



## BÖLÜM 19

# PRİMER VE SEKONDER VASKÜLTİTLER

Fatma Gülhan ŞAHBAZ<sup>1</sup>

### GİRİŞ

Vaskülit; damar duvarının inflamasyonu ile karakterize olan , iskemi ve endotel hücre nekrozuyla sonuçlanabilen, uç organ hasarına sebep olabilen ve çoğunlukla multisistemik tutulumla seyreden heterojen bir hastalıktır (1). Damardaki inflamasyon; damar duvarında kalınlaşma, lümende daralma ve skarlaşma gibi birçok fibrinoid değişiklikleri içermektedir.Vaskülitin alt tipine göre tutulan damarın boyutu, tipi ve lokasyonu farklılık göstermektedir.Vaskülitler primer olarak ortaya çıkabildikleri gibi başka hastalıklara sekonder olarak da izlenebilmektedir.Günümüzde birçok farklı sınıflama yapılmıştır. Damar çapına göre, lokasyona göre, patolojik özelliklerine göre birçok gruba ayırmak mümkündür.Vaskülit için Tanı ve Sınıflandırma Kritlerleri çalışması (DCVAS) neticesinde daha verimli sınıflandırmalar yapılması beklenmektedir (2). Vaskülitler çeşitli semptomlarla karşımıza çıkabilmektedir.Ciddi nörolojik klinik tablolara ve mortaliteye sebep olmaları nedeniyle bu hastalıkların hızlı tanı ve tedavisi önem kazanmıştır.Günümüze kadar birçok şekilde farklı sınıflandırma yapılmıştır.Aşağıdaki tablo 1 ve tablo 2'de sinir sistemini etkileyen vaskülitlerin genel sınıflandırılması ve son yapılan revizyon gösterilmektedir (2,3 ).

<sup>1</sup>Uzm. Dr., Afyonkarahisar Devlet Hastanesi, Nöroloji Kliniği, gul\_shbz@hotmail.com

Son olarak vaskülitler neoplastik ve paraneoplastik sendrom şeklinde de karşımıza çıkabilmektedir. Immun kompleks ilişkili vaskülitler (lökositoklastik vaskülit, henoç-schönlein purpura, kriyoglobulinemi), Anca ilişkili vaskülitler (mikroskopik pan, churg-strauss sendrom, granülomatöz polianjit), dev hücreli arterit, primer SSS vaskülit ve PAN bu grupta sayılabilir. Tromboembolizm, hipеркоагюлопати ve antifosfolipid anitkor pozitiflikleri eşlik edebilir (61).

## SONUÇ

Vaskülitler anlaşıldığı üzere primer veya başka bir hastalığa sekonder olarak karşımıza çıkabilmektedir. Birden fazla sistemi etkilemesi nedeniyle çok farklı klinik tablolarla prezente olabilir. Hastalığı tanıyalımak güç olmakla birlikte nonspesifik multisistem tutulumlarının birarada görüldüğü durumlarda, yardımcı testleri kullanarak, doğru tanıya ulaşmak mümkündür. Tedavide ilk seçenek olarak kortikosteroidler kullanılsa da relaps ve inflamasyonu baskılamak amacıyla antikor ilişkili olan bu otoimmun hastalıklarda immunsupresif ilaçlar sıkılıkla kullanılmaktadır. Multidisipliner tedavi yaklaşımı önerilmektedir.

## KAYNAKLAR

1. Watts RA, Robson J. Introduction, epidemiology and classification of vasculitis. Best Pract Res Clin Rheumatol. 2018 Feb;32(1):3-20. doi: 10.1016/j.berh.2018.10.003. Epub 2018 Nov 16. PMID: 30526896.
2. Craven A, Robson J, Ponte C, Grayson PC, Suppiah R, Judge A, et al. ACR/EULAR-endorsed study to develop Diagnostic and Classification Criteria for Vasculitis (DCVAS) Clin Exp NEphrol. 2013;17(5):619-21. Doi:10.1007/s10157-013-0854-0.
3. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, Flores-Suarez LF, Gross WL, Guillevin L, Hagen EC, Hoffman GS, Jayne DR, Kallenberg CG, Lamprecht P, Langford CA, Luqmani RA, Mahr AD, Matteson EL, Merkel PA, Ozen S, Pusey CD, Rasmussen N, Rees AJ, Scott DG, Specks U, Stone JH, Takahashi K, Watts RA. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum. 2013 Jan;65(1):1-11. doi: 10.1002/art.37715.
4. Byram K, Hajj-Ali RA, Calabrese L. CNS Vasculitis: an Approach to Differential Diagnosis and Management. Curr Rheumatol Rep. 2018 May 30;20(7):37. doi: 10.1007/s11926-018-0747-z. PMID: 29846828.
5. Salvarani C, Brown RD Jr, Hunder GG. Adult primary central nervous system vasculitis. Lancet. 2012 Aug 25;380(9843):767-77. doi: 10.1016/S0140-6736(12)60069-5. Epub 2012 May 9.

