# CHAPTER 4

# BEHCET'S DISEASE (M35.2)

Nazlı CAF[1]
Defne ÖZKOCA[2]

## REMEMBER

- Behçet's disease (BD) is a chronic, recurrent, autoinflammatory and multisystemic disease. In fact, patients have a systemic vasculitis.
- Because BD is more common in countries on the Silk Road, it was formerly known as "Silk Road Disease".
- The prevalence of BD disease varies between countries and races. The prevalence ranges from 1 per 100,000 to 70 per 100,000.
- HLA-B51 and HLA-B5 genes are predisposing for the disease.
- It is known that molecules such as interleukin (IL)-1, IL-2, IL-38, IL-10, IL-18, IL-23R, IL12-RB2 play a role in the pathogenesis of BD.
- Behçet's disease is a neutrophilic disease and other neutrophilic dermatoses may accompany this disease (e.g., Sweet's syndrome)
- Recurrent oral aphthae (painful) is an indispensable parameter in the diagnostic criteria. Minor, major and/or herpetiform aphthae can be observed, but major aphthae are the most common. Oral ulcers are most commonly appear on the lips, buccal mucosa and gingiva.

[1] MD., Basaksehir Cam and Sakura City Hospital, Dermatology Clinic, drnazlicaf@gmail.com
[2] MD., Zonguldak Atatürk State Hospital, Dermatology Clinic, defneozkoca@yahoo.com

- Deep vein thrombosis and thrombophlebitis are the most common forms of vascular involvement.
- Pulmonary artery involvement can be seen and usually presents with hemoptysis.
- Superior and inferior vena cava syndromes and Budd-Chiari syndrome may develop.

### Urology

- Recurrent epididymo-orchitis attacks can be observed in male patients.
- Mucosal ulcerations may also occur in the bladder causing hematuria.
- Neurogenic bladder and related micturition disorders (such as residual urine, detrusor muscle hyperactivity, and decreased bladder compliance) may develop due to neurological involvement.

## REFERENCES


Hatemi G, Seyahi E, Fresko I, Talarico R, Hamuryudan V. One year in review 2020: Behçet's syndrome. Clin Exp Rheumatol. 2020;38(5):3–10.

Bettiol A, Prisco D, Emmi G. Behçet: The syndrome. Rheumatol (United Kingdom). 2020;59:iii101–7.

Karadag O, Bolek EC. Management of Behcet's syndrome. Rheumatol (United Kingdom). 2020;59(September 2019):iii108–17.

Talarico R, Palagini L, D'Ascanio A, Elefante E, Ferrari C, Stagnaro C, et al. Epidemiology and management of neuropsychiatric disorders in behçet's syndrome. CNS Drugs. 2015;29(3):189–96.

Dursun R, Uguz F, Kaya N, Cilli AS, Endogru H. Psychiatric disorders in patients with Behçet's disease. Int J Psychiatry Clin Pract. 2007;11(1):16–20.

Seyahi E, Yazici H. Behçet's syndrome: Pulmonary vascular disease. Curr Opin Rheumatol. 2015;27(1):18–23.

Aslam F, Bandeali SJ, Crowson C, Alam M. Cardiac Function and Diastolic Dysfunction in Behcet's Disease: A Systematic Review and Meta-Analysis. Int J Rheumatol. 2016;2016.

Mogulkoc N, Burgess MI, Bishop PW. Intracardiac thrombus in Behcet's disease: A systematic review. Chest. 2000;118(2):479–87.

Bonitsis NG, Luong Nguyen LB, Lavalley MP, Papoutsis N, Altenburg A, Kötter I, et al. Gender-specific differences in Adamantiades-Behçet's disease manifestations: An analysis of the german registry and meta-analysis of data from the literature. Rheumatol (United Kingdom). 2014;54(1):121–33.

Kai-Wen Chuang H-CC. Risk of ischaemic heart diseases and stroke in behçet disease: A systematic review and meta-analysis. Eur J Clin Invest. 2022;52(8).

