Bölüm
9

İNTRAABDOMİNAL/ RETROPERİTONEAL FİBROZİS TEDAVİSİ

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GİRİŞ VE PATOGENEZ

Retroperitoneal fibrozis(RF), fibrozis ve inflamasyonun eşlik ettiği, retroperitoneal dokunun sıklıkla abdominal organları ve yapıları (ör. abdominal aorta, üreterler) sardığı oldukça ender görülen bir hastalıktır(1).

Toplumdaki görülme sıklığı tahminen her 100.000 kişide 1.38'dir. Etnik dağılımı henüz belirlenememiştir(2). Erkeklerde kadınlara göre 2 veya 3 kat daha fazla görülmektedir. Hastalığın başlangıç yaşı genellikle 55-60 yaş arasıdır(3,4).

RF idiopatik veya sekonder olarak iki gruba ayrılabilir. İdiopatik formu %70-75 oranında görülmektedir. Sekonder grupta; maligniteler, enfeksiyonlar, travma, radyoterapi ve belirli ilaç grupları yer almaktadır(5). Idiopatik RF'nin ilk tanımını 1905'te Fransız ürolog olan Albarran yapmıştır. Albarran vakasında üreteral obstruksiyona yol açan yaygın fibrotik retroperitoneal dokunun cerrahi tedavisini raporlamıştır(6). Sonrasında, Ormond'un 1948'de iki vakasını yayınlamasıyla birlikte idiopatik RF'nin anlaşılmasında ve tedavisinde büyük gelişmeler kaydedilmiştir(7).

Yaklaşık 30 yıl önce, birkaç çalışma serisi idiopatik RF'yi kronik periaortit, inflamatuvar abdominal aort anevrizması ve perianevrizmal retroperitoneal fibrozis çatısı altında incelenebileceğine odaklanmıştır(6,7). Bu üç antitenin de histopatolojik özellikleri benzerdir.

Hastalık retroperitoneal fibrozis olarak adlandırılsa da patolojiyi tam olarak yansıtmamaktadır. Patogenezi tam olarak ortaya konulamamış olsa da son 2 dekadda yapılan çalışmalar, okside düşük dansiteli lipoproteinlere(LDL)ve B ve T lenfositler tarafından salından seroidlere(LDL oksidasyonundan oluşan lipopro-

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KAYNAKÇA

1. Gilkeson GS, Allen NB. Retroperitoneal fibrosis: a true connective tissue disease. Rheum Dis Clin North Am 1996; 22: 23–38.
2. Vaglio A, Corradi D, Manenti L, et al. Evidence of autoimmunity in chronic periaortitis: a prospective study. Am J Med 2003; 114: 454–62.
3. Kermani TA, Crowson CS, Achenbach SJ, et al: Idiopathic retroperitoneal fibrosis: a retrospective review of clinical presentation, treatment, and outcomes. Mayo Clin Proc 86: 297–303, 2011.
4. Vaglio A, Palmisano A, Alberici F, et al: Prednisone versus tamoxifen in patients with idiopathic retroperitoneal fibrosis: an open-label randomised controlled trial. Lancet 378: 338–346, 2011.
5. Koep L, Zuidema GD. The clinical significance of retroperitoneal fibrosis. Surgery 1977; 81: 250–57.
6. Albarran J. Retention renale par peri-ureterite: liberation externe de l'uretere. Assoc Fr Urol 1905; 9: 511–17.
7. Ormond JK. Bilateral ureteral obstruction due to envelopment and compression by an inflammatory process. J Urol 1948; 59: 1072–79.
8. Mitchinson MJ. Chronic periaortitis and periarteritis. Histopathology 1984; 8: 589–600.
9. Parums DV. The spectrum of chronic periaortitis. Histopathology 1990; 16: 423–31.
10. Vaglio A, Buzio C. Chronic periaortitis: a spectrum of diseases. Curr Opin Rheumatol 2005; 17: 34–40.
11. Khosroshahi A, Carruthers MN, Stone JH, et al: Rethinking Ormond's disease: "idiopathic" retroperitoneal fibrosis in the era of IgG4-related disease. Medicine (Baltimore) 92: 82–91, 2013.
12. Mahajan VS, Mattoo H, Deshpande V et al: IgG4-related disease. Annu Rev Pathol 9: 315–347, 2014.
13. Oshiro H, Ebihara Y, Serizawa H, et al. Idiopathic retroperitoneal fibrosis associated with immuno-hematological abnormalities. Am J Med 2005; 118: 782–86.
14. Galdoni M, Bonini S, Urban ML, et al. Asbestos and smoking as risk factors for idiopathic retroperitoneal fibrosis: A case control study. Ann Intern Med 2014;161:181-8.
15. Koep L, Zuidema GD. The clinical significance of retroperitoneal fibrosis. Surgery 1977; 81: 250–57.
16. Demko TM, Diamond JR, Groff J. Obstructive nephropathy as a result of retroperitoneal fibrosis: a review of its pathogenesis and associations. J Am Soc Nephrol 1997; 8: 684–88.
17. Waters VV. Hydralazine, hydrochlorothiazide and ampicillin associated with retroperitoneal fibrosis: case report. J Urol 1989; 141: 936–37.
18. Kottra JJ, Dunnick NR. Retroperitoneal fibrosis. Radiol Clin North Am 1996; 43: 1259–75.
19. Thomas MH, Chisholm GD. Retroperitoneal fibrosis associated with malignant disease. Br J Cancer 1973; 28: 453–58.
20. Seth A, Ansari MS, Trikha V, et al. Retroperitoneal fibrosis: a rare complication of Pott's disease. J Urol 2001; 166: 622–23.
21. Moul JW. Retroperitoneal fibrosis following radiotherapy for stage I testicular seminoma. J Urol 1992; 147: 124–26.
22. Corradi D, Maestri R, Palmisano A et al: Idiopathic retroperitoneal fibrosis: clinicopathologic features and differential diagnosis. Kidney Int 72: 742–753, 2007.
23. Clevenger JA, Wang M, MacLennan GT, et al: Evidence for clonal fibroblast proliferation and autoimmune process in idiopathic retroperitoneal fibrosis. Hum Pathol 43: 1875–1880, 2012.
24. Weyand CM, Kurtin PJ, Goronzy JJ: Ectopic lymphoid organogenesis: a fast track for autoimmunity. Am J Pathol 159: 787–793, 2001.
25. Zen Y, Onodera M, Inoue D, et al: Retroperitoneal fibrosis: a clinicopathologic study with respect to im- munoglobulin G4. Am J Surg Pathol 33: 1833–1839, 2009.