6. Salvarani C, Brown RD Jr, Calamia KT, Christianson TJ, Weigand SD, Miller DV, Giannini C, Meschia JF, Huston J 3rd, Hunder GG. Primary central nervous system vasculitis: analysis of 101 patients. *Ann Neurol.* 2007 Nov;62(5):442-51. doi: 10.1002/ana.21226. PMID: 17924545.
7. Calabrese LH, Duna GF, Lie JT. Vasculitis in the central nervous system. *Arthritis Rheum.* 1997 Jul;40(7):1189-201. doi: 10.1002/1529-0131(199707)40:7<1189::AID-ART2>3.0.CO;2-4.
8. Brannagan TH 3rd. Retroviral-associated vasculitis of the nervous system. *Neurol Clin.* 1997 Nov;15(4):927-44. doi: 10.1016/s0733-8619(05)70356-x. PMID: 9367973.
9. Nogueras C, Sala M, Sasal M, Viñas J, Garcia N, Bella MR, Cervantes M, Segura F. Recurrent stroke as a manifestation of primary angiitis of the central nervous system in a patient infected with human immunodeficiency virus. *Arch Neurol.* 2002 Mar;59(3):468-73. doi: 10.1001/archneur.59.3.468.
10. van der Ven AJ, van Oostenbrugge RJ, Kubat B, Tervaert JW. Cerebral vasculitis after initiation antiretroviral therapy. *AIDS.* 2002 Nov 22;16(17):2362-4. doi: 10.1097/00002030-200211220-00028.
11. Scolding NJ, Joseph F, Kirby PA, Mazanti I, Gray F, Mikol J, Ellison D, Hilton DA, Williams TL, MacKenzie JM, Xuereb JH, Love S. Abeta-related angiitis: primary angiitis of the central nervous system associated with cerebral amyloid angiopathy. *Brain.* 2005 Mar;128(Pt 3):500-15. doi: 10.1093/brain/awh379. Epub 2005 Jan 19. PMID: 15659428.
12. Salvarani C, Brown RD Jr, Calamia KT, Christianson TJ, Huston J 3rd, Meschia JF, Giannini C, Miller DV, Hunder GG. Primary central nervous system vasculitis: comparison of patients with and without cerebral amyloid angiopathy. *Rheumatology (Oxford).* 2008 Nov;47(11):1671-7. doi: 10.1093/rheumatology/ken328. Epub 2008 Aug 27.
13. Iwase T, Ojika K, Mitake S, Katada E, Katano H, Mase M, Yoshida S, Ueda R. Involvement of CD45RO+ T lymphocyte infiltration in a patient with primary angiitis of the central nervous system restricted to small vessels. *Eur Neurol.* 2001;45(3):184-5. doi: 10.1159/000052120.
14. Williams PL, Leib SL, Kamberi P, Leppert D, Sobel RA, Bifare YD, Clemons KV, Stevens DA. Levels of matrix metalloproteinase-9 within cerebrospinal fluid in a rabbit model of coccidioidal meningitis and vasculitis. *J Infect Dis.* 2002 Dec 1;186(11):1692-5. doi: 10.1086/345365. Epub 2002 Nov 1.
15. Winkler DT, Bondolfi L, Herzig MC, Jann L, Calhoun ME, Wiederhold KH, Tolnay M, Staufenbiel M, Jucker M. Spontaneous hemorrhagic stroke in a mouse model of cerebral amyloid angiopathy. *J Neurosci.* 2001 Mar 1;21(5):1619-27. doi: 10.1523/JNEUROSCI.21-05-01619.2001.
16. Giannini C, Salvarani C, Hunder G, Brown RD. Primary central nervous system vasculitis: pathology and mechanisms. *Acta Neuropathol.* 2012 Jun;123(6):759-72. doi: 10.1007/s00401-012-0973-9. Epub 2012 Mar 16. PMID: 22421812.
17. Hajj-Ali RA, Calabrese LH. Primary angiitis of the central nervous system. *Autoimmun Rev.* 2013 Feb;12(4):463-6. doi: 10.1016/j.autrev.2012.08.004. Epub 2012 Aug 16.

