thromboembolic disease.

A heart with two tragedies: a Behçet's disease revealed by an infectious endocarditis.

Ichrak Hajaj
Co-Authors: yassmina chhih, Khibri Hajar, Fari Safae, Sara Chadli, Mouatassim Naima, Ammouri Wafaa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida

Introduction: Cardiovascular involvement is a rare systemic manifestation in Behçet's disease. Primarily thrombotic and predominantly affects the right chambers. We report a case of Behçet's disease with cardiac involvement revealed by an infectious endocarditis.

Method: A 21-year-old patient with recurrent bipolar aphthosis presented with fever and general deterioration over the past few weeks. Clinical examination 5 scrotal scars. Inflammatory markers elevated. The rest of the infectious workup was negative. Transthoracic echocardiography: 2 thrombus of 7x15mm and 18x14mm attached to the right ventricle and the right atrium respectively. A cervico-thoraco-abdomino- pelvic angio-scanner: thrombosis of the suprahepatic veins. Ophthalmological examination normal. Treatment: appropriate antibiotic therapy for his endocarditis. Subsequently: corticosteroids at a dose of 1mg/kg/day, therapeutic anticoagulation, and monthly cyclophosphamide therapy for 6 months followed by azathioprine.

Discussion: Cardiac manifestations of Behçet's disease can be pericardial (29%), endocardial (25%), thrombotic (29%), myocardial (myocardial infarction (15%), fibrotic (endomyocardial fibrosis 8%), and aneurysmal. The presence of cardiac involvement with intracardiac thrombosis has a better prognosis compared to myocardial infarction, but its association with venous involvement worsens the prognosis by increasing the risk of death by 4. The presence of suprahepatic vein thrombosis is also a major prognostic factor, increasing the risk of death by 9. Immunomodulation: the cornerstone of therapeutic strategy (corticosteroids alone or in combination with colchicine and/or azathioprine, cyclophosphamide, cyclosporine). Surgery: cases resistant to medical treatment.

Conclusion: The discovery of serious complications such as intracardiac thrombus in a young male without cardiovascular risk factors in the Mediterranean region should raise suspicion of Behçet's disease.

A Rare Cause of Portal Hypertension: Porto-sinusoidal Vascular Disease (PSVD) and Behçet's Disease: A Case Report

Ichrak Hajaj, N. Mouatassim, H. Khibri, W. Ammouri, M. Maamar, H. Harmouche, Z. Tazi-Mezalak, M. Adnaoui
Co-Authors: Yassmina chhih, Khibri Hajar, Fari Safaa, Chadli Sara, Mouatassim Naima, Ammouri Wafaa, Maammar Mouna, Harmouche Hicham, Adnaoui Mohamed, Tazi Mezalek Zoubida

Introduction: PSVD: Variety of diseases characterized by abnormalities of the small vessels of the liver. - Portal hypertension (PHT): Unusual mode of discovery. - Several pathologies/ associated with PSVD. - Here we report a case of angio-Behçet revealed by PSVD.

Method: Patient aged 46 years. No particular medical history. History of illness: generalized abdominal pain, recurrent bipolar aphthosis (6 episodes/year). Clinical examination: hepatomegaly at 18cm, pseudofolliculitis, and scar of genital aphthae. Investigations: marked inflammatory syndrome. Liver function tests: PAL 6*N without cytolysis, PT at 64% with normal INR, albumin at 62g. Hepatitis viral serologies, iron overload studies, and autoimmune panel were normal. Abdominal angioscanner: liver showing chronic hepatopathy accompanied by thrombosis of the spleno-mesenteric trunk with portal hypertension and hypersplenism. Liver biopsy: PSVD without cirrhosis. Digestive exploration: grade II esophageal varices. Neoplastic, acquired thrombophilic, and constitutional causes ruled out. Diagnosis of PSVD on underlying angio-Behçet established. Treatment: oral corticosteroid therapy,curative and prophylactic treatment for variceal rupture, curative anticoagulation. Induction therapy: 6 monthly cycles of intravenous cyclophosphamide. Maintenance therapy: quarterly cyclophosphamide. Progression: improvement Follow-up of 13 months without complications.

Discussion: PSVD results: intrahepatic vasculopathy+ systemic diseases that include vasculitis processes. Regeneration with compression of the central veins: portal hypertension without cirrhosis. The association of PSVD with Behçet's disease extremely rare, reported only 2 times in the literature so far.

Conclusion: - Portal hypertension in Behçet's disease: various processes but PSVD is rare - Active investigation into treatable causes of non-cirrhotic portal hypertension= improve the prognosis.

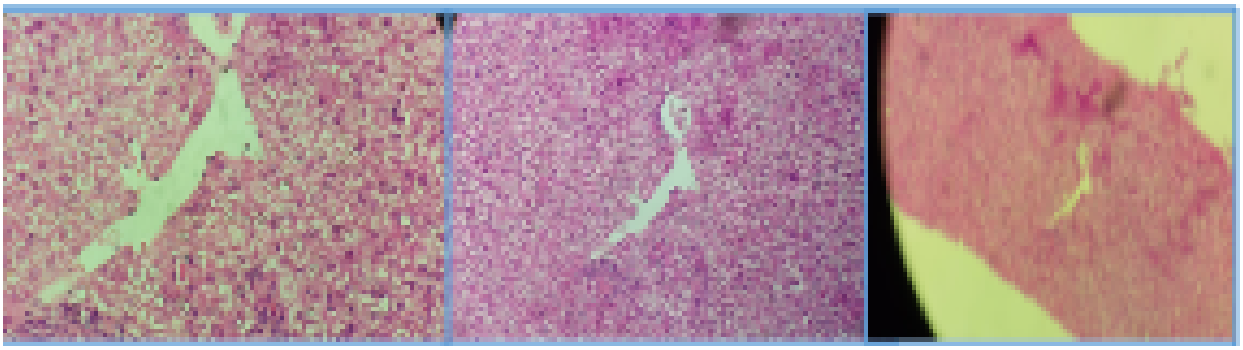

Figure 1. Liver biopsy showing architecture conserved with a vascular structure abnormal lobular.

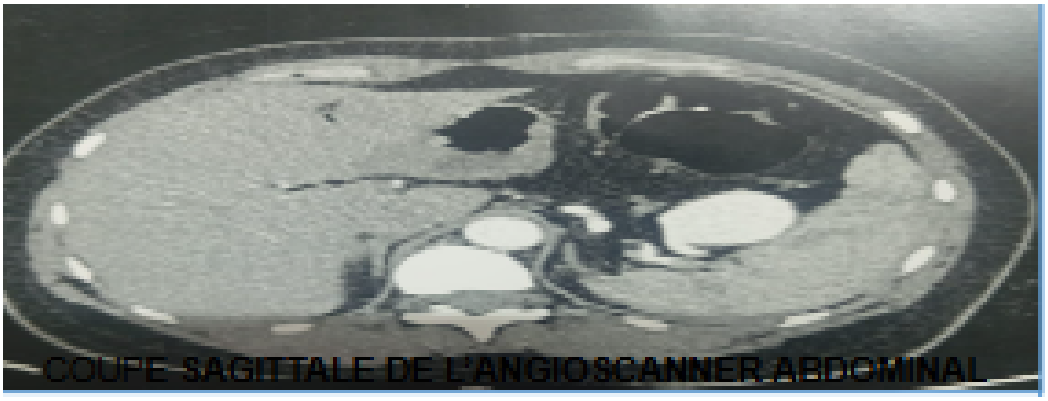

Figure 2. Abdominal ct sagittal section showing spleno-mesaraic trunk thrombosis.

Spectacular image of a Giant False Aneurysm of the Celiac Artery during Behçet's disease

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Co-Authors: Nissrine Bahadi, Oumama Jamal, Zineb EL Bougrini, Farah Ahallat, Bilal Talamoussa, Nawal Sahel, Ilyass El Kassimi, Adil Rkiouak, Youssef Sekkach

Introduction: Alongside venous thrombosis, aneurysmal damage and large-vessel stenosis are the most frequent manifestation in angioBehçet's disease.we report the observation of a young Moroccan man treated for Behçet's disease with a giant false aneurysm of the celiac artery.

Method: we report the observation of a young Moroccan man treated for Behçet's disease with a giant false aneurysm of the celiac artery.

Discussion: Young 17-year-old patient, recently diagnosed with behçet's disease with mucosal cutaneous tropism, presented with intense epigastric and periombilical abdominal pain. abdominal ultrasonography revealed an anechogenic image at the expense of the celiac artery, additional Doppler confirmed the aneurysmal nature of the lesion and its celiac origin. Angioscan with MIP construction(Fig 1) confirmed the presence of a giant false aneurysm at the level of the celiac trunk(5 cm in diameter), which had not ruptured. Biologically, the patient presented with an inflammatory syndrome (CRP at 180 mg/l). Immunosuppressive medications were administered with high- dose corticosteroid therapy (methylprednisolone 1 g/day for 3 days), followed by prednisolone and with oral cyclophosphamide as a steroid-sparing immunosuppressive drug, followed by endovascular treatment with aortic stent grafting. The evolution was marked by clinical and biological improvement, The postoperative course was uneventful, and control imaging showed an intact repair.(Fig 2)

Conclusion: The occurrence of false celiac artery aneurysms in patients with Behçet's disease is extremely rare. Epigastric discomfort remains the predominant symptom. in young patients with false aneurysmal in the course of Behçet's disease, early treatment is indicated particularly in large and symptomatic aneurysms, since the rupture is the main cause of death in such patients.

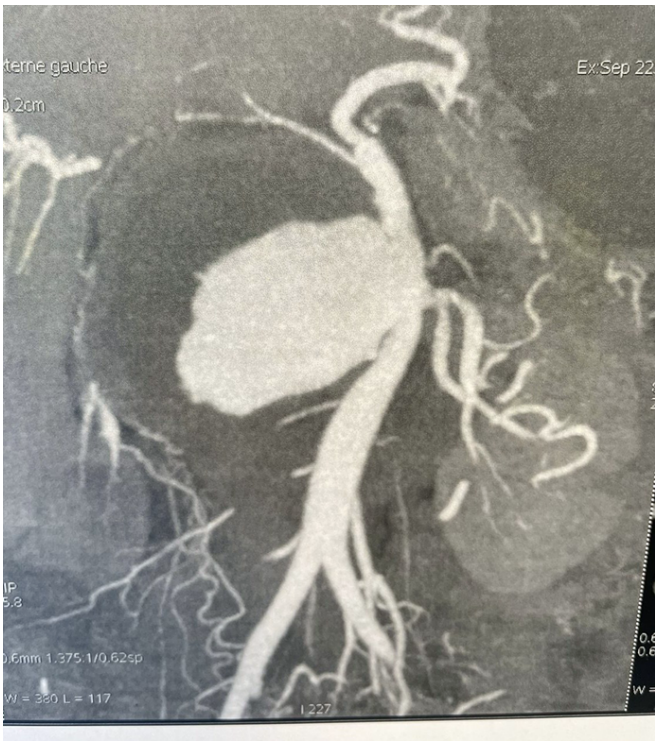

Figure 1. Abdominal angioscan with MIP reconstruction showing a celiac trunk false aneurysm f measuring 50 mm.

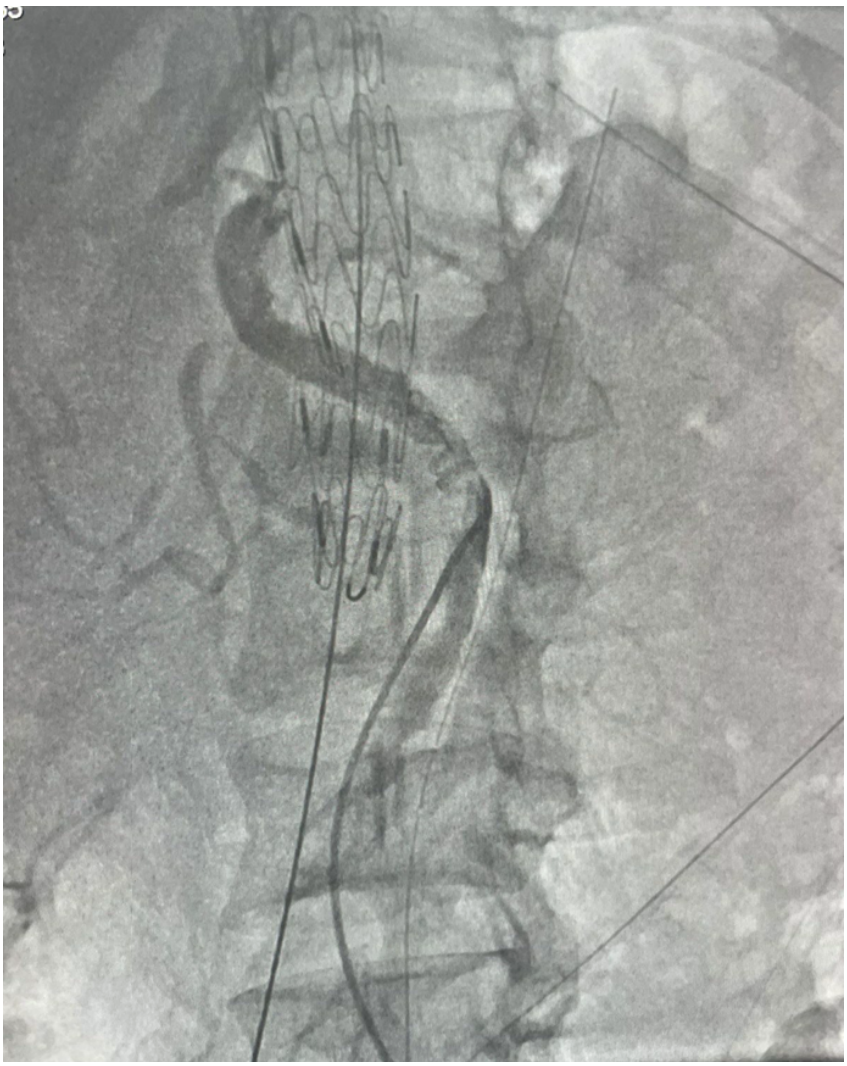

Figure 2. Abdomen scan after endovascular exclusion of the false aneurysm.

Behçet uveitis presenting with or without retinitis: A comparative study

Moncef Khairallah, Faculty of Medicine of Monastir, Tunisia
Co-Authors: Nesrine Abroug, Oumaima Allagui, Wijdene Nabi, Imen Ksiaa, Béchir Jelliti

Introduction: Retinitis occurs in more than 50% of patients with Behçet uveitis. Comparative studies on clinical pattern of patients presenting initially with retinitis (retinal infiltrates) as compared to those presenting without retinitis are lacking.

Method: We described and compared the clinical and multimodal imaging findings between patients with BD uveitis associated with retinal infiltrates (Group 1) and those with BD uveitis without associated retinal infiltrates (Group 2).

Discussion: Our study included 91 patients: 37 patients (49 eyes) in group 1 and 54 patients (114 eyes) in group 2. The mean age was 32.5 ± 10.5 years in group 1 and 30 ± 10.50 years in group 2 (p=0.20). Comparative analysis of extra-ocular manifestations showed that group 2 patients exhibited more frequently clinical signs of neuro-Behçet and angio-Behçet than the group 1 patients (p=0.01). There was no significant difference in initial visual acuity between the two groups. However, retinal hemorrhages, vascular sheathing, retinal arterial involvement, occlusive retinal vasculitis, and vitreous exudates were significantly more frequent in group 1 (respectively p=0.01; p=0.01; p=0.04). There was no significant difference in the rate of fern-like capillaritis between the two groups (31.30% vs 38.10%; p= 0.09). Macular edema on OCT was slightly more frequent in group 2 patients (36.20% vs 21.90%; p=0.06).