Talarico R, Elefante E, Parma A, Taponeco F, Simoncini T, Mosca M. Sexual dysfunction in Behçet's syndrome. Rheumatol Int [Internet]. 2020;40(1):9–15. Available from: https://doi.org/10.1007/s00296-019-04455-w

Alizadeh F, Khorrami MH, Izadpanahi MH, Nouri-Mahdavi K, Sichani MM. Bladder involvement in Behcet's disease. Urol J. 2012;9(1):347–50.

Tong B, Liu X, Xiao J, Su G. Immunopathogenesis of Behcet's disease. Front Immunol. 2019;10(MAR):1–12.

Nelson CA, Stephen S, Ashchyan HJ, James WD, Micheletti RG, Rosenbach M. Neutrophilic dermatoses: Pathogenesis, Sweet syndrome, neutrophilic eccrine hidradenitis, and Behçet disease. J Am Acad Dermatol [Internet]. 2018;79(6):987–1006. Available from: https://doi.org/10.1016/j.jaad.2017.11.064

Vargas-Mora P, Valenzuela F, Kaplan V, Carreño L. Subcutaneous Sweet's syndrome associated with the onset of Behcet's disease. An Bras Dermatol. 2021;96(5):644–5.

Kalay Yildizhan İ, Boyvat A. Diagnostic sensitivity of different applications of pathergy test for behçet's disease. Arch Rheumatol. 2020;35(1):29–34.

BÖLÜM 5

# BEHÇET HASTALIĞI (M35.2)

Nazlı CAF[1]
Defne ÖZKOCA[2]

## HATIRLA

- Behçet Hastalığı (BH) kronik, tekrarlayıcı otoinflamatuvar ve multisistemik bir hastalıktır. Hastalarda sistemik vaskülit vardır.
- BH, İpek yolu üzerindeki ülkelerde daha sık görüldüğünden eskiden "İpek Yolu Hastalığı" olarak bilinirdi.
- Hastalık prevalansı ülkeler ve ırklar arasında farklılıklar göstermektedir. Prevalans 100.000'de 1 ile 100.000'de 70 arasında değişmektedir.
- HLA-B51 ve HLA-B5 genleri hastalık için predispozandır.
- Interlökin (IL)-1, IL-2, IL-38, IL-10, IL-18, IL-23R, IL12-RB2 gibi moleküllerin hastalık patogenezide rol oynadığı bilinmektedir.
- BH nötrofilik bir hastalıktır ve diğer nötrofilik dermatozlar bu hastalığa eşlik edebilir (ör: Sweet sendromu)
- Rekürren oral aftlar (ağrılı) tanı kritlerleri içerisinde olmazsa olmaz parametredir. Minör, major ve/veya herpetiform aftlar izlenebilir ancak en sık major aftlar görülür. Oral ülserler en sık dudaklarda, yanak mukozasında ve jijivalarda izlenir.

[1] Dr., TC Sağlık Bakanlığı Çam ve Sakura Şehir Hastanesi, Dermatoloji Kliniği, drnazlicaf@gmail.com
[2] Dr., TC Sağlık Bakanlığı Zonguldak Atatürk Devlet Hastanesi, Dermatoloji Kliniği, defneozkoca@yahoo.com

## Kardiyoloji/Kalp Damar Cerrahisi

- Behçet Hastalığı'nda kardiyak tutulum oranları değişkendir ve %7 ile %46 arasındadır.
- Hastalarda kalp kapakçık hastalıkları, endokardit, miyokardit, perikardit, intrakardiyak trombozlar, konjestif kalp yetmezliği ve myokart efraktüsü meydana gelebilir.
- Derin ven trombozu ve tromboflebit en sık vasküler tutulum şeklidir.
- Pulmoner arter tutulumu görülebilir ve genellikle hemoptizi ile prezente olur.
- Superior ve inferior vena cava sendromu ile Budd-Chiari sendromu gelişebilir.