▲ Demyelinizan Hastalıklar

18. Hajj-Ali RA, Calabrese LH. Central nervous system vasculitis: advances in diagnosis. *Curr Opin Rheumatol.* 2020 Jan;32(1):41-46. doi: 10.1097/BOR.0000000000000676. PMID: 31652165.
19. de Boysson H, Zuber M, Naggara O, Neau JP, Gray F, et al. French Vasculitis Study Group and the French NeuroVascular Society. Primary angiitis of the central nervous system: description of the first fifty-two adults enrolled in the French cohort of patients with primary vasculitis of the central nervous system. *Arthritis Rheumatol.* 2014 May;66(5):1315-26. doi: 10.1002/art.38340. PMID: 24782189.
20. Salvarani C, Brown RD Jr, Calamia KT, Christianson TJ, Huston J 3rd, Meschia JF, Giannini C, Miller DV, Hunder GG. Primary central nervous system vasculitis presenting with intracranial hemorrhage. *Arthritis Rheum.* 2011 Nov;63(11):3598-606. doi: 10.1002/art.30594.
21. de Boysson H, Boulouis G, Dequatre N, Godard S, Néel A, et al; French Vasculitis Study Group. Tumor-Like Presentation of Primary Angiitis of the Central Nervous System. *Stroke.* 2016 Sep;47(9):2401-4. doi: 10.1161/STROKEAHA.116.013917. Epub 2016 Jul 28.
22. Messmer B, Butts M. Relapsing Primary Central Nervous System Vasculitis Treated With Rituximab. *J Clin Rheumatol.* 2020 Sep;26(6):e206-e207. doi: 10.1097/RHU.0000000000001075.
23. de Boysson H, Arquian C, Touzé E, Zuber M, Boulouis G, Naggara O, Guillemin L, Aouba A, Pagnoux C. Treatment and Long-Term Outcomes of Primary Central Nervous System Vasculitis. *Stroke.* 2018 Aug;49(8):1946-1952. doi: 10.1161/STROKEAHA.118.021878. PMID: 29986936.
24. Malani Shukla N, Lotze TE, Muscal E. Inflammatory Diseases of the Central Nervous System. *Neurol Clin.* 2021 Aug;39(3):811-828. doi: 10.1016/j.ncl.2021.04.004. Epub 2021 Jun 9.
25. Shavit E, Alavi A, Sibbald RG. Vasculitis-what do we have to know? A review of literature. *Int J Low Extrem Wounds.* 2018;17(4):218-26. doi:10.1177/1534734618804982.
26. Ferro F, Quartuccio L, Monti S, Delvino P, Di Cianni F, Fonzetti S, et al. One year in review 2021: systemic vasculitis. *Clin Exp Rheumatol.* 2021;39 Suppl 129(2):3-12.
27. Frumholtz L, Laurent-Roussel S, Lipsker D, Terrier B. Cutaneous Vasculitis: Review on Diagnosis and Clinicopathologic Correlations. *Clin Rev Allergy Immunol.* 2021 Oct;61(2):181-193. doi: 10.1007/s12016-020-08788-4.
28. Greco A, Marinelli C, Fusconi M, Macri GF, Gallo A, De Virgilio A, Zambetti G, de Vincentiis M. Clinic manifestations in granulomatosis with polyangiitis. *Int J Immunopathol Pharmacol.* 2016 Jun;29(2):151-9. doi: 10.1177/0394632015617063. Epub 2015 Dec 18. PMID: 26684637; PMCID: PMC5806708.
29. Akarken D, Tarhan EF. Granülomatöz polianjiti. Tarhan EF, editör. Küçük Damar Vaskülitleri. 1. Baskı, Ankara: Türkiye Klinikleri; 2022. p.33-41.
30. Taştekin F, Kerim D, Keser G. Eozinofilik granülomatozis polianjiti. Tarhan EF, editör. Küçük Damar Vaskülitleri. 1. Baskı, Ankara: Türkiye Klinikleri; 2022. p.42-50.
31. Chung SA, Langford CA, Maz M, Abril A, Gorelik M, Guyatt G, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the management