Conclusion: Patients with Behçet uveitis presenting with retinitis had lower rates of neurological and vascular involvement. Conversely, retinal hemorrhages, retinal vascular sheathing, retinal arterial involvement, occlusive retinal vasculitis, and vitreous exudates are more frequent in patients with retinitis than in those without retinitis.

Behcet's Disease and IgA Vasculitis: A Real Assosciation or Quincidence ?

Bouzgarou Mohamed, Resident

Introduction: Behçet's disease is a chronic inflammation of the vessels of unknown etiology characterized by recurrent oral and genital aphtosis associated with ocular, cutaneous, vascular, digestive or joint manifestations. Its association with another systemic vasculitis is possible we report a case in an adult presenting with Behçet's disease associated with IgA vasculitis.

Method: A 48-year-old man, followed in our department for Behçet's disease, diagnosed for: oral and genital aphtosis,pseudofolliculitis with positif pathergy test, treated with colchicine. The patient presented an episode of vascular purpura with abdominal pain. he was afebrile, hemodynamically stable with a preserved general condition He had infiltrated petechial lesions in both lower limbs. The abdomen was tender without guarding or contracture The rest of the physical examination was without abnormalities. The biology report, particularly renal, was normal. An abdominal CT angiogram performed did not show any abnormalities. A skin biopsy was performed showing leukocytoclastic vasculitis with high intensity deposit of IgA and C3. The diagnosis of IgA vasculitis was made Corticosteroid therapy at a dose of 0.5 mg/kg/day with a gradual decline over 6 months was started; with a good clinical evolution

Discussion: The association between Behçet's disease and IgA vasculitis is rarely described Renal damage in this case conditions the prognosis Strict nephrological monitoring is recommended **Conclusion:** The association between Behçet's disease and IgA vasculitis is rarely described Renal damage in this case conditions the prognosis Strict nephrological monitoring is recommended

Hughes-Stovin syndrome about 3 cases and literature review.

Fatima Zahra Boucham, Internal Medicine
Co-Authors: Charaf Eddine Amezyane, Mohamed Jira, Fadoua Mekouar, Nawal El Omri, Jamal Fatihi

Introduction: Hughes-Stovin syndrome is a rare disease characterized by the association of isolated or multiple pulmonary arterial aneurysms and systemic or intracardiac deep vein thrombosis. Often considered a severe form of Behçet's disease without cutaneous-mucosal or ocular manifestations, it mainly affects young people and carries a high risk of sudden hemoptysis, especially in the absence of early diagnosis and treatment.

Method: A 20-year retrospective review of 3 cases, explored clinical and morphological features, therapeutic strategies, and challenges in managing Hughes-Stovin Syndrome, along with factors affecting prognosis.

Discussion: We proposed diagnostic criteria for HSS, emphasizing the importance of right intracardiac thrombosis and other rare but suggestive features. Combined immunosuppressive therapy has shown effectiveness, while the use of
anticoagulants must be carefully evaluated. Thrombectomy using AngioVac is an alternative in case of hemorrhagic risk, and urgent endovascular embolization is warranted for pulmonary arterial aneurysms (PAAs) at risk of rupture. Prognosis depends on periodic assessment of hemorrhagic risk, with massive hemoptysis being a frequent cause of death. Early combined immunosuppressive treatment is associated with reduced mortality and should not delay endovascular embolization if necessary.

Conclusion: Hughes-Stovin Syndrome (HSS) is a rare life-threatening condition that can be fatal in cases of sudden hemoptysis without early diagnosis and treatment. The use of anticoagulants remains controversial.

The characteristics of a northern Israeli cohort of patients with Behcet's syndrome

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Introduction: Behcet's syndrome (BS) is a multisystem disease that typically manifests as recurrent oral and genital ulcer, along with other systemic manifestaions. BS is believed to prevalent among Muslims and Druze in Israel. Very few studies describing the characteristics of BS among Israeli patients have been published

Method: we retrospectively reviewed electronic medical records of our BS cohort. Demographic, clinical, laboratory and medications data were extracted. We further compared the Jewish and Arabic subpopulations.

Discussion: 86 patients were included. 43 were males (50%), mean age at diagnosis was 29.9 years old. 61 (70.9%) Arab (39 (45.3%) Muslims, 18 (20.9%) Druze and 4 (4.7%) Christians). Oral and genital ulcers were evident in 82 (95.3%) and 47 (54.7%) patients, respectively. Skin, joints, eye, GI tract, neurologic and vascular systems were involved in 35 (40.7%), 51 (59.3%), 41 (47.7%), 6 (7%), 7 (8.1%), 12 (14%), respectively. 4 (4.6%) had cardiac involvement, one patient had pulmonary hemorrhage and 2 patients had epididymitis. HLA B51 was performed in 32 (37.2%) patients and was positive in 20 (66.7%). Pathergy test was performed in 9 (10.5%) patients and was positive in 6 (66.7%). Colchicine was used in 72 (82.5%) cases, AZA 42 (48.9%), MTX 15 (17.4%), apremilast 7 (8.1%), CsA 7 (8.1%), adalimumab 23 (26.7%), infliximab 10 (14.6%), CYC 1 (1.2%), tocilizumab 2 (2.3%) and anti-coagulation 3 (3.5%). Arabs and Jews were significantly different in male proportion only and the 35 (57.4%) vs 8 (32%), p=0.033.

Conclusion: BS is more common among Arabs in northern Israel, especially among Druze and Muslims, but no significant differences were found except for higher proportion of males among Arabs. Further studies are warranted to understand the true prevalence and genetic bases among the Arab, especially Druze, subpopulation.

Behcet and Hypnos: evaluation of sleep quality in Behcet's disease in a Tunisian population

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Co-Authors: Donia Dridi, Ines Naceur, Maysam Jridi, Imed Ben Ghorbel, Monia Smiti, Fatma Said

Introduction: Behcet's disease (BD) is a systemic vasculitis of unknown etiology. The quality of life can be altered during this pathology and essentially sleep. The purpose of this study is to investigate poor sleep quality in BD patients and to examine the relationship with clinical manifestations.

Method: We conducted a transversal monocentric study in our internal medicine department enrolling BD patients aged over 18 years. The Pittsburgh Sleep Quality Index was applied to determine the sleep quality. Poor sleep quality is defined by a score ≥ 9.

Discussion: Forty one patients were included in this study. The mean age was 43.1±11.5 years. Average disease duration was 8.9±9.1 years. Male to female ratio was 3.2. Oral and genital ulcers were observed respectively in 83,3% and 64.3% of the patients. Articular and vascular involvements were noted respectively in 33.3% and in 31.7% BD patients. Ocular involvement was detected in 28.7% of patients with bilaterality observed in 21.4% of cases.Neurological manifestations were detected in 19.5% of the patients, half of whom displayed parenchymal involvement, while the remaining half presented non-parenchymal manifestations. A poor sleep quality was revealed in 43.9% of the patients. Sleep quality was notably diminished in patients with vascular involvement (30.8% vs. 67.9%; p=0.026) while there were no significant associations with articular involvement (p=0.34) neither neurological nor ocular involvement (p=0.42 and p=0.38, respectively). There were also no statistically significant associations with bipolar aphthous (p=1 and p=0.74 respectively for genital and oral ulcers)

Conclusion: Our study reveals a high prevalence of poor sleep quality in BD patients, particularly among those with vascular involvement, suggesting a potential link between vascular manifestations and sleep disturbances. Hence, further research is needed to better understand and address sleep disorders in BD.

Assessment of fatigue in Tunisian Behcet's patients: prevalence and associated factors

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Introduction: Fatigue is a common symptom of chronic inflammatory diseases such as Behçet's disease (BD) which might have serious implications on the quality of life. The purpose of this study is to investigate severe fatigue in BD patients and its associated factors.

Method: We conducted a transversal monocentric study in our internal medicine department enrolling BD patients aged over 18 years fulfilling the international criteria for Behcet's disease. The Functional Assessment of Chronic Illness Therapy – Fatigue Scale was used in our study.

Discussion: A total of 41 patients were included with a mean age of 43.1±11.5 years. Male to female ratio was 3.2.Mean disease duration was 8.9±9.1 years. Vascular involvement was observed in 31.7% of the patients with 19.5% presenting with lower limb deep vein thrombosis. Vascular aneurysms were diagnosed in 9.8% of cases. Pulmonary embolism and Hughes-Stovin syndrome were identified in 2.4% and 4.9% of patients, respectively. Ocular involvement was observed in 28.7% of patients. Neuro-Behçet was observed in 19.5% of patients. Corticosteroids were administered to 59.5% of patients, while 26.2%, 21.1%, and 2.4% of patients received cyclophosphamide, azathioprine, and infliximab, respectively.Severe fatigue was present in 19.5 % of the patients. The level of fatigue was higher in female patients (p=0.017) and in patients with vascular involvements (p=0.04).There were no significant associations with ocular(p=0.39) and neurological involvements(p=0.17). Neither the duration of the disease (p=0.88) nor the treatment type showed an association with fatigue severity: cyclophosphamide,azathioprine, colchicine, corticosteroids, and infliximab (p=1, p=1, p=0.56, p=0.2 and p=0.41 respectively).

Conclusion: Our study underscores Behçet's diverse manifestations, notably in vascular and ocular areas, with severe fatigue prevalent, especially among females and those with vascular issues, indicating the necessity for holistic management addressing symptoms and fatigue.

Behcet's disease: how does it begin?

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Introduction: Behcet's disease (BD) is a multisystem inflammatory vasculitis of unclear etiology. The disease might include several manifestations namely recurrent oral and genital ulcers, uveitis, arthritis, and others with a variable chronological sequence of occurrence. Our study aims to determine the different revealing modes of the disease.

Method: We conducted a retrospective monocentric study in our internal medicine department enrolling BD patients aged over 18 years fulfilling the international criteria for Behcet's disease from the period spanning 2021 to 2024.

Discussion: Our study enlisted 32 patients, with a male to female ratio of 2.2. The mean age upon diagnosis of the disease was 35.4 years [20-58]. Bipolar aphthous was the most common revealing mode, observed in 12 patients. Vascular involvement revealed the disease in 11 patients, among whom 3 patients presented with pulmonary embolism, 6 patients had lower limb deep vein thrombosis and one patient presented with Budd-Chiari syndrome. Hughes-Stovin syndrome was the revealing mode in one patient. Ocular involvement was the mode of disease onset in 7 patients in whom six patients exhibited bilateral panuveitis with retinal vasculitis in 5 of them. Isolated bilateral anterior uveitis was observed in one patient.

Conclusion: The discovery modes of BD are diverse and potentially severe with sometimes a life- threatening prognostic implication. Hence the importance of recognizing the varied clinical manifestations of BD for prompt diagnosis and optimal management.

Case Report of a Patient with Recurrent Clinical Neuro-Behçet's Disease and Persistently Normal MRI Findings

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Introduction: Behcet's disease is a chronic relapsing inflammatory disease of unknown etiology, characterized by recurrent episodes of remission and exacerbation of various symptoms, Parenchymal involvement is usually nonvasculitic, demyelinating lesions mainly involves the brainstem and epithalamus.

Method: case report

Discussion: Patient followed in our training for neurobehcet since 2014 retained in front of bipolar aphthosis, uveitis + neurological sign, type dysarthria and right hemyparesis, where the clinical examination found a right hemipyramidal SD + with ophthalmology of the left eye.CSF study showed pleocytosis, HLA B51 was positive,the brain MRI returned normal with no abnormalities. in 2023 admitted for the progressive installation of diplopia with oral aphthosis and dysarthria with accentuation of the balance disorder, brain MRI: without abnormalities. in 2024: the patient presents with acute onset paraparesis with diplopia and dysarthria associated with a s urinary incontinence concomitant with bipolar aphthosis with on neurological examination: parapyramidal syndrome & pseudobulbar syndrome + ophthalmoplegia of the 6th left cranial pair. In front of the spinal cord signs, a complementary spinal MRI was carried out, revealing no abnormality. 3.Disscussion: MRI studies of patients helps clinicians in understanding the neuropsychiatric abnormalities in the neuro-Behçet's disease. MRI criteria was developed for neuro-Behçet's disease by the International Study Group. the neuro-imaging features were described in detail by Kidd et al.(68%) had lower brainstem lesions and 61% had lesions involving the hemispheres. Spinal cord lesions were seen in 36% with pons lesions detected in 19%, However, the MRI findings in neuro-Behçet's can be norma.

Conclusion: Brain MRI and MRA are the best exploration methods for the detection and monitoring of parenchymal lesions of the NB. although the absence of abnormality on MRI is admissible and in no way distorts the diagnosis.

Association study of HLA-A and HLA-B alleles with clinical manifestations of Behçet's disease in a Japanese population

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Introduction: Behçet's disease (BD) is strongly associated with HLA-B*51 in many ethnic groups. HLA-A*26, is also strongly associated with BD in several ethnic groups. Additionally, some HLA-A and -B alleles are reportedly associated with BD and specific clinical manifestations. In this study, we investigated the association of HLA-A and -B alleles with clinical manifestations of BD in a Japanese population.

Method: 608 Japanese patients with BD and 744 Japanese healthy controls were enrolled. For the genotyping of HLA-A and -B alleles, we performed HLA imputation using SNP2HLA with our existing GWAS data.

Discussion: HLA-B*51 showed the most significant association with BD (Pc=1.2×10-54, odds ratio [OR]=3.72). Among HLA-A alleles, HLA-A*26 showed the most significant association with BD (Pc=1.1×10-10, OR=1.85), and the association between A*26 and BD was stronger in B*51-negative cases (Pc=6.0×10-15, OR=2.51). A*31, B*39, B*51, B*55, and B*59 were also significantly associated with the risk of BD, and A*11, A*33,
B*44, B*52, B*54, B*56, and B*67 were protective alleles. B*51 showed no significant association with specific clinical manifestations. The allele frequency of B*51 was significantly increased in male cases (31.8%) compared to female cases (22.4%) (OR=1.62). A*26 was significantly associated with the risk of ocular (OR=2.96) and neurological (OR=3.13) lesions, with a greater OR in the B*51-negative cases (OR=4.42 and 4.85, respectively). In other alleles, B*35 significantly increased the risk of ocular lesions (OR=4.44), with a greater OR in the B*51-negative cases (OR=7.25). In addition, A*02 was significantly associated with the risk of arthritis (OR=1.99), B*15 with epididymitis (OR=4.58), and A*24 with vascular lesions (OR=3.93) in the B*51-negative cases.

Conclusion: This study suggests that some HLA-A and -B alleles contribute to the risk of specific clinical manifestations of BD. To validate our findings, further studies with other independent cohorts are needed.