## Üroloji

- Erkek hastalarda tekrarlayan epididimoorşit atakları izlenebilir.
- Mesane mukozasında de ülserasyonlar meydana gelebilir. Hematüri görülebilir.
- Nörolojik tutuluma bağlı nörojen mesane ve buna bağlı miksiyon bozuklukları (rezidü idrar, detrusor kas hiperaktivitesi ile mesane kompliyansında azalma gibi) meydana gelebilir.

# KAYNAKLAR


Hatemi G, Seyahi E, Fresko I, Talarico R, Hamuryudan V. One year in review 2020: Behçet's syndrome. Clin Exp Rheumatol. 2020;38(5):3–10.

Bettiol A, Prisco D, Emmi G. Behçet: The syndrome. Rheumatol (United Kingdom). 2020;59:iii101–7.

Karadag O, Bolek EC. Management of Behcet's syndrome. Rheumatol (United Kingdom). 2020;59(September 2019):iii108–17.

Talarico R, Palagini L, D'Ascanio A, Elefante E, Ferrari C, Stagnaro C, et al. Epidemiology and management of neuropsychiatric disorders in behçet's syndrome. CNS Drugs. 2015;29(3):189–96.

Dursun R, Uguz F, Kaya N, Cilli AS, Endogru H. Psychiatric disorders in patients with Behçet's disease. Int J Psychiatry Clin Pract. 2007;11(1):16–20.

Seyahi E, Yazici H. Behçet's syndrome: Pulmonary vascular disease. Curr Opin Rheumatol. 2015;27(1):18–23.

Aslam F, Bandeali SJ, Crowson C, Alam M. Cardiac Function and Diastolic Dysfunction in Behcet's Disease: A Systematic Review and Meta-Analysis. Int J Rheumatol. 2016;2016.

Mogulkoc N, Burgess MI, Bishop PW. Intracardiac thrombus in Behcet's disease: A systematic review. Chest. 2000;118(2):479–87.

Bonitsis NG, Luong Nguyen LB, Lavalley MP, Papoutsis N, Altenburg A, Kötter I, et al. Gender-specific differences in Adamantiades-Behçet's disease manifestations: An analysis of the german registry and meta-analysis of data from the literature. Rheumatol (United Kingdom). 2014;54(1):121–33.

Kai-Wen Chuang H-CC. Risk of ischaemic heart diseases and stroke in behçet disease: A systematic review and meta-analysis. Eur J Clin Invest. 2022;52(8).

Talarico R, Elefante E, Parma A, Taponeco F, Simoncini T, Mosca M. Sexual dysfunction in Behçet's syndrome. Rheumatol Int [Internet]. 2020;40(1):9–15. Available from: https://doi.org/10.1007/s00296-019-04455-w

Alizadeh F, Khorrami MH, Izadpanahi MH, Nouri-Mahdavi K, Sichani MM. Bladder involvement in Behcet's disease. Urol J. 2012;9(1):347–50.

Tong B, Liu X, Xiao J, Su G. Immunopathogenesis of Behcet's disease. Front Immunol. 2019;10(MAR):1–12.

Nelson CA, Stephen S, Ashchyan HJ, James WD, Micheletti RG, Rosenbach M. Neutrophilic dermatoses: Pathogenesis, Sweet syndrome, neutrophilic eccrine hidradenitis, and Behçet disease. J Am Acad Dermatol [Internet]. 2018;79(6):987–1006. Available from: https://doi.org/10.1016/j.jaad.2017.11.064

Vargas-Mora P, Valenzuela F, Kaplan V, Carreño L. Subcutaneous Sweet's syndrome associated with the onset of Behcet's disease. An Bras Dermatol. 2021;96(5):644–5.

Kalay Yildizhan İ, Boyvat A. Diagnostic sensitivity of different applications of pathergy test for behçet's disease. Arch Rheumatol. 2020;35(1):29–34.