- of antineutrophil cytoplasmic antibody-associated vasculitis. *Arthritis Rheumatol.* 2021;73(8):1366-83. doi:10.1002/art.41773.
32. Alba MA, Espígol-Frigolé G, Prieto-González S, Tavera-Bahillo I, García-Martínez A, Butjosa M, Hernández-Rodríguez J, Cid MC. Central nervous system vasculitis: still more questions than answers. *Curr Neuropharmacol.* 2011 Sep;9(3):437-48. doi: 10.2174/157015911796557920.
  33. Greco A, De Virgilio A, Rizzo MI, Gallo A, Magliulo G, Fusconi M, Ruoppolo G, Tombolini M, Turchetta R, de Vincentiis M. Microscopic polyangiitis: Advances in diagnostic and therapeutic approaches. *Autoimmun Rev.* 2015 Sep;14(9):837-44. doi: 10.1016/j.autrev.2015.05.005. Epub 2015 May 17.
  34. Geetha D, Jefferson JA. ANCA-Associated Vasculitis: Core Curriculum 2020. *Am J Kidney Dis.* 2020 Jan;75(1):124-137. doi: 10.1053/j.ajkd.2019.04.031. Epub 2019 Jul 26.
  35. Demirci Yıldırım T, Can G. Mikroskopik polianjitit. Tarhan Ef, editör. Küçük Damar Vaskülitleri. 1. Baskı, Ankara: Türkiye Klinikleri; 2022. p.51-7.
  36. Yaseen K, Herlitz LC, Villa-Forte A. IgA Vasculitis in Adults: a Rare yet Challenging Disease. *Curr Rheumatol Rep.* 2021;23(7):50. doi:10.1007/s11926-021-01013-x.
  37. Maritati F, Canzian A, Fenaroli P, Vaglio A. Adult-onset IgA vasculitis (Henoch-Schönlein): Update on therapy. *Presse Med.* 2020;49(3):104035. doi:10.1016/j.pmed.2020.104035.
  38. Taştekin F, Karabulut G. Küçük damar vaskülitleri, IgA vaskülitleri. Tarhan Ef, editör. Küçük Damar Vaskülitleri. 1. Baskı, Ankara: Türkiye Klinikleri; 2022. p.58-64.
  39. de Boysson H, Guillemin L. Polyarteritis Nodoso Neurologic Manifestations. *Neurol Clin.* 2019 May;37(2):345-357. doi: 10.1016/j.ncl.2019.01.007. Epub 2019 Mar 16. PMID: 30952413.
  40. Karadag O, Jayne DJ. Polyarteritis nodosa revisited: a review of historical approaches, subphenotypes and a research agenda. *Clin Exp Rheumatol.* 2018 Mar-Apr;36 Suppl 111(2):135-142. Epub 2018 Feb 20. PMID: 29465365.
  41. Singh S, Jindal AK, Pilania RK. Diagnosis of Kawasaki disease. *Int J Rheum Dis.* 2018 Jan;21(1):36-44. doi: 10.1111/1756-185X.13224. Epub 2017 Nov 13.
  42. Rife E, Gedalia A. Kawasaki Disease: an Update. *Curr Rheumatol Rep.* 2020 Sep 13;22(10):75. doi: 10.1007/s11926-020-00941-4.
  43. Younger DS. Giant Cell Arteritis. *Neurol Clin.* 2019 May;37(2):335-344. doi: 10.1016/j.ncl.2019.01.008. Epub 2019 Mar 16.
  44. Keser G, Aksu K, Direskeneli H. Takayasu arteritis: an update. *Turk J Med Sci.* 2018 Aug 16;48(4):681-697. doi: 10.3906/sag-1804-136. PMID: 30114347.
  45. Wolf J, Schmitt V, Palm F, Grau AJ, Bergner R. Peripheral neuropathy as initial manifestation of primary systemic vasculitides. *J Neurol.* 2013 Apr;260(4):1061-70. doi: 10.1007/s00415-012-6760-7. Epub 2012 Dec 2
  46. Koike H. [Non-systemic Vasculitic Neuropathy]. *Brain Nerve.* 2016 Mar;68(3):233-41. Japanese. doi: 10.11477/mf.1416200383.
  47. Hadden RDM, Collins MP, Živković SA, Hsieh ST, Bonetto C, Felicetti P, Marchionne P, Santuccio C, Bonhoeffer J; Brighton Collaboration Vasculitic Peripheral Neuropathy Working Group. Vasculitic peripheral neuropathy: Case definition and guidelines for