Autoimmune manifestations in the clinical spectrum of haploinsufficiency A20: not to be confused with the diagnosis of Behçet

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Introduction: Studies published over 30 ya suspected Mendelian genet transmission in families affected at an early age by Behçet's disease-BD.We report here the case-C of a French family with at least 3 members presenting HA20 & initially diagnosed in advance BD

Method: C1:A 66 yo woman,clinical picture-pic of a Behçet-like-BL dating back to early childhood,presenting autoimmune hepatitis-HAI in -23 on liver biopsy,treated with steroid with good evolution for the moment.C2:Her 47 yo daughter,with BL cutaneous-articular disease since the age of 6 presenting immunological thrombocytopenia in -22. She received steroid for ITP with good response.C3:A 54 yo man,brother of C1,presenting with a pic of BD revealed at an early age/10yr associated with inflammatory colitis.In 2023,he developed multiple infectious complications:yersinosis,clostridial colitis & pneumopathy,resulting in a pic of immune deficiency.Anti-TNF treatment was offered to the patient in the event of a new digestive flare-up,& despite the advanced endoscopic lesions,he remains under regular,close surveillance.HLAB51 antigen is absent in the family.Genet analysis of the TNF/AIP3 gene revealed c.1880_1881 del variant on a single allele,resulting in the modification p.Cys627Phefs*44 in the aminoAC chain of the A20 protein.This mutation,present in the heterozygous state in all 3 patients,points to autosomal dominant transmission of the disease

Discussion: HA20 clinical hallmark is early onset aphtosis dominantly inherited.Febrile inflammatory outbreaks & joint involvement affect half of all patients,uveitis are rare, AI conditions usually absent in BD can be seen (ITP,HAI). Severe digestive disease is prominent,affecting more than 2/3 of C & potentially severe & haemorragic

Conclusion: The presence of a familial history of AI or inflammatory manifestations outside the classical spectrum of BD,plus an in advance of symptoms,should alert the clinician.Early diagnosis of HA20 is key for identifying complications promptly the best treatment and follow-up strategy

Gender influence in Behçet Syndrome: a Tunisian study

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Introduction: Male gender is inconsistently reported as having more severe involvement in Behçet syndrome (BS) according to the studied ethnic group. We aimed to study gender influence in BS patients in the Tunisian context

Method: Retrospective comparative study of BS patients, followed up in the Internal Medicine and the Ophthalmology Departments of Fattouma Bourguiba University Hospital (Monastir, Tunisia), according to gender.

Discussion: A total of 449 BD patients were included: 137 females (30.5%) and 312 males (69.5%) (Sex ratio M/F was 2.27). Mean age at diagnosis was 32 ±10.5 years. Family history of BD was recorded in 12.1% of patients. Oral aphthosis was detected in 92.6 % of patients at presentation, genital ulcers in 70.5%, pseudofolliculitis in 70.6% and erythema nodosum in 9.9%. Ocular inflammation was found in 38.5% of the cases, neurological and vascular involvements were found in 12.5% and 38.1% of patients, respectively. Comparative study between the two studied groups revealed that males were more prone to develop pseudofolliculitis (74.9% vs 60.7%; p= 0.003), ocular manifestations (43.6% vs 27%; p= 10-3). Conversely, females have more frequently developped erythema nodosum, oral and genital aphtosis (17% vs 6.8%; p= 0.001), (97.1% vs 90.7%; p=0.018) and (79.4%, vs 67%; p=0.008) respectively.

Conclusion: In the Tunisian context, BS is characterized by a male predominance. Males are more prone to develop ocular manifestations whereas females are more likely to develop milder manifestations dominated by muco-cutaneous involvement.

Failure of Conventional Immunosuppressive Therapy Among Patients with Behçet Uveitis: Causes and Management Modalities

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Introduction: According to the EULAR recommendations, treatment of posterior uveitis related to Behçet disease (BD) is based on corticosteroids associated to immunosuppressants, and biological agents in a therapeutic escalation according to the severity of the condition. We aimed to assess the causes of failure of conventional therapy among patients with Behçet uveitis (BU) and to describe subsequent management modalities in a referral center in Tunisia.

Method: Retrospective descriptive study from the Ophthalmology Department of Fattouma Bourguiba University Hospital, Monastir, Tunisia, of BD patients with posterior uveitis, during the decade of 2013 – 2023 with at least a follow-up period > 6 months.

Discussion: A total of 101 patients were selected. The mean age was 31 years (range, 7–61). There were 78 males (77.2 %) and 23 females (22.8 %). All patients were treated with oral corticosteroids and immunosuppressants (IS), including Azathioprine in 97 patients (96%), Cyclosporine A in 6 patients (5.9%), and Mycophenolate Mofetil in one patient (1%). Of 101 patients, 68 (67.3 %) achieved a sustained remission during a mean follow-up period of 4 years (range: 0.9 to 10.7 years), and 33 (32.7 %) had uncontrolled disease activity. The underlying causes of treatment failure were medication-nonadherence in 4 patients (12.12 %), drug-induced severe anemia in one patient (3.03 %) and resistance to the treatment in 28 patients (84.85 %). The subsequent therapeutic approaches included switching to another IS (3.03 %), adding another IS in 9 patients (27.27 %), using periocular steroid injection in 10 patients (30.3 %) and switching to biologic agents in 26 patients (78.79 %).

Conclusion: During the study period, the rate of failure of conventional immunosuppressive therapy among patients with BU was 32.7%. The main cause of failure was uncontrolled disease activity, with subsequent use of biologic agents in most cases.

Psycho-cognitive disorders in patients with neuro-Behçet

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Introduction: Behçet disease is a systemic inflammatory disease. Parenchymal or vascular neurological damage can be observed in 17% to 44% of cases. Psychocognitive impairment is noted in 44% to 53% in cases of neurobehcet. The objective of this study is to evaluate the psycho-cognitive impairment in a sample of Casablanca patients suffering from Neuro-Behçet.

Method: This is a retrospective, descriptive study at the neurology department of Chu Ibn Rochd in Casablanca in patients diagnosed with neuro-behçet between 2016 and 2022. Each patient benefited from a neuro-psychological evaluation based on the following scores: the Mini-Mental State Examination (MMSE) and the Montreal Cognitive Assessment (MoCA). The digit memory test (Digit Span), the Rey figure, the clock test and the Trail making test (TMT A and B). The neuro-psychiatric assessment consisted of the assessment of depression (Beck Depression Inventory) and anxiety (Beck Anxiety Inventory).

Discussion: A review of 74 cases of neurobehçet showed that 87% of patients presented a deficiency particularly in memory and executive functions with relative preservation of language and visuospatial abilities. One of the explanations for cognitive disorders is dysfunction of frontal and temporal functions due to subcortical lesions. In our study, patients with a vascular form had a clear impairment of memory while those with a parenchymal form had a predominance of impairment of executive functions.

Conclusion: Our study revealed that psycho-cognitive impairment is common in patients with neuro-Behçet. The two main areas affected are memory and executive functions; Memory impairment is predominant in patients with vascular disease. On the other hand, the impairment of executive functions is more evident in patients with parenchymal damage.

Neurobehçet revelation mode

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Introduction: Behçet's disease (BD) is a rare systemic vasculitis predominantly affecting males. Neurological manifestations are not rare with a prevalence varying between 5.3% and 50%, and can even be the inaugural event of the disease in 7.5%.The objective of this development is to describe the spectrum of neurological manifestations of Behçet's disease, in order to optimize rapid recognition and therapeutic management.

Method: This is a retrospective cross-sectional study carried out in the Neurology department of the IBN ROCHD University Hospital on patients suffering from BD with neurological manifestations ranging from 2016 until November 2023. All patients met the criteria of the international group of study on Behçet's disease: the International Consensus Recommendations (ICR).

Discussion: Neurobehcet (NB) according to the ICR is all neurological damage occurring in a patient meeting the international criteria for BD, not explained by other systemic or neurological diseases, nor induced by treatment. Generally occurs 4 to 5 years after the first clinical sign of the disease explained by the neglect of the cutano-mucosal signs if it is not inaugural. The clinical picture of NB is characterized by a large polymorphism, which can be classified between damage to the CNS which arises from two main mechanisms: macrovascular venous damage (exceptionally arterial) and parenchymal damage. Rhombencephalitis, although with a predilection for the brainstem and diencephalon, and TVC are the most common disorders of NB. More rarely, cognitive disorders, internuclear ophthalmoplegia, attacks of ataxia- dysarthria, sensory disorders, abnormal movements, cranial nerve damage and strokes can be observed.

Conclusion: Neurological damage in BD is polymorphic and constitutes a serious criterion that can jeopardize the vital and functional prognosis of patients. The objective of this development is to describe the spectrum of neurological manifestations of BD, in order to optimize rapid recognition and therapeutic management.

Importance of Antibodies Against Phospholipids in the thrombotic Vascular Manifestations of Behçet's Disease

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Introduction: Behçet's syndrome (BS) is characterized by clinical heterogeneity, including vascular manifestations. Antibodies against phospholipids (aPL) are detected in various SADs and contribute to the occurrence of thrombosis. In BS, a higher frequency of aPL has been observed compared to healthy controls, although their relevance is unclear. Most thromboses in BS are attributed to endothelial damage secondary to vasculitis and respond better to immunosuppressants than to anticoagulants.

Method: Retrospective observational study of causasian patients diagnosed with BS (2013 ICBD criteria), in the Rheumatology Department of the Hospital of Leon (Spain). APL were tested, and if positive, retested after 12 weeks.

Discussion: APL were tested in 45 patients, all of Caucasian origin, 75.5% women, mean age of 40.5 ± 11.5 years, with 15.6% (7/45) testing positive (Table 1). The most frequent antibody was LA. In our sample, there were a total of 11 vasculothrombotic events in 7 patients (15%): two superficial venous thromboses and three deep vein thromboses, one myocardial infarction, one stroke, one transient ischemic attack, one patient with ischemic brain lesions on MRI, one case of pulmonary aneurysms, and one case of focal arteritis of the posterior tibial artery. The clinical characteristics of our cohort are described in Table 1. A trend towards an association between male sex and aPL positivity was observed (p=0.07). Although not statistically significant (small sample size), 71.4% of aPL (+) patients had the HLA-B51 allele, compared to 37.1% of aPL (-) patients. No relationship was found between aPL and the different clinical phenotypes, including vascular. Likewise, no association was observed between thrombotic events and the presence of aPL.

Conclusion: In our sample, there was no greater number of vascular events in patients with aPL. There appears to be a trend towards an association between male sex and HLA-B51 with aPL positivity, but studies with larger sample sizes are needed to confirm these data.

	APL (+) n=7	APL (-) n=38	p-value
Age	42.6 (37.3-58.7)	38.3 (33-44.8)	NS
Female sex	4 (57.1)	31 (81.6)	0.07
Type of Antiphospholipid Antibodies			
Lupus anticoagulant	4	-	
IgG anticardiolipin	1	-	
IgM anticardiolipin	2	-	
IgG anti-β2-glycoprotein	1	-	
IgM anti-β2-glycoprotein	0	-	
Clinical phenotypes of BD			
Mucocutaneous	7 (100)	38 (100)	NS
Articular	2 (28.6)	17 (44.7)	NS
Vascular	1 (14.3)	6 (15.8)	NS
Gastrointestinal	1 (14.3)	5 (13.2)	NS
Neurological	0 (0)	2 (5.3)	NS
Ocular	0 (0)	3 (7.9)	NS
HLAB51 positive/tested	5/7 (71.4)	13/35 (37.1)	0.103
Patients with thrombotic events	1 (14.3)	6 (15.8)	NS

Table 1. Clinical and analytical variables of 45 patients with BD tested for aPL. Values are shown as median (IQR) or number (%).

Rare Association of Behçet's Disease and Marfan Syndrome: Diagnostic and therapeutic challenge

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Introduction: Behçet's disease (BD) is an auto-inflammatory disease characterised by skin, mucous membranes and eyes, as well as vascular, neurological and cardiac disorders. Marfan syndrome (MS) is a genetic disorder of connective tissue, manifested by large stature, ligament hyperlaxity and cardiovascular abnormalities. The overlap of vascular and cardiac involvement in these two conditions can complicate diagnosis and delay treatment specific to each disease.

Method: We present here the case of a 30-year-old patient with such an association

Discussion: A 30-year-old patient initially presented with right facial hemiplegia with aphasia and dysarthria. Clinical examination revealed a high stature with ligament laxity (positive Wrist and thumb sign), right hemi-pyramidal syndrome and left central facial paralysis as well as bipolar aphthosis and pseudofolliculitis, the recurrent nature of which confirmed upon retaking history. A cerebral CT scan revealed a hemorrhagic stroke with no signs of associated vascular malformation, cerebral MRI angiography showed mesencephalo-diencephalic lesions with a left ponto-mesencephalic atrophy and lumbar puncture showed aseptic meningitis, all in favor of a Neuro-Behçet's. BD's workup included an ophthalmological examination revealing bilateral ectopia lentis, complicated by retinal detachment, and an echocardiogram showing a hypokinetic cardiomyopathy with an LVEF of 54%. The diagnoses of Neuro-Behçet's and MS with ocular involvement have been made, while the precise precise cause of its vascular and cardiac involvement remains to be determined. the patient was put on systemic corticosteroids and cyclophosphamide, followed by azathioprine, in addition to colchicine, with a favorable outcome

Conclusion: The association of BD and MS is rare but can present diagnostic and therapeutic challenges due to overlapping clinical manifestations. A multidisciplinary approach and individualized management are essential to optimize clinical outcomes in these patients

Comparison of Diagnostic Criteria in Behçet's Disease in a Moroccan population

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Introduction: Behçet's disease (BD) is a systemic vasculitis identified by recurring oral and genital ulcers, along with skin lesions. Since its initial description by Hulsi Behcet, various sets of criteria have been proposed for its diagnosis, with the latest being the ICBD criteria revised in 2014. Given the lack of specific Moroccan criteria, this study aimed to evaluate the specificity and sensitivity of Five criteria sets in a Moroccan population

Method: This study included 175 patients diagnosed with BD by expert rheumatologists and internists, recruited between December 2018 and December 2023, as well as 87 control patients who had diagnoses mimicking BD, or presented with at least one major sign of BD. The ISG (1990), ICBD (2014), revised Korean (2003), revised Japanese (2003), and Mason and Barnes (1969) diagnostic criteria for BD were applied to all patients and compared among them

Discussion: A total of 175 patients (93 men/82 women) were included in the study, with a mean age at diagnosis of 35.3+/-11.2 years. The ICBD criteria demonstrated the highest sensitivity at 94.9%, followed by the Korean criteria at 86.9%, the Japanese at 62.9%, the ISG at 60%, and Mason and Barnes at 48.6%. Conversely, the ISG criteria showed the highest specificity at 97.7%, followed by Mason and Barnes at 95.5%, the Japanese at 93.2%, the ICBD at 81.8%, and the Korean at 88.6%.

Conclusion: The ICBD criteria demonstrated the best compromise between sensitivity and specificity in a Moroccan population.

Aortic abdominal aneurysm and splanchnic vein thrombosis in Behçet's syndrome: an unusual association

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Introduction: Behçet's syndrome is a chronic, multisystem vasculitis with a relapsing and remitting course. Arterial involvement in behçet's disease is rare.This case is interesting because the patient had multiple aneurysms, rarely presenting in behçet's disease, associated with other venous thrombotic manifestations which are not devoid of several complications. It also highlights the diversified nature of vascular involvement which should be identified by the clinician in order to ensure an appropriate and early management.

Method: A 37 years old with a 5-year history of behçet's syndrome was admitted. Physical examination showed pallor,ascitis grade II, collateral circulation and edema of the right leg.

Discussion: Computed tomography showed budd-chiari syndrome, thrombosis of inferior vena cava, partially thrombosed aneurysm of the abdominal aorta measuring 15mm x 25mm x 11cm, partially thrombosed aneurysm of right common iliac artery measuring 17mm. Doppler ultrasound of lower limbs showed thrombosis. The patient was started on pulse corticosteroid therapy 1g during 3 days followed by oral corticosteroids. The patient was also started on immunosuppressive therapy and received his first cycle of cyclophosphamide. The patient responded well and was discharged. An endovascular surgery was scheduled. Vascular involvement of behçet's disease occurs late, five to ten years after its onset. The aortic localization, and above all, the risk of rupture can be serious. The optimal treatment is based on corticosteroids pulse therapy associated to immunosuppressive drugs. After the control of the inflammatory phase, endovascular surgery may be needed. Immunosuppressive treatment before and after the procedure is essential to avoid post-operative complications such as anastomotic pseudo-aneurysms.

Conclusion: Behçet's disease is a multi-systemic vasculitis of unknown etiology. Vascular involvement is rare but considered as critical sign of the clinical course of patients with Behçet's disease.

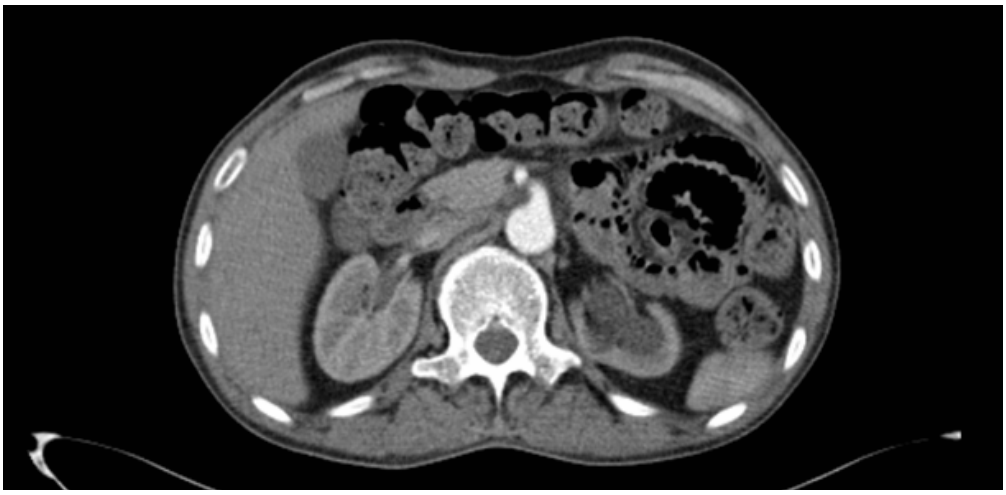

Figure 1. Fusiform abdominal aortic aneurysm.

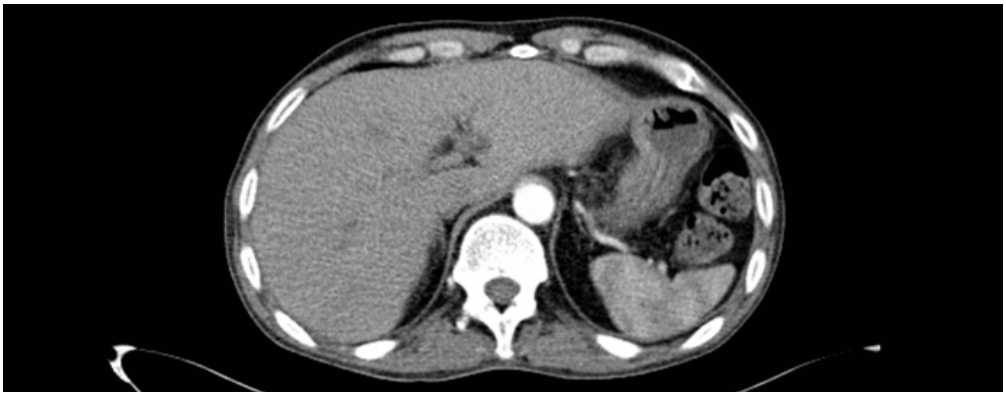

Figure 2. Lack of opacification of hepatic veins compatible with Budd-chiari syndrome.

Evaluation Of Body Image Perception and Social Appearance Anxiety in Behcet's Disease

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Introduction: Behçet's Disease (BD) is a multisystemic, inflammatory disease characterized by recurrent oral & genital ulcers. Its dermatological manifestations and frequent occurrence in young adults can lead to consequences affecting body image (BI) perception in BD. Our aim is to evaluate BI & social appearance concerns in different BD phenotypes; & to assess the relationship between BI, social appearance concerns, quality of life, anxiety, & depression.

Method: Our study was a cross-sectional survey of patients with BD & healthy controls (HC). We administered the Behçet's Disease Current Activity Form, Body Image Scale, Social Appearance Anxiety Scale, Short From-36 & Hospital Anxiety and Depression Scale.

Discussion: Worse social appearance anxiety (SAA) was observed in BD (42.74±12.16) compared to HC (27.29±17.06) (p<0.001). BI was worse in BD (6.57±7.36) compared to HC (2.41±3.93) (p<0.001). SAA was worse in patients with active disease (51.43±16.22) compared to those without active mucocutaneous involvement in the last month (40.62±8.96) (p=0.043). BI perceptions were better in patients without active symptoms in the last month (5.53±3.77) or with musculoskeletal involvement (2.28±2.05) compared to patients with multisystemic (12.76±7.48) or mucocutaneous involvement (13.06±7.46) (p<0.001).

Conclusion: SAA, BI, depression, & anxiety are worse in BD compared to HC. In patients with active mucocutaneous involvement in the last month, BI, SAA, anxiety, & depression are worse. Results indicate, despite the absence of life or organ threatening risks such as major organ involvement, mucocutaneous involvement negatively affects quality of life by creating negative BI & high social appearance anxiety in patients with BD. These observations should be considered in BD follow-up and treatment.

Figure 1. Table showing general information and survey score averages for Behçet's Patients and Healthy Control groups

		Behcet's Disease		Healthy Control	
N	Female	32	100	34	100
	Male	68		66	
Age		44,14		44,07	
Education Level	Illiterate	3		3	
	Literate	4		0	
	Primary School	29		28	
	Secondary School	25		21	
	High School	29		28	
	University or Higher Education	10		20	
BIS		6,569 ± 7,36		2,41 ± 3,93	
SAAS		42,74 ± 12,16		27,29 ± 17,06	
HADS	HADS Anxiety	8 ± 5,18	15,3 ± 9,63	4,08 ± 4,99	6,17 ± 8,09
	HADS Depression	7 ± 5,03		2,09 ± 3,46	
SF-36	Mental Health	63,04 ± 21,63		-	
	Social Functioning	65,12 ± 29,9			
CRP		6,99 ± 16,43			

BIS: Body Image Scale
HADS: Hospital Anxiety and Depression Scale
SAAS: Social Appearance Anxiety Scale
SF-36: RAND 36-Item Health Survey

Knowledge of Tunisian primary care professionals regarding Behçet disease

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Introduction: Behçet disease (BD) is a rare vasculitis more frequently found in mediterranean countries such as Tunisia. Yet, diagnostic delay is still common and tunisian patients are referred from primary care in late stages. The objective of our work was to evaluate primary care physicians knowledge regarding BD.

Method: Cross-sectional study in April 2024, based on a Google questionnaire distributed to family doctors, via social networks. Participation in this study was voluntary and responses were treated anonymously.

Discussion: Thirty-two participants responded who were distributed as follows: family medicine residents (n=18); family doctors (n=14). The average length of service in the specialty was 2,2 years [1;5]. Eighteen participants (56,2%) did a rotation in internal medicine during their residency. The approximate average number of BD patients seen during the past year was 2,3 patients [0;20] with 11 participants not seeing any (34,3%). The average response for "the epidemiological data" section was 3/5 [0;5]. The average for the "when to think of BD" was 4,37/6 [2/6]. "Manifestations of BD" mean of answers was 5,15/6 [3;6]. As for "diagnosis confirmation", the average score was 6,6/ [2;10]. For the "biological activity markers", the average was 1,5/3 [0;3]. The average response for the "immunological assessment" section was 2,2/5 [0;5]. As for "BD treatments", the average response was 2,5/4 [0;4]. For vaccination, the average response was 1,46/" [0/3]. Twenty-two participants know that BD represents a cardiovascular risk factor (68,75%). All participants judged that the management of BD patients would be better in specialized centers.

Conclusion: Participants' knowledge regarding BD manifestations was satisfying. This might be due to the internal medicine rotation in their residency. On the other hand, average responses to questions regarding vaccination weren't. A policy of collaboration between the Primary and tertiary care physicians must be studied in a more elaborate manner.

Comparison of inflammatory markers in Behçet's disease

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Introduction: Behçet's disease (BD) is a systemic variable vasculitis. Estimation of the disease activity in BD is difficult. There are no reliable laboratory markers of BD activity. Erythrocyte sedimentation rate (ESR), C - reactive protein (CRP), neutrophil- lymophocyte ratio (NLR), and systemic immune-inflammation index (SII) are known indicators of inflammation that are elevated in active Behçet's disease. The aim of this study was to compare the NLR, SII, ESR, and CRP in measuring BD activity.

Method: This study included 90 BD patients. The median age of patients was 32 years [26; 37], and the median disease duration was 11 years [5; 15]. The activity of BD was determined using the Behçet's disease current activity form (BDCAF). High disease activity was defined as a BDCAF score ≥4. NLR was calculated using the formula: neutrophils/lymphocytes. SII was calculated as neutrophil count × platelet count/lymphocyte count. A full blood count was determined on the SYSMEX XN 1000 hematology analyzer (Japan). The ESR values were measured by the Westergren method (N≤ 20 mm/h), and the CRP levels were measured by the immunonephelometric method (N≤ 5 mg/L).

Discussion: The areas under the ROC curve (AUC) for NLR, SII, CRP, and ESR were 0.721 (95% CI: 0.598–0.843), 0.705 (95% CI: 0.577–0.832), 0.787 (95% CI: 0.671–0.903), and 0.689 (95% CI: 0.556–0.821), respectively (Figure 1). The optimal cut-off values for NLR, SII, CRP, and ESR were 2.05 (60.9% sensitivity, 70.1% specificity), 571.65 (73.9% sensitivity, 62.7% specificity), 5.25 mg/L (73.9% sensitivity, 79.1% specificity), and 20.5 mm/h (52.2% sensitivity, 83.6% specificity), respectively. Fig.1 Comparison of area under ROC curves for NLR, SII, CRP, and ESR for determining the high disease activity

Conclusion: NLR, SII, ESR, and CRP are useful biomarkers for determining high BD activity. CRP had the largest area under the ROC curve (AUC=0.787). The optimal cut-off point of CRP was 5.25 mg/L with 73.9% sensitivity and 79.1% specificity.

A rare case of Behçet's disease with severe vascular involvement.

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Introduction: Vascular involvement is common in Behçet's disease and can present in a wide variety of forms. Venous involvement is much more frequent and arterial involvement is rare. We report a case of Behçet's disease with severe vascular involvement, which was revealed by rupture of an aneurysm of the internal iliac artery.

Method: We collected the patient's clinical data and medical history through a detailed review of the medical records. Behçet's disease was diagnosed according to international criteria.

Discussion: OBSERVATION A 58-year-old man with a history of stroke and recent pulmonary embolism presented with fever, abdominal pain, and shock. Initial investigations revealed an inflammatory syndrome, anemia, and elevated levels of inflammatory markers. A clinical examination revealed several genital aphthous scars. Subsequent imaging revealed pelvic hematoma with active bleeding from the internal iliac artery. Behçet's disease with severe vascular involvement was suspected and emergency surgery was performed to address the ruptured aneurysm. The patient initially responded well to corticosteroid and cyclophosphamide treatment. However, he tragically passed away two months later from unexplained cardiac arrest.

Conclusion: Arterial involvement in Behçet's disease presents a significant clinical challenge, necessitating heightened vigilance and prompt therapeutic interventions. While our patient survived aneurysm rupture, their unexplained death underscores the gravity and complexity of this disease, emphasizing the importance of close monitoring and appropriate management of cardiovascular complications in patients with Behçet's disease.

Vascular Behçet's Disease: Focus on aneurysms

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Introduction: Behçet's Disease (BD) is a rare, chronic inflammatory disorder. While deep vein thrombosis is more common, arterial involvement is a distinctive feature of BD. This study aims to delineate the epidemiological and clinical profiles of arterial aneurysms in BD.

Method: We retrospectively analyzed BD patients with arterial aneurysms over 24 years (1999-2023).

Discussion: Forty three patients were included with a male predominance (88.37%) and a mean age at onset of 38.6 years (18-59). Most patients (65.12%) had concurrent deep vein thrombosis. Multiple aneurysms were present in 20.93% of cases, primarily affecting the infrarenal abdominal aorta (30.23%) and pulmonary artery (25.58%). Iliac and femoral arteries accounted for 11.63% each, followed by popliteal artery (9.3%), subclavian and right coronary arteries (6.98% each), renal and carotid arteries (4.62% each), inferior-mesenteric, superior-mesenteric, celiac, splenic, and descending thoracic aorta (2.32% each). Abdominal pain (30%) and chest pain (18.6%) were common, along with intermittent claudication of limbs (18.6%) and hemoptysis (14%). Neurological manifestations (2%) and hemorrhagic shock (2%) were infrequent. Routine screening led to the discovery of aneurysms in 7% of cases. Concomitant mucocutaneous manifestations were found in 97.67% of patients, followed by neurological (32.56%) and ocular manifestations (27.91%), and less frequently renal and digestive manifestations (4.65% each). All patients received corticosteroids. Forty were treated with cyclophosphamide and three with azathioprine. Anti-Tumor-Necrosis-Factor agents were administered in one case resistant to first-line-therapy. Surgical interventions were performed in 18 cases. Outcomes varied significantly, with 46.5% of recovery, 13.95% of relapse and 13.95% of local complications.

Conclusion: This study underscores the heterogeneous nature of arterial aneurysm presentations in BD, emphasizing the importance of considering arterial involvement in the diagnostic algorithm.

Prevalence of Behçet's disease in 384 patients with recurrent oral aphtosis followed in liberal outpatient internal medicine

Said El Kettani, Liberal

Introduction: Behçet's disease is a systemic vasculitis, responsible for various systemic manifestations. Its etiology is unknown. Recurrent oral aphtosis is one of the three major clinical manifestations. OBJECTIVE This work was undertaken to determine the epidemiological, clinical, diagnostic and prognostic characteristics of patients with recurrent oral aphtosis according to whether they have Behçet disease or not, followed in a liberal internal medicine practice in Settat.

Method: This is a prospective descriptive study conducted from September 2009 to December 2023. It involved 384 patients with recurrent oral aphtosis. Patients are on average 38.2 11.8 years old, 57% of whom are female. The diagnosis of Behçet's disease was selected on the International Classification Criteria for Behçet's disease, revised in 2013. Statistical analysis was performed under SPSS version 20.

Discussion: The prevalence of Behçet's disease in patients with recurrent oral aphtosis is 50.5%. It is not influenced by age or sex. The group of patients with recurrent oral aphtosis with Behçet disease is significantly characterized by the exclusive presence of genital aphtosis and ocular damage ($p<0.000$) and a significantly higher frequency of skin damage ($p = 0.001$) and the positivity of the pathergic test (0.009). Regular monitoring of patients with oral aphtosis is imperative to detect Behçet disease in time.

Conclusion: The prevalence of Behçet's disease in patients with recurrent oral aphtosis is 50.5%. This prevalence is not influenced by age or gender.

Serum calprotectin as a biomarker of disease activity in Behçet's disease

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Introduction: Behçet's disease (BD) is a systemic neutrophilic vasculitis characterized by recurrent oral ulcers, genital ulcers, and internal organ damage. Calprotectin (CLP) is a marker of neutrophil activation and NETosis. Currently, there is insufficient data on the role of serum CLP in determining the activity of BD. Aim: To measure the levels of serum calprotectin in BD patients and to assess its association with disease activity.

Method: This study included 90 BD patients and 30 healthy controls. High and low disease activity were defined as BDCAF score ≥4 or <4, respectively. CLP was measured in serum by ELISA according to the manufacturer's protocol (Bulhmann Laboratories AG).