▲ Demyelinizan Hastalıklar

- collection, analysis, and presentation of immunisation safety data. *Vaccine*. 2017 Mar 13;35(11):1567-1578. doi: 10.1016/j.vaccine.2015.11.047. Epub 2015 Dec 2.
48. Koçer N, Islak C, Siva A, Saip S, Akman C, Kantarci O, Hamuryudan V. CNS involvement in neuro-Behçet syndrome: an MR study. *AJNR Am J Neuroradiol*. 1999 Jun-Jul;20(6):1015-24. PMID: 10445437;
49. Saip S, Akman-Demir G, Siva A. Neuro-Behçet syndrome. *Handb Clin Neurol*. 2014;121:1703-23. doi: 10.1016/B978-0-7020-4088-7.00110-3.
50. Kiriakidou M, Ching CL. Systemic Lupus Erythematosus. *Ann Intern Med*. 2020 Jun 2;172(11):ITC81-ITC96. doi: 10.7326/AITC202006020.
51. Sammaritano LR. Antiphospholipid syndrome. *Best Pract Res Clin Rheumatol*. 2020 Feb;34(1):101463. doi: 10.1016/j.berh.2019.101463. Epub 2019 Dec 19.
52. Ngian GS. Rheumatoid arthritis. *Aust Fam Physician*. 2010 Sep;39(9):626-8. PMID: 20877764.
53. Makol A, Matteson EL, Warrington KJ. Rheumatoid vasculitis: an update. *Curr Opin Rheumatol*. 2015 Jan;27(1):63-70. doi: 10.1097/BOR.0000000000000126. PMID: 25405822.
54. Ienopoli S, Carsons SE. Extraglandular manifestations of primary Sjögren's syndrome. *Oral Maxillofac Surg Clin North Am*. 2014 Feb;26(1):91-9. doi: 10.1016/j.coms.2013.09.008.
55. Perzyńska-Mazan J, Maślińska M, Gasik R. Neurological manifestations of primary Sjögren's syndrome. *Reumatologia*. 2018;56(2):99-105. doi: 10.5114/reum.2018.75521. Epub 2018 May 9. PMID: 29853725; PMCID: PMC5974632.
56. Voortman M, Drent M, Baughman RP. Management of neurosarcoidosis: a clinical challenge. *Curr Opin Neurol*. 2019 Jun;32(3):475-483. doi: 10.1097/WCO.0000000000000684.
57. Bradshaw MJ, Pawate S, Koth LL, Cho TA, Gelfand JM. Neurosarcoidosis: Pathophysiology, Diagnosis, and Treatment. *Neurol Neuroimmunol Neuroinflamm*. 2021 Oct 4;8(6):e1084. doi: 10.1212/NXI.0000000000001084.
58. Cacoub P, Comarmond C, Domont F, Savey L, Saadoun D. Cryoglobulinemia Vasculitis. *Am J Med*. 2015 Sep;128(9):950-5. doi: 10.1016/j.amjmed.2015.02.017. Epub 2015 Mar 30. PMID: 25837517.
59. Teng GG, Chatham WW. Vasculitis related to viral and other microbial agents. *Best Pract Res Clin Rheumatol*. 2015 Apr;29(2):226-43. doi: 10.1016/j.berh.2015.05.007. Epub 2015 Jun 11. PMID: 26362741.
60. Radić M, Martinović Kaliterna D, Radić J. Drug-induced vasculitis: a clinical and pathological review. *Neth J Med*. 2012 Jan;70(1):12-7.
61. Park HJ, Ranganathan P. Neoplastic and paraneoplastic vasculitis, vasculopathy, and hypercoagulability. *Rheum Dis Clin North Am*. 2011 Nov;37(4):593-606. doi: 10.1016/j.rdc.2011.09.002. PMID: 22075199.