Discussion: serum CLP levels were higher in patients with BD compared to healthy controls (4.08 [2.81; 7.25] µg/mL vs. 2.86 [2.15; 3.92] µg/mL, p = 0.003). The concentration of CLP was higher in patients with high disease activity than in patients with low disease activity (6.47 [3.9; 11.68] µg/mL vs. 3.16 [2.69; 6.44] µg/mL, p = 0.003). A direct correlation was found between calprotectin and the BDCAF index (rs=0.415, p<0.0001). The sensitivity and specificity of CLP for differentiating BD patients with high disease activity from low activity patients using a cutoff value of 3.85 µg/mL were 78.3% and 53.7%, respectively. The area under the ROC curve of CLP was 0,709, 95%% CI: 0.586-0.833, p = 0.003.

Conclusion: The serum CLP levels were significantly higher in BD patients compared to controls. High levels of CLP were associated with high disease activity with 78.3% sensitivity and 53.7% specificity.

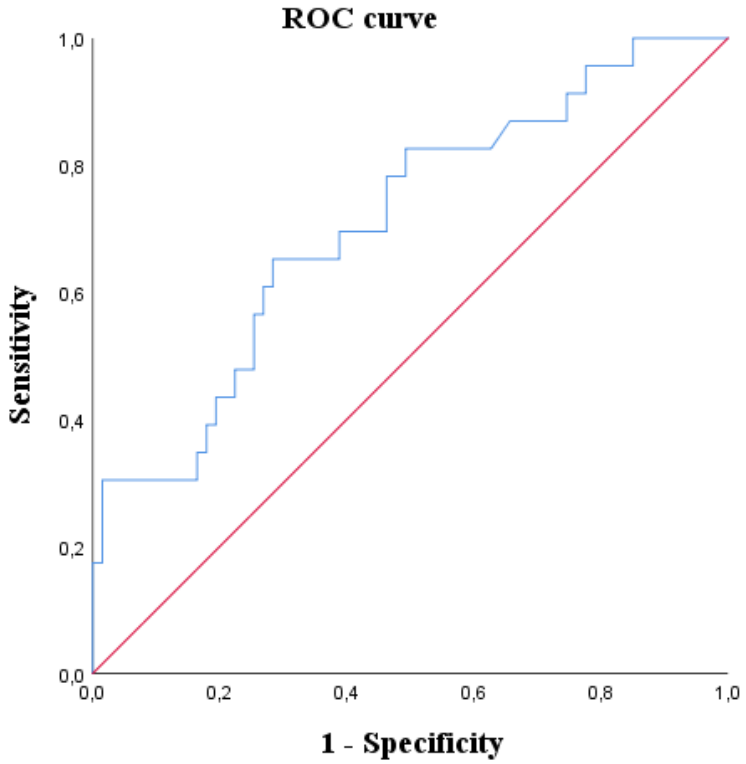

Figure 1. ROC curve of CLP for determining the high disease activity of BD.

Serum calprotectin in Behçet's disease

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Introduction: Behçet's disease (BD) is a chronic systemic vasculitis characterized by neutrophil activation and recurrent oral aphthous ulcers, genital ulcers, uveitis, and other clinical symptoms. Calprotectin (CLP) is a marker of neutrophil activation and netosis. Currently, there is insufficient data on the association of CLP with clinical and laboratory manifestations of BD. Aim: To study the relationship between high levels of CLP and manifestations of BD.

Method: This study included 90 BD patients and 30 healthy controls. The median age of patients was 32 years [26; 37], the median disease duration was 11 years [5; 15]. The activity of BD was determined using the Behçet's disease current activity Form (BDCAF). High disease activity was defined as BDCAF score ≥4. CLP was measured in serum by enzyme-linked immunosorbent assay according to the manufacturer's protocol (Bulhmann Laboratories AG, Switzerland). The upper limit of the CLP was determined by the 95th percentile of healthy control values.

Discussion: serum CLP levels were higher in patients with BD compared to healthy controls (4.08 [2.81; 7.25] μg/mL vs. 2.86 [2.15; 3.92] μg/mL, p=0.003). 23 (26%) of 90 patients with BD had elevated serum CLP levels. High CLP levels were associated with pustulosis (OR=3.41; 95% CI: 1.05-11.13, p=0.044), arthritis (OR=13.89; 95% CI: 1.47-131.82, p=0.014), uveitis (OR=4.74; 95% CI: 1.55-14.48, p=0.011), and high disease activity (OR=3.195; 95% CI: 1.149-8.887, p=0.029). CLP correlated with leukocyte count (rs=0.527, p<0.0001), neutrophil count (rs=0.656, p<0.0001), erythrocyte sedimentation rate (rs=0.357, p=0.001) and C-reactive protein (rs=0.466, p<0.0001) in patients with BD.

Conclusion: High levels of CLP were associated with the presence of pustulosis, arthritis, uveitis, and high overall clinical and laboratory activity of BD.

Serum MPO-DNA complex in Behçet's disease

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Introduction: Behçet's disease (BD) is a systemic vasculitis that is characterized by neutrophil activation and NETosis. There is limited data on the study of the specific marker of NETosis, the MPO-DNA complex, in BD. The aim of the work was to investigate the levels of MPO-DNA complex in serum in patients with BD.

Method: This study included 50 patients with BD and 20 healthy donors. The median age was 31,5 [26; 39] years, and the median disease duration was 9,5 [5; 14] years. Disease activity was assessed using Behçet's Disease Current Activity Form (BDCAF), whose values were considered low (0–1 points), moderate (2–3 points), and high (4–12 points). Serum MPO-DNA complex levels were determined via an enzyme-linked immunosorbent assay (ELISA). The reference values corresponding to the 5th percentile and 95th percentile of healthy controls were 0.0292-0.09335 OD450

Discussion: The levels of MPO-DNA complex in serum were not significantly different between patients and healthy controls (0.051 [0.043; 0.079] OD450 vs. 0.0485 [0.041; 0.060] OD450, p=0.164). Nine (18%) of 50 patients had MPO-DNA complex levels above reference values. No significant associations were found between high MPO-DNA complex concentrations and clinical manifestations or BD activity. No significant correlation was found between the MPO-DNA complex and leukocytes, neutrophils, ESR, and CRP levels in BD.

Conclusion: In our study, no significant increase in serum MPO-DNA complex was found in BD patients. In addition, no correlation between high MPO-DNA complex levels and BD manifestations was found.

NLR and SII in Behçet's disease

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Introduction: Behcet's disease (BD) is an immunoinflammatory disease characterized by recurrent oral aphthous ulcers, genital ulcers, and other clinical manifestations. Disease activity in Behçet's disease is difficult to determine due to a lack of laboratory markers that can reflect clinical activity; therefore, the search for new available markers is required. The neutrophil-lymphocyte ratio (NLR) and systemic immune- inflammation index (SII) are novel inflammatory markers. The aim of the study is to evaluate the association between NLR, SII and disease activity and clinical features of BD.

Method: This study included 90 BD patients and 30 controls. The median age of patients was 32 years [26; 37], and the median disease duration was 11 years [5; 15]. The activity of BD was determined using the Behçet's disease current activity Form (BDCAF). High disease activity was defined as a BDCAF score ≥4. NLR was calculated using the formula: neutrophils/lymphocytes. SII was calculated using the formula: neutrophils x platelets/lymphocytes. The upper limits of the NLR and SII were determined by the 95th percentile of healthy control values.

Discussion: High NLR was found in 26 (29%) and SII in 18 (20%) of 90 patients with BD. Patients with high NLR were more likely to have pustulosis (OR=6.35; 95% CI: 1.73-23.31, p=0.003), a positive pathergy test (OR=3.67; 95% CI: 1.13-11.96, p=0.035), and high disease activity (OR=13.095; 95% CI: 1.66-103.18, p=0.003). Patients with high SII were more likely to have genital ulcers (OR = 6.57; 95% CI: 1.32–32.66, p = 0.028), a positive pathergy test (OR = 4.0; 95% CI: 1.16–13.74, p = 0.048), and high BD activity (OR = 7.48; 95% CI: 0.94-59.77, p = 0.034). NLR correlated with C- reactive protein (CRP) (rs=0.42, p<0.0001) and erythrocyte sedimentation rate (ESR) (rs=0.30, p=0,004). SII also correlated with CRP (rs=0.47, p<0.0001) and ESR (rs=0.39, p<0.0001).

Conclusion: high NLR and SII levels were associated with high clinical and laboratory activity of BD.

Anticoagulant treatment in addition to immunosuppressives decreases the relapse rate in Pulmonary Arterial Involvement of Behçet's Disease

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Introduction: Vascular inflammation in Behcet's Disease (BD) is one of the most important causes of mortality due to pulmonary artery involvement (PAI) or Budd-Chiari syndrome. In this study, we aimed to retrospectively evaluate the clinical features, course and factors affecting the recurrence risk of BD-associated PAI.

Method: BD patients followed up between 1990-2022 were included. All data were acquired from the patient charts. Factors affecting the risk of relapses were determined using multivariate Cox regression analysis.

Discussion: Among 1350 BD patients, 110 (8.1%) had PAI. The mean age (SD) of patients with PAI was 42.4 (11.6) years, and the male/female ratio was 2.2 (76/34). Thirty-two (29.1%) of 110 patients were asymptomatic. Symptomatic patients were significantly older (p=0.031), and female gender (p=0.001) and recurrence (p=0.019) rates were higher than asymptomatic patients. Thrombotic involvement was seen in 104 (94.5%) and aneurysms in 9 (6.6%) patients. (Figure.1) Relapses were observed at least once in 31 (28.2%) patients. In multivariate analysis, the Cox regression model was significant (p=0.015) and not starting anticoagulants (HR 5.11, 95% CI 1.21- 21.6), p=0.026), independently increased the relapse risk. (Table.1)

Conclusion: PAT is the main presentation type of PAI in BD while aneurysmatic formation is rare. In one-third of patients with PAI, relapses develop during follow-up despite immunosuppressive treatment. When added to immunosuppressive treatment, anticoagulant therapy significantly decreases the relapse rate in BD patients with PAI.

Table 1. Factors associated with pulmonary thrombi relapses in BD patients with PAI

	Univariate		Multivariate	
Parameters	HR (95% CI)	*P* value	HR (95% CI)	*P* value
Age at the time of PAI	1.03 (1-1.06)	***0.031****	0.96 (0.91-1.01)	0.1
Female sex	1.59 (0.78-3.27)	0.205		
Age at diagnosis	1.04 (1-1.08)	0.085		
Disease duration at first PAI	1 (1-1.01)	0.285		
Smoking ever	1.13 (0.5-2.59)	0.769		
BMI	1.01 (0.93-1.1)	0.821		
Presence of symptoms at first PAI	3.09 (1.08-8.86)	***0.036****	2.45 (0.28-21.54)	0.419
Concurrent DVT	1.09 (0.47-2.56)	0.839		
Segmentary and above at CTPA	0.7 (0.34-1.43)	0.326		
Multiple involvement	1.07 (0.37-3.06)	0.907		
CRP at first PAI	1 (0.99-1.01)	0.430		
N-terminal proBNP at first PAI	1 (1-1,01)	***0.028****	1 (1-1.002)	0.254
Failure to initiation of anticoagulation	-2.85 (1.39-5.84)	***0.004****	-5.11 (1.21-21.6)	***0.026****
Initial corticosteroid dose	1 (1-1,01)	0.098		
PAH at first PAI	2.18 (1.08-4.43)	***0.030****	6.9 (0.67-71.47)	0.105

(PAI: Pulmonary arterial involvement, BMI: Body mass index, DVT: Deep vein thrombosis, CTPA: Computed tomography pulmonary angiography, CRP: C-Reactive protein, PAH: Pulmonary arterial hypertension)

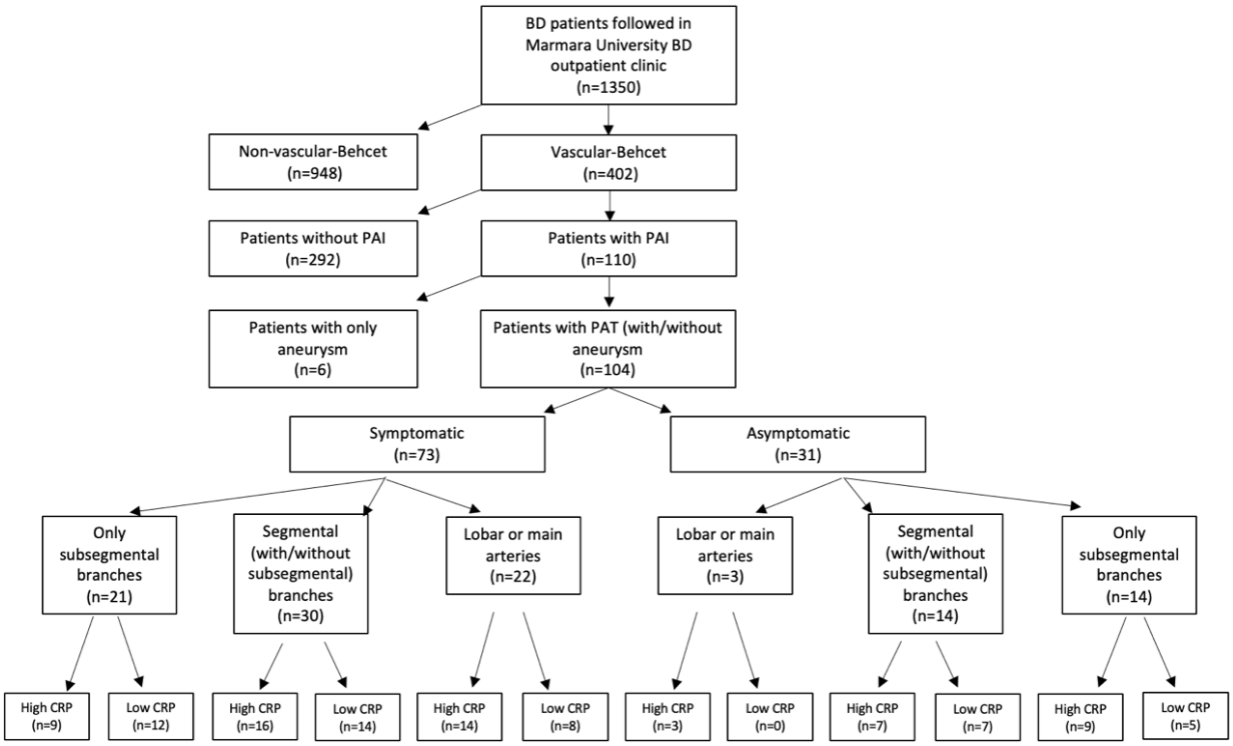

Figure 1. The distribution of patients is classified according to symptom status, size of involved vessels and acute phase reactant levels at the time of diagnosis of PAI.

Systemic Immune-Inflammation Index (SII) Role in Diagnosis of Behçet Disease

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Introduction: Behçet disease (BD) is classified as an auto-inflammatory disease in which dysregulation of the immune system can cause systemic inflammation and various end-organ damages. Since the major part of the treatment is directed toward the regulation of the immune system, it is crucial to accurately estimate the disease activity. The systemic immune-inflammation index (SII) is purposed to detect disease activity in various malignancies and rheumatologic diseases. This study aims to investigate the role of some parameters derived from complete blood cell count (CBC) including SII in patients with BD.

Method: The study was performed prospectively in the Behçet clinic of Shariati Hospital, Tehran University of Medical Sciences during 2022-2023. Accordingly, 112 BD and 61 healthy cases were enrolled in the study. SII was calculated as Neutrophil*Platelet/Lymphocyte. Subsequently, other CBC indices were calculated, as well. All the indices were compared between the groups.

Discussion: Neutrophil to lymphocyte ratio (NLR), Monocyte to RDW ratio (MONO/RDW), Neutrophil to RDW ratio (NEUT/RDW), and SII were reported to be statistically different in patients with BD and healthy controls (p-value= 0.015, 0.034, 0.001 and 0.004, respectively). Moreover, SII and Neutrophil to RDW ratio (NEUT/RDW) were significantly higher in patients with ophthalmic disease (p-value= 0.03 and 0.018, respectively). Moreover, SII was reported to be higher in BD patients with vascular involvement, yet not statistically significant (p-value= 0.07). Ultimately, using ROC analysis, a cut-off value of 538 was estimated for SII (AUC= 64.66%, SPE= 89% and SEN= 41%).

Conclusion: CBC-derived indices are feasible tools for estimating the disease activity of BD due to their high availability and low cost. This study revealed SII and Neutrophil to RDW ratio as parameters of CBC which are increased in BD patients and can be used as adjunct tools in disease diagnosis.

Betacellulin is the major ligand of epidermal growth factor receptor pathway in the exacerbation of experimental autoimmune uveoretinitis

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Introduction: Uveitis comprises various types, some of which have an acute onset, such as Behçet's disease. Recently, the epidermal growth factor receptor (EGFR) pathway has been impricated in the exacerbation of inflammatory diseases. We investigated the involvement of the EGFR pathway in experimental autoimmune uveoretinitis (EAU), an animal model of uveoretinitis.

Method: To induce EAU, 7-week-old C57BL/6 mice were immunized with interphotoreceptor retinoid binding protein derived peptide and complete Freund's adjuvant. Pertussis toxin was also injected intraperitoneally.

Discussion: EAU-induced mice were treated with intraperitoneal gefitinib, an EGFR tyrosine kinase inhibitor, and clinical and histological scores were evaluated. On day 14 after immunization, EGFR and EGF family mRNA in the retinochoroid were measured. On day 16 after immunization, betacellulin (BTC) protein level in the retinochoroid was measured by ELISA, and immunostaining of EGFR and BTC in EAU-induced mice was performed. Clinical severity of EAU was milder in gefitinib-treated mice (0.41 ±0.51) than that of untreated EAU-induced mice (3.2 ±1.1) on day 16 after immunization ($p<0.01$). The histological severity of EAU was also milder in gefitinib-treated mice (0.42 ±0.63) than that of untreated EAU-induced mice (1.6 ±0.51) on day 19 after immunization ($p<0.01$). Among EGF families, only BTC mRNA expression was upregulated in the retinochoroid of EAU-induced mice. Relative mRNA expression compared to control was 2.45 ±0.34 ($p<0.01$). Retinochoroidal protein level of BTC was significantly higher (355 ±100 pg/mg) in EAU-induced mice than that of controls (211 ±53 pg/mg, $p<0.01$). Immunostaining showed that EGFR was localized in ganglion cell layer (GCL) and BTC was localized in GCL, inner plexiform layer and inner nuclear layer in EAU-induced mice. BTC and EGFR were merged in GCL.

Conclusion: Our results suggest that BTC is the major ligand of the EGFR pathway involved in the pathogenesis of EAU in mice.

CitH3 as a specific marker of NETosis in Behçet's disease

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Introduction: Behçet's disease (BD) is a systemic variable vasculitis that is characterized by increased production of neutrophil extracellular traps (NETs). There is no data on the study of a specific marker of NETosis, citrullinated histone H3 (CitH3), in peripheral blood in BD. The aim of the work was to investigate the levels of CitH3 in serum in patients with BD.

Method: This study included 74 patients with a reliable diagnosis of BD and 30 healthy donors. The median age was 32 [27; 39] years, and the median disease duration was 10 [5; 17] years. Disease activity was assessed using Behçet's Disease Current Activity Form (BDCAF), whose values were considered low (0–1 points), moderate (2–3 points), and high (4–12 points). CitH3 was measured in serum by enzyme-linked immunosorbent assay according to the manufacturer's protocol (BlueGene Biotech, China).

Discussion: CitH3 levels were significantly higher in patients with BD compared to healthy controls (243.34 [124.38; 373.1] ng/mL vs. 106.09 [90.46; 192.09] ng/mL, p<0.0001). ROC analysis was performed to assess the diagnostic value of CitH3 in differentiating patients with BD from healthy controls.The area under the ROC curve (AUC) was 0.762, 95% CI: 0.673-0.851, p<0.0001. At a cut-off point of 238.79 ng/mL, CitH3 allows differentiation of BD patients from healthy donors with a sensitivity of 51.4% and specificity of 96.7%. Patients with moderate and high disease activity (BDCAF≥2 points) had higher CitH3 levels than those with low activity (255.01 [174.85; 390.95] ng/mL vs. 98.55 [90.99; 339.75] ng/mL, p=0.024).

Conclusion: An increase in the specific marker of NETosis, CitH3, was observed in BD patients, indicating activation of PAD4-mediated NETosis in BD.

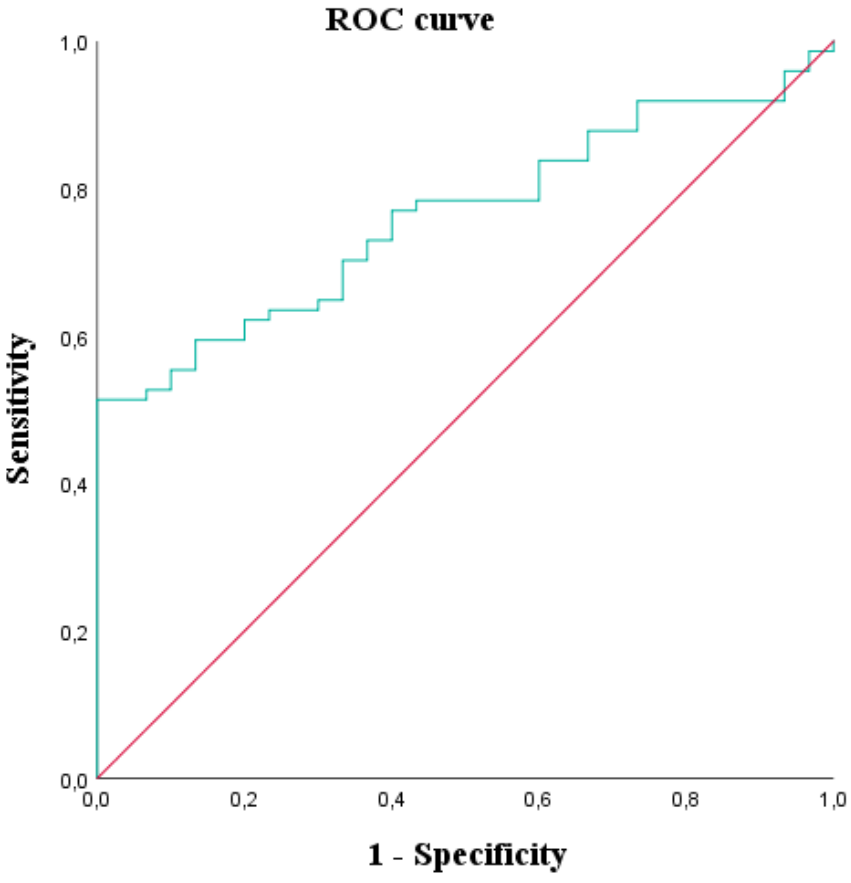

Figure 1. ROC curve of CitH3 in differentiating patients with BD from healthy controls.

Does Vein Wall Thickness have prognostic value in Behçet's Disease? A prospective follow-up study

Fatma Alibaz-Oner
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Introduction: We showed that increased CFV thickness is a distinctive feature of BD, rarely present in other inflammatory or vascular diseases with a specificity higher than 80% for the cut-off value of ≥ 0.5 mm. However, the association between CFV thickness and any organ involvement, disease course or treatment during disease course has not been demonstrated so far. This study aimed to assess the longitudinal course and prognostic value of CFV thickness measurement during a prospective follow-up.

Method: In this study, we included 195 patients with a diagnosis of BD. Bilateral CFV thickness was measured with ultrasonography (US) by an experienced radiologist at patients' routine visit day. Patients were started to follow up prospectively with 3-6 months intervals and in any urgent visit.

Discussion: Of 195 patients, 167 had prospective clinical follow-up data with a mean (SD) of 49.8 (22.3) months. New major organ involvement or relapse leading to treatment change was seen in 50 (29.9%) patients (Table 1). In 48 patients who initially had no major organ involvement, six vasculitis and one uveitis developed at the end of a mean (SD) 53.9 (28.5) month follow-up period. These seven patients had statistically significantly higher baseline CFV thicknesses compared to patients without major organ involvement, (0.87 mm vs 0.72 mm for right CFV, p=0.036 and 0.83 mm vs 0.71 mm for left CFV, p=0.161) In 47 patients, the second CFV thickness measurement was done with a mean 19.8 months after the first visit. There was no statistically significant difference between the first and second CFV wall thickness measurements for both right and left CFVs (First vs. second for right CFV: 0.79 vs. 0.76 mm, p=0.26; for left CFV: 0.79 vs. 0.75 mm, p=0.26).

Conclusion: CFV wall thickness measurement does not show a major change over time. Also, our results suggest that mucocutaneous BD patients with higher CFV thickness may have a higher risk for the development of major organ involvement during follow-up.

Predictive model for long-term visual prognosis of Behcet's uveitis using machine learning

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Introduction: Artificial intelligence will become increasingly important not only in diagnostic imaging but also in clinical data research. We investigated the independent variables of clinical data set related to visual prognosis in patients with uveitis after long-term follow-up using machine learning.

Method: A total of 102 patients with the three major etiologies of uveitis, 198 eyes (65 with sarcoidosis, 71 with Vogt-Koyanagi-Harada disease, and 62 with Behcet's disease), who had been observed for more than 100 months at our Hospital were included.

Discussion: The dependent variable was visual acuity at the last visit and the independent variables were gender, age at the first visit, visual acuity at the first examination, location of uveitis, and history of ocular complications (glaucoma, cataract, cystoid macular edema, epiretinal membrane, macular hole, vitreous hemorrhage, and rhegmatogenous retinal detachment; all of which were performed with or without surgery). Analysis was performed by machine learning using the Categorical Data Analysis Program, CATDAP.Overall, complicated cataract had the best prognostic value for visual acuity (Akaike information criteria, AIC: -31.84) followed by visual acuity at the first visit (AIC: -5.91). Sub-analysis by disease showed that the age of disease onset in sarcoidosis (AIC: -10.68), cataract in VKH disease (AIC: -18.33), and also complicated cataract in Behcet's disease (AIC: -10.78) as the factors most closely related to visual prognosis for each disease.

Conclusion: Behcet's disease had the poorest visual prognosis among 3 major etiologies of uveitis in Japan. Complication of cataract showed very high predictive performance regardless of surgical history. The presence of complicated cataract could be a representative indicator of the course of treatment, including recurrence or prolongation of ocular inflammation and total dose of corticosteroid medication.

Articular involvement during Behçet's disease in private outpatient internal medicine in 197 patients

Said El Kettani, Liberal

Introduction: Behçet's disease is a vasculitis, responsible for various systemic manifestations. Articular involvement represents one of the minor clinical manifestations. The published series concern almost exclusively patients being treated in hospital. The purpose of this study is to determine the epidemiological, clinical, diagnostic and prognostic features of articular lesions in patients treated in a private internal medicine practice in Settat.

Method: This is a prospective descriptive study conducted from September 2009 to December 2023. It involved 197 patients suffering from Behçet's disease, with an average age of 39.06 ± 11.32 years, with extremes of 15 to 70 years. The diagnosis relies on the International Classification Criteria for Behçet's disease, revised in 2013. The patients were divided into two groups, the 1st group composed of 35 patients (17.8%) with articular involvements and the 2nd group composed of 162 patients (82.2%) without articular manifestations. The statistical analysis was carried out using SPSS version 20.

Discussion: The Articular involvement occupies third place (17.8%) after mucocutaneous lesions (98.5%) and ocular manifestations (39.6%). The Patients with articular involvement are slightly older than the others (42.4 ± 9.9 years vs 38.4 ± 11.5 years (p = 0.067)). it is more prevalent among men than woen (19.8% vs 16% (p = 0.668)). The articular manifestations are: - Arthralgia in 20% of cases. - Arthritis in 42.9% of cases, they are non-deforming and non-destructive, mainly affecting the knees and ankles. - unilateral or bilateral effusion of the knees was observed in 11 patients (31.4%). The evolution under colchicine was constantly favorable.

Conclusion: Articular manifestations are observed in 17.8% of cases. They mainly affect the large joints of the lower limb. They are of arthritis type (42.9%), arthralgia (20%) and hydrarthrosis of the knee (31.4%).

Ocular manifestations of Behçet's disease (about 197 patients consulting internal medicine in a private outpatient setting)

Said El Kettani

Introduction: Behçet's disease is a systemic vasculitis. Ocular involvement represents one of the major diagnostic and prognostic criteria. The published series concern almost exclusively patients being treated in hospitals. The aim of this study is to determine the clinical, diagnostic and prognostic features of ocular involvment in patients treated in a private internal medicine practice located in Settat,a small morocan city.

Method: This is a prospective study spanned multiple years, carried out from September 2009 to December 2023. It involved 197 patients suffering from Behçet's disease, with an average age of 39.06 ± 11.32 years, with extremes of 15 to 70 years, a proportion of 53.5% is female. Behçet's disease was addressed according to the International Classification Criteria for Behçet's Disease, revised in 2013. The patients were divided into two groups, the first group composed of 78 patients (39.6%) with ocular involvement and the second group composed of 119 patients (60.4%) without ocular involvement.

Discussion: Ocular lesions occupies second place (39.6%), after mucocutaneous lesions (98.5%) and before articular manifestations (17.2%). Patients in group 1 are significantly older ($p = 0.003$), they have significantly a lower incidence of genital aphthosis ($p < 0.000$) and less positivity of the pathogenic test ($p = 0.014$). The Ocular lesion is bilateral in 41.9% of patients. Ocular manifestations are predominantly characterized by uveal involvement (90.3%). Retinal vasculitis was observed in 9.7% and exclusively in male patients. The choice of treatment depended on the type of ocular involvement. This is a particularly serious disease, 22.6% of patients experience blindness with a higher frequency observed in men.

Conclusion: Ocular involvement in an outpatient internal medicine situated, in a small (medium) city, is notable for its frequency and severity.

Behçet Disease in the Elderly

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Introduction: Behçet disease (BD) is a vasculitis of unknown etiology and pathogenesis. BD typically affects patients aged 20 to 40 years, with a male predominance. It is considered rare beyond the age of 65.

Method: A retrospective descriptive study over 18 years. All included patients were 65 years or older at the time of diagnosis. Diagnosis was established based on ISGBD and ACR 2007 criteria, after excluding differential diagnoses.

Discussion: Four elderly patients out of a total of 563 patients were identified, accounting for 0.71%. The mean age at onset of clinical signs was 58.75 years (range 47 to 66 years). Only one patient developed clinical symptoms after the age of 65; for others, the disease manifested at a younger age. Oral aphthosis was the initial manifestation in all patients, followed by genital aphthosis in 50% of cases. Ocular involvement was the most frequent presenting manifestation (75%). Necrotizing pseudo-folliculitis was found in 25% of cases, as well as asymmetric oligoarthritis of the lower limbs. Skin hypersensitivity testing was negative in all patients. Laboratory findings showed normal CBC, platelets, absence of inflammatory syndrome, normal vitamin B9 and B12, negative syphilitic, HIV, and herpetic serologies. HLA-B51 antigen testing was performed only in one patient, yielding a positive result. All patients were malnourished. Therapeutically, all patients received colchicine. Patients with ocular involvement were treated with corticosteroids/immunosuppressants. Dietary enrichment, meal fractionation, and oral nutritional supplements were recommended for all patients. All patients showed favorable outcomes under treatment.

Conclusion: BD is rare in the elderly; however, its occurrence in this age group remains possible. Treatment of BD leads to a good clinical outcome and helps prevent and/or correct malnutrition caused or exacerbated by oral aphthosis.

Clinicopathological Characteristics of Behçet disease with Severe Aortic Regurgitation

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Introduction: Behçet's disease (BD) with severe aortic regurgitation (AR) is rare but has a high mortality rate, attributed to complications like paravalvular leakage (PVL). Preoperative biologics reduce systemic inflammation and disease activity and improve prognosis, but their pathologic involvement is unknown.

Method: BD patients diagnosed with severe AR who underwent surgery at our institution were included. Pathological specimens of the aortic valve and/or aortic wall were re- analyzed based on their preoperative disease activity and treatment strategy.

Discussion: 22 BD patients with severe AR with a mean age of 39.6±13.1 years were enrolled. Seven (31.8%) underwent cardiac surgery during the active phase due to uncontrolled disease progression, while the remaining 68.2% underwent surgery during remission. Pathologically, BD with AR is characterized by mixed inflammatory cell infiltration, with higher neutrophil infiltration (60% vs. 7.7%, p=0.044) in the aortic valve of active cases. Regarding the aortic wall, acute inflammation was prominent in active cases. Detailed analysis of the three aortic layers(intima, media, and adventitia) revealed more diffuse inflammatory infiltration in the intima (40% vs. 8.3%) and media (14.3% vs. 0%), and increased neutrophil infiltration (71.4% vs. 36.4%), more frequent granulation tissue (57.1% vs. 16.7%), vasa vasorum mucoid degeneration (57.1% vs. 27.3%), vasculitis (100% vs. 72.7%)in the adventitia of active cases. Of note, preoperative biologics significantly reduced necrosis (71.4% vs. 0, p=0.023) in the aortic valves, and vasa vasorum mucoid degeneration (85.7% vs. 20%, p=0.017), neutrophil infiltration (61.5% vs. 0%), neurofibril thickening (71.4% vs. 0), and granulation tissue (42.9% vs. 0) in the aortic adventitia.

Conclusion: Overall, our study highlights the pathology of BD-induced AR as a mixed inflammatory infiltration and provides the first pathologic rationale for achieving preoperative remission and early biologic therapy to improve BD prognosis.

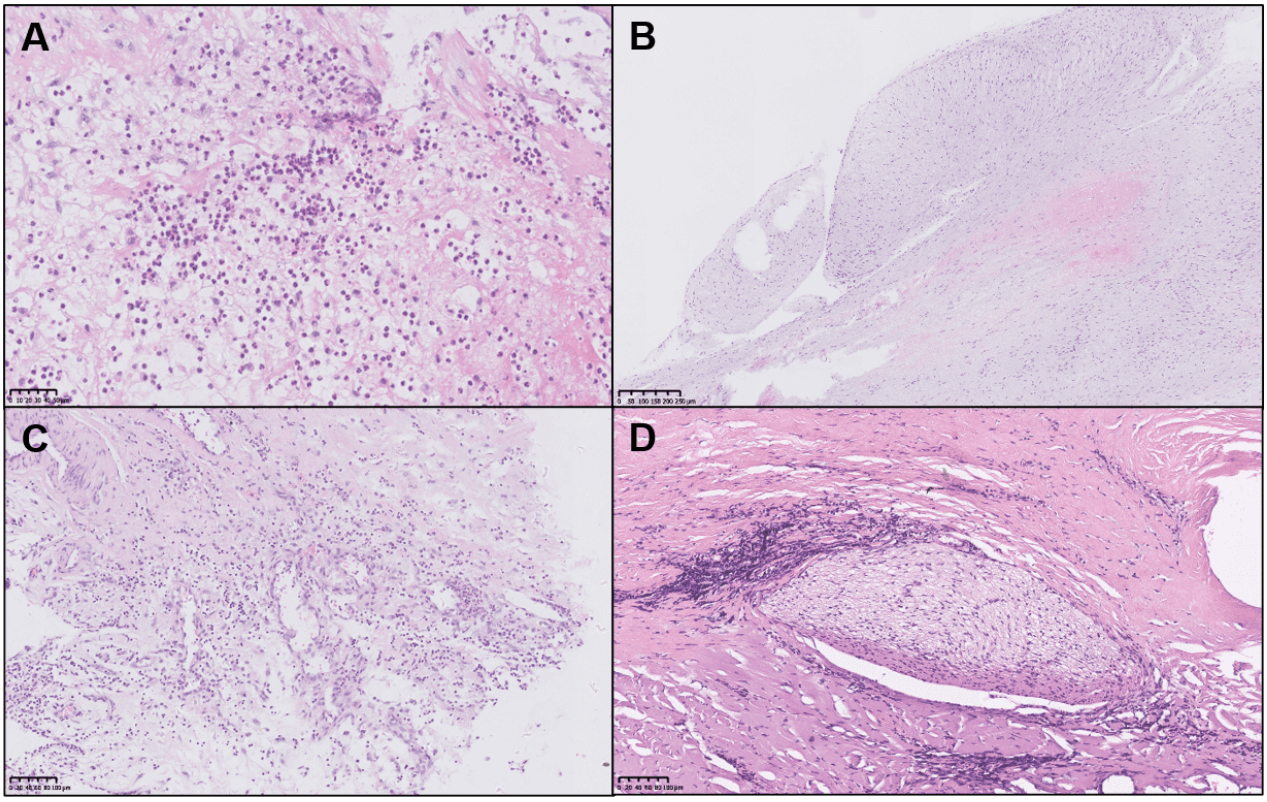

Figure 1. Representative image of aortic valve and aortic wall in active BD with AR. A, The representative image of neutrophils diffusing across the inflammatory exudate and necrosis around the valve. B, The representative image of mucoid degeneration in the aortic valve with slight necrosis. C, The representative image of neutrophils diffusing across the adventitia of aortic wall. D, The representative HE image of abnormal vasa vasorum accompanied by both sever mucoid degeneration and vasculitis with lymphocytes aggregating, wall thickening and lumen narrowing.

ABO Blood Groups and Increased Risk for Vascular Involvement in the Patients with Behçet Disease

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Introduction: Recent studies suggest an association between ABO blood groups and thrombotic vascular disease in particular in those carrying non-O (A, B, and AB) blood groups. We herein aimed to investigate the potential contribution of ABO blood groups to the risk of vascular involvement in patients with BD.

Method: Patients fulfilling the ISG Criteria for diagnosis of BD were screened, and those with information about ABO blood groups were included in the study. The presence or absence of vascular involvement and its features were recorded using a standard form.

Discussion: The study group consisted of 411 patients with available blood group data [143 (34.8%) were carrying O, and 268 (65.2%) were carrying non-O blood groups] (Table 1). There was no statistical significance between O and non-O groups regarding the potential confounding factors affecting the risk for vascular disease, including sex, age at diagnosis, family history, HLA-B51 positivity, smoking, comorbidities, and prothrombotic mutations. Vascular involvement was observed in 39 (27.3%) patients with blood group O [venous in 35 (24.5%), and arterial in 11 (7.7%)], whereas 109 patients (40.7%) with non-O blood groups had vascular involvement [venous in 95 (35.4%), and arterial in 38 (14.2%)]. The frequencies of total vascular and venous involvement between the two groups were significantly different (p=0.007, p=0.023, respectively). Unadjusted and adjusted ORs with different models in the multivariate logistic regression analyses are shown in Table 2. After adjustments for age, sex, and comorbidities, the risk for arterial disease was also found to be increased in association with non-O blood groups.

Conclusion: The results of this preliminary study support the previous reports revealing the potential contribution of ABO blood groups in the development of thrombotic vascular disease in general, and suggest an approximately two-fold increased risk for vascular involvement for BD patients for non-O blood groups.

	All patients (N=411)	O Group (N=143)	non-O Group (N=268)	p (χ^2)
Age at diagnosis, y, mean ± SD	30.94 ± 9.03	31.38 ± 10.13	30.70 ± 8.40	0.469
BD duration, m, median (IQR)	153 (98-219)	167 (107-251)	148 (92-204)	**0.047**
Male, n (%)	244 (59.4)	83 (58.0)	161 (60.1)	0.689
HLA B*51, n (%)	57‡ (53.3)	20¤ (50.0)	37§ (55.2)	0.600
Smoking, n (%)	131¥ (31.9)	44† (41.5)	87¶ (39.4)	0.711
Comorbidity, n (%)				
Diabetes mellitus	22 (5.4)	9 (6.3)	13 (4.9)	0.536
Hypertension	34 (8.3)	16 (11.2)	18 (6.7)	0.117
Hyperlipidemia	20 (4.9)	7 (4.9)	13 (4.9)	0.984
CVD	16 (3.9)	9 (6.3)	7 (2.6)	0.066
Malignancy	12 (2.9)	3 (2.1)	9 (3.4)	0.555
Genetic Trombophilia, n (%)				
FVL	22Œ (37.3)	6¢ (42.9)	16ƒ (35.6)	0.622
Protrombin Gene	10Œ (16.9)	1¢ (7.1)	9ƒ (20.0)	0.425
Combined	4Œ (6.8)	1¢ (7.1)	3ƒ (6.7)	1.000

Table 1. Characteristics of the patients. Abbreviations: BD, Behçet's Disease; CVD, cardiovascular disease; FVL, factor V leiden; y, years; m, months; N, number of evaluable patiens; n, number of patients meetings baseline situaition. (‡ N=107, ¤ N=40, § N=67, ¥ N=327, † N=106, ¶ N=221, Œ N=59, ¢ N=14, ƒ N=45)

	Overall involvement		Venous involvement		Arterial involvement	
	p	OR (CI %95)	p	OR (CI %95)	p	OR (CI %95)
Model 1	0.007	1.8 (1.2-2.8)	0.023	1.7 (1.1-2.7)	0.057	1.9 (0.9-4.0)
Model 2	0.007	1.9 (1.2-3.0)	0.024	1.7 (1.1-2.8)	0.062	1.9 (0.9-4.0)
Model 3	0.005	1.9 (1.2-3.2)	0.022	1.8 (1.1-2.9)	0.038	2.2 (1.0-4.7)
Model 4	0.006	1.9 (1.2-3.2)	0.023	1.7 (1.1-2.9)	0.046	2.1 (1.0-4.6)
Model 5	0.029	1.8 (1.1-3.2)	0.038	1.8 (1.0-3.2)	0.102	1.9 (0.9-4.5)

Abbreviations: OR, odds ratio; CI, confidence intervals

Table 2. Logistic regression analysis to estimate unadjusted and adjusted risk for vascular events comparing 0 with non-0 blood groups. Model 1: ABO blood group, Model 2: Model 1 plus age at diagnosis and sex, Model 3: Model 2 plus comorbidities, Model 4: Model 3 plus malignancy, Model 5: Model 4 plus smoking (missing value for 84 patients).

Medical practice assessment on anticoagulation in Behçet's disease (BD): Primary results from a Mediterranean survey.

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Introduction: Deep vein thrombosis during Behçet disease is an important manifestation and the underlying physiopathology is the vessel wall inflammation rather than blood clotting. Therapeutic agents are debated.

Method: A Google form questionnaire in two versions (English and French) was created. It consisted of four clinical scenarios. The questionnaire was diffused internationally, to internists via e-mail and social media.

Discussion: Fifty-one answers were collected. In the first clinical scenario: Only one doctor chose not to anticoagulate. Thirty-three chose low molecular weight heparin and vitamin K antagonists (VKA) whilst 17 opted for DOA. Seven chose anticoagulation alone.
Sixteen chose oral route glucocorticoids alone, and 28 added immunosuppressive agents with the glucocorticoids. When anticoagulation was started before admission: it was immediately discontinued (n=1). Yet, it was continued for six months (n=24), three months(n=23) or until proof of reperfusion on ultrasound (n=3). In the second clinical scenario: 29 chose anticoagulation alone using: curative anticoagulation with low molecular weight heparin and VKA (n=22) or DOA (n=7). Twenty-two chose to associate glucocorticoids and immune suppressive therapy with curative anticoagulation. In the third clinical scenario, the answers were: VKA for six months +Infliximab (n=16), long-term anticoagulation + glucocorticoids + infliximab (n=13), long-term VKA + continue Azathioprine (n=11) and anticoagulate for six months+ glucocorticoids+ continue Azathioprine (n=11). In the fourth clinical scenario, answers were: discontinue anticoagulation + high dose glucocorticoids + Cyclophosphamide (n=34), add to the latter pulmonary artery embolization (n=5), maintain anticoagulation + add high dose glucocorticoids + Cyclophosphamide (n=10) and discontinue anticoagulation but continue azathioprine (n=2).

Conclusion: Almost all answers agree on anticoagulation. The duration, the molecules and other circumstances showed diverse clinical approaches.

Behçet's Disease and Celiac Disease: Coexistence Case

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Introduction: Behçet's disease and celiac disease are both autoimmune disorders, yet they target different bodily systems. Currently, there isn't a direct proven association between these two conditions. Nevertheless, it's conceivable for an individual to concurrently experience both Behçet's disease and celiac disease, as autoimmune diseases can occasionally coincide within the same person.

Method: A 26-year-old man with a confirmed diagnosis of Behçet's disease presents with a history of recurrent oral ulcers, genital ulcers, skin lesions, and thrombosis of the median vein of the left forearm. He now reports worsening fatigue and unintentional weight loss.

Discussion: The patient's presentation raises concerns for the possible coexistence of Behçet's disease and celiac disease. Symptoms such as recurrent oral ulcers, genital ulcers, and thrombosis align with Behçet's disease, while fatigue and unintentional weight loss raise suspicion for celiac disease. Considering the patient's history of Behçet's disease, additional evaluation for concurrent celiac disease involves serological testing for celiac-specific antibodies (such as anti-tissue transglutaminase) followed by confirmation through small intestine biopsy to assess for villous atrophy and crypt hyperplasia.

Conclusion: While there are numerous hypothetical cases that could further illustrate the diverse presentations and potential overlap between Behçet's disease and celiac disease, it's essential to emphasize the necessity for additional research. Comprehensive studies are required to fully understand any potential connections between these conditions and to improve diagnosis and management strategies.

Analysis of two cases of false aneurysm in Behçet's disease

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Introduction: Behçet's disease is a vasculitis of unknown etiology. Vascular involvement is rare and dominated by venous thrombosis, but may be inaugural in many cases.

Method: we report two cases of false aneurysm of different topography revealing Behçet's disease.

Discussion: Case1: A 28-year-old patient presented with subumbilical abdominal pain. The physical examination revealed a painful beating mass to palpation. Ultrasound and angioscanographic confirmed the topography of a false aneurysm of the left iliac axis. The patient's history revealed recurrent bipolar aphthosis.Behçet disease diagnosis was made.High-dose steroid intra venous therapy and immunosuppressive was started with endovascular stent grafting of the external iliac artery opposite the false aneurysm. Case2: 40-year-old female patient with a history of bipolar aphthosis and recurrent deep vein thrombosis, presenting with a lateral cervical mass that increased in volume.Morphological assessment revealed a false carotid aneurysm.The surgical treatment, in addition to corticotherapy and immunosuppressive therapy, consisted in flattening the false aneurysm and then performing a prosthetic replacement.

Conclusion: False aneurysm remains exceptional in Behçet's disease but significantly reduces the vital prognosis of patients suffering from this vasculitis, it can be treated either surgically with a flattening procedure or endovascularly with a stent graft. Perioperative medical treatment combining corticosteroids and immunosuppressive drugs appears to reduce postoperative recurrence.

Online support group for Behçet Diseases patients: the Tunisian experience

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Introduction: Patient support groups (PSG) play a big role in patient therapeutic education in rare diseases. In Tunisia, a group dedicated to patients with Behçet disease (BD) was created a few years ago.The aim of our work was to assess the impact of PSG for Tunisian BD patients.

Method: A qualitative cross-sectional study carried out in April 2024, based on a Google Forms questionnaire distributed in the Facebook group supporting BD patients. Answers were open and treated anonymously. Participation was voluntary.

Discussion: 14 members participated. Gender ratio (M/F) was 0,27. Mean age when answering was 43,8 years [28;69]. Mean disease duration was 9,6 years [1;20]. The average group membership duration was 2.35 years [0.5;7]. Answers to the question 'Why did you join the group?" were: Looking for people with BD (n=8); helping others (n=5); better understanding BD (n=4); Looking for supplies in medicine (n=1). Answers on 'how did the group help you?' were: psychological support (n=5); Understanding better BD (n=4); not feeling alone (n=3); no help (n=3); Benefiting of medicines donations (n=3). Eight participants double-checked medical information communicated by members. Reported group limitations were: Weak members' interactivity (n=3); Absence of in-person activities (n=3); lack of financial support for patients in need (n=1). Thirteen participants consider that doctors are not present enough in the group. Expectations about doctors were: answering medical questions and giving advices (n=10); Giving scientific updates about BD and its treatments (n=4); correcting false information communicated by members (n=4); showing more psychological support (n=3); nothing (n=3); producing educational material for the group (n=1).

Conclusion: Members' expectations of the group seem to be mainly medical with a complementary role to the medical consultation. An active presence of the medical professionals could correct this false assumption and emphasize on the social and political role of these groups instead.

Subclinical herpesvirus activity prevents remission of uveitis in patients with Behcet's disease

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Introduction: Behcet's disease (BD) is a systemic variable vasculitis of unknown etiology, almost 70% of patients develop uveitis. BD pathogenesis is complex, human herpesviruses (HHV) play an important role among infectious trigger factors. Our aim was to study the possible impact of HHV reactivation: herpes simplex virus type 1 (HSV 1), type 2 (HSV 2), cytomegalovirus (CMV) and Epstein - Barr virus (EBV) on the clinical course of uveitis in BD.

Method: Serum samples of 106 BD patients (Mean age 39,2±9,88 years; 67% are men) with uveitis were examined for the presence of antibodies - serological markers of chronic HHV infection and reactivation.

Discussion: In 65 (61,3%) patients (25 with active uveitis (UA), 40 with uveitis remission (UR)), HHV reactivation was detected (mainly HSV 1, less often HSV 2, CMV and EBV - in individual cases). 41 (38,7%) patients had chronic HHV (17 with UA, 24 with UR). Certain clinical symptoms of uveitis were found to depend on HHV activity. Clinical signs of active uveitis (cells in the vitreous body), as well as severe irreversible changes (social blindness and low vision) were significantly more frequently detected in cases of HHV reactivation ($p < 0.05$) than in chronic HHV. The data obtained allow us to suggest, with some caution, that a subclinical HHV reactivation which stays after the onset of clinical uveitis remission, is an important factor of postuveal complications that are mainly manifested during remission. These complications include epiretinal fibrosis, pronounced optic nerve atrophy, and vasculitis (arterial and venous occlusion).

Conclusion: The remission period in patients with subclinical herpesvirus activity is less favorable than in patients without serological markers of HHV reactivation. HHV reactivation that persists after uveitis activity is stopped can contribute to sudden exacerbations of uveitis

The efficacy of an anti-inflammatory peptide in a mouse model of Behçet's disease induced by herpes simplex virus

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Introduction: Behçet's disease (BD) is a chronic, recurrent, intractable inflammatory disease. Despite the use of several drugs, there remains a need for new treatments due to considerations of cost and efficacy.

Method: Peptides were externally administered to mice with BD symptoms for 10 days. Symptom changes were observed and recorded. Immune organs were analyzed using flow cytometry, real-time PCR, confocal, and electron microscopy.

Discussion: The peptide suppressed IL-17 and RORγt mRNA genes in a dose-dependent manner under in vitro IL-17 induction conditions. After in vivo administration to symptomatic mice, IL-17, RORγt, and TNFα mRNA were suppressed in peripheral blood leukocytes and spleen. Additionally, the frequency of CD83-positive leukocytes was downregulated. These effects resulted in the shrinking of ulcers and improvement of symptoms in mice. In splenocyte primary culture, the peptide was delivered intracellularly and reached the mitochondria and endoplasmic reticulum. When administered to mouse skin, the peptide reached the dermis. Transmission electron microscopy analysis revealed that macrophages in the peritoneum of BD mice administered the peptide were restored to a level similar to that of macrophages in normal mice. In contrast, macrophages from untreated BD mice were filled with intracellular vesicles and were significantly larger in size. The peptide remained detectable for more than 48 hours in the peritoneum of mice.

Conclusion: A peptide was discovered that has the function of suppressing pathology-related cytokine molecules that cause the deterioration of BD symptoms. This peptide improved the symptoms of ulcers in BD mice and was found to be safe in a single toxicity test, confirming its potential to be developed as a treatment in the future.

Cytokine signature differences in major phenotypic groups of Behçet's disease

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Introduction: Behçet's disease (BD) has heterogeneous presentations including mainly mucocutaneous, vascular, and ocular manifestations, and the mechanisms associated with different phenotypes have not been clarified yet. We aimed to investigate the expression of innate and adaptive immunity-related cytokines in these 3 main BD phenotypes in active, untreated states and remission after treatment to be able to develop a cytokine-based treatment algorithm.

Method: Serum samples were isolated from 41 patients with active-BD(aBD), which consisted of 19 mucocutaneous-aBD(m-aBD), 11 ocular-aBD(o-aBD), and 11 vascular-aBD(v- aBD) patients, 35 patients in remission(rBD), and 9 healthy controls(HC). Serum levels of each cytokine were measured with sandwich ELISA and analyzed as both raw measurements and corrected levels for each one million white blood cells.

Discussion: Fourty-one aBD patients(F/M:9/32, median age 29), 35 rBD(F/M:9/26, median age 29), and 9 HC (F/M:3/6, median age 28) were enrolled. The serum IFN-gamma level was significantly higher in the aBD group than in the rBD(116 vs 92 pg/ml, p=0.022). The serum IL-35 level was significantly higher in the HC group compared to aBD and rBD(p=0.05). IL-17-related cytokines were lower in a-oBD. With treatment they increased in o-aBD but decreased in m-aBD and v-aBD patients.

Conclusion: This study supports the role of both innate and Th1-predominated adaptive immune responses in all BD phenotypes. IL-17 and Th17-related immunity seem less overt in the ocular BD group, which might explain the failure of IL-17 blockade in ocular BD. These results support further studies using more comprehensive gene expression analyses for the development of targeted treatment strategies for BD phenotypes.

Efficacy and tolerability of low-dose roflumilast in refractory oral ulcers in Behçet's disease and recurrent aphthous stomatitis

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Introduction: Recently, the efficacy of apremilast, a PDE4 inhibitor, has been confirmed for improving oral ulcers (OU) in Behçet's disease (BD). However, apremilast is not yet available in many countries including Korea, making its use difficult in practice. Roflumilast, which is used for chronic obstructive pulmonary disease, is also a PDE4 inhibitor. Therefore, verification of the efficacy of roflumilast as an alternative to apremilast is needed.

Method: We conducted a single-center, single-arm, retrospective, observational investigation of low-dose roflumilast for refractory OUs in BD and RAS with 46 subjects.

Discussion: During the 12 weeks study protocol, the subjects received roflumilast at a dosage of 0.25 mg a day. Clinical response and adverse events monitored through the study protocol. At week 12, 71.7% of patients responded positively to roflumilast, with 30.4% achieving complete remission. Adverse events were reported in 76.1% of the 46 subjects with follow-up visits, primarily gastrointestinal (71.7%) and neurological symptoms (17.4%). Among the cohort, 78.3% of patients tolerated roflumilast without discontinuation, including 15.2% with dose reduction, while 21.7% discontinued due to intolerable adverse events.

Conclusion: Roflumilast exhibited fast-acting and sustained efficacy for reducing oral ulcers in BD and RAS. Despite adverse events, primarily gastrointestinal symptoms, most were tolerable and manageable. Differences in response rates between BD and RAS warrant further investigation. While the study has limitations, including its retrospective observational nature and small sample size, it suggests roflumilast as a potential treatment alternative for refractory oral ulcers, deserving further research.

Screening for familial disease presence in first-degree relatives of Behçet's disease patients: Is measurement of common femoral vein wall thickness valuable for the diagnosis ?

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Introduction: Familial aggregation was shown in Behçet's disease (BD) previously. In this study, we aimed to assess first-degree relatives (FDRs) of BD patients for the presence of clinical symptoms and signs of BD and evaluate common femoral vein (CFV) wall thickness measurement for the diagnosis of familial cases.

Method: Patients with BD (n=129) and FDRs (n=230) of these patients were included in the study. FDRs were questioned in terms of clinical symptoms of BD by phone. Pathergy test and CFV wall thickness measurement by doppler ultrasonography (US) were performed among 111 FDRs who accepted the clinical assessment. Clinical assessment group were classified according to the criteria sets for BD. FDRs (n=111) who did not meet the criteria sets and had at least one clinical finding in addition to oral aphthae (OA) were categorized as "suspected BD" group.

Discussion: We observed increased frequency of isolated BD manifestations in FDRs of BD patients (OA: 46.5%, joint involvement: 31.3%, folliculitis: 28.3%, erythema nodosum (EN): 11.8% and genital ulcer (GU): 7%). Ten FDRs were diagnosed with BD during clinical assessment. CFV wall thickness was ≥0.5 mm cut-off value in 8 of these 10 (80%) patients. Significantly increased CFV wall thickness was observed in FDRs of BD patients fulfilling diagnostic BD criteria ($p<0.001$ for both sides) and also in those with suspected BD group ($p<0.05$ for both sides)(Table 1). Additionally, presence of OA, GU, folliculitis or EN were associated with increased CFV wall thickness ($p<0.05$ for each).

Conclusion: We confirmed a high presence of familial BD and increased isolated Behçet's manifestations in the FDRs of BD patients. We also found that there is increased CFV wall thickness in FDRs of BD patients fulfilling diagnostic criteria for BD. Increased CFV wall thickness is also associated with the number of clinical findings in FDRs with suspicious BD symptoms. Our results suggest that CFV wall thickness measurement can be used for the diagnosis of familial BD.

	Left CFV wall thickness		Right CFV wall thickness	
	Median (min-max)	**P value**	**Median (min-max)**	**P value**
Cases fulfilling ISG and/or ICBD criteria sets (n=10)	0.5 (0.3-0.9)	**<0.001**	0.5 (0.3-0.8)	**<0.001**
Cases not fulfilling ISG or ICBD criteria sets (n=96)	0.2 (0.2-0.6)		0.2 (0.2-0.5)	
Suspected BD group (n=30)	0.3 (0.2-0.5)	**0.027**	0.3 (0.2-0.5)	**0.003**
Non-suspected BD group (n=66)	0.2 (0.2-0.6)		0.2 (0.2-0.5)	

Table 1. Common femoral vein wall thickness measurements in clinical assessment group.

Evaluation of Thrombophilia Risk Factors in Behçet's Patients with Vascular Involvement

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Introduction: Although the cause of thrombosis in BD is unknown, it is considered to be a thrombosis model caused by inflammation. Our aim was to evaluate the presence of acquired or hereditary thrombophilic risk factors in BD with vascular involvement. We also aimed to determine whether the presence of thrombophilia risk factors contributes additionally to the development of vascular relapse.

Method: 225 BD patients with vascular involvement were analyzed. All data was obtained from the medical charts. Acquired/hereditary thrombosis predispositions were evaluated.

Discussion: Patients (n=225) comprised 29.3% females, 70.7% males, median age 43 (20-75). Relapse occurred in 45.3%. 382 vascular events were noted; deep vein thrombosis in 43.9%, pulmonary artery thrombosis in 38.2%. Among 141 patients fully investigated, 42.5% had ≥1 risk factor; 28.4% had hereditary, 23.4% acquired. In this group, relapse rate was 46%. Presence of risk factors was similar between relapse and non-relapse groups.. Although no significant difference was found, non-relapse group had fewer without risk factors; relapse group had higher rates of ≥2 risk

factors and total risk factors.

Conclusion: Our study is the first comprehensively evaluating both detailed acquired and hereditary thrombophilic risk factors in Behçet's disease with thrombotic vascular involvement. The presence of hereditary risk factors during thrombotic vascular incidents were found to be more frequent than in the general population but lower than in venous thromboembolisms in the general population. The presence of acquired or hereditary thrombophilic risk factors during the thrombotic events in vascular BD had no effect on the development of relapse. Deep vein thrombosis (DVT) is the most common presentation in our study despite the lower rates compared to current literature. Pulmonary arterial thrombosis was found to be the second most common presentation of vascular involvement, occurring in approximately one-third of patients.

CHARACTERISTICS OF THROMBOPHILIC FACTORS IN GROUPS

	NON-RELAPS (N=75,54%)	RELAPS (N=66, 46%)	P VALUE
Individuals without risk factors	49 (65,3)	33 (50)	0,066
Individuals with two or more risk factors	7 (9,3)	10 (15,2)	0,290
Individuals with two or more acquired risk factors	2 (2,7)	2 (3)	0,999
Individuals with two or more hereditary risk factors	0	2 (3)	0,217
Total risk factors	0,47 ± 0,74	0,65 ± 0,73	0,072

Figure 1. Characteristics of thrombophilic factors in groups.

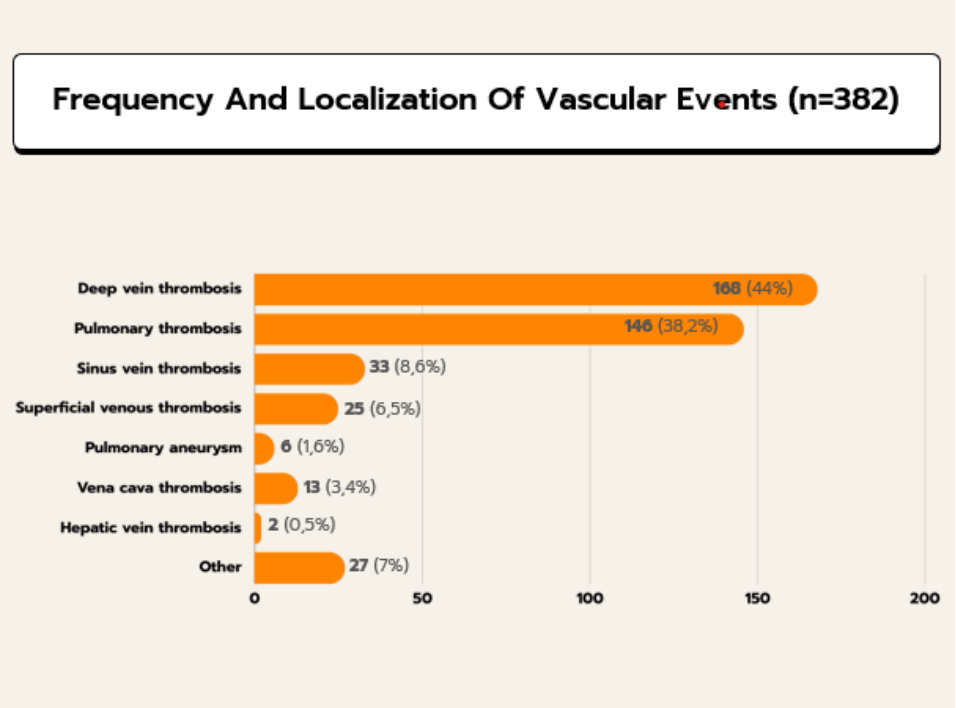

Figure 2. Frequency and localization of vascular events.