26. Parums DV, Chadwick DR, Mitchinson MJ: The localisation of immunoglobulin in chronic periaortitis. Atherosclerosis 61: 117– 123, 1986.
27. Ramshaw AL, Parums DV: The distribution of adhesion molecules in chronic periaortitis. Histopathology 24: 23–32, 1994.
28. Ramshaw AL, Roskell DE, Parums DV: Cytokine gene expression in aortic adventitial inflammation associated with advanced atherosclerosis (chronic periaortitis). J Clin Pathol 47: 721–727, 1994.
29. Vaglio A, Salvarani C, Buzlo C. Retroperitoneal fibrosis. Lancet. 2006 Jan 21;367(9506):241-51.
30. Yamada H, Komatsu R, Nagae H, et al. Idiopathic retroperitoneal fibrosis with duodenal obstruction successfully treated with corticosteroids. Intern Med 1998; 37: 592–98.
31. Baker LRI, Mallinson WJW, Gregory MC, et al. Idiopathic retroperitoneal fibrosis: a retrospective analysis of 60 cases. Br J Urol 1988; 60**:** 497–503.
32. van Bommel EF, Jansen I, Hendriksz TR, et al: Idiopathic retroperitoneal fibrosis: prospective evaluation of incidence and clinicoradiologic presentation. Medicine (Baltimore) 88: 193–201, 2009.
33. S Liao, Y Wang, K Li, et al(2017): Idiopathic retroperitoneal fibrosis: a cross-sectional study of 142 Chinese patients, Scandinavian Journal of Rheumatology. Doi:10.1080/03009742.2017.13 63280.
34. Vaglio A, Catanoso MG, Spaggiari L, et al. Interleukin-6 as an inflammatory mediator and target of therapy in chronic periaortitis. Arthritis Rheum 65: 2469–2475, 2013.
35. Ceresini G, Urban ML, Corradi D, et al: Association between idiopathic retroperitoneal fibrosis and autoimmune thyroiditis: a case-control study. Autoimmun Rev 14: 16–22, 2015.
36. Vaglio A, Palmisano A, Ferretti S, et al. Peripheral inflammatory arthritis in patients with chronic periaortitis: report of five cases and review of the literature. Rheumatology (Oxford) 47: 315–318, 2008.
37. Vaglio A, Greco P, Corradi D, et al. Autoimmune aspects of chronic periaortitis. Autoimmun Rev 2006;5(7);458-64.
38. Saldino RM, Palubinskas AJ. Medial placement of the ureter: a normal variant, which may simulate retroperitoneal fibrosis. J Urol 1972; 107: 582–85.
39. Vaglio A, Maritati F. İdiopathic retroperitoneal fibrosis. J Am Soc Nephrol 27: 1880–1889, 2016
40. Scheel PJ Jr, Feeley N. Retroperitoneal fibrosis. Rheum Dis Clin North Am. 2013 May;39(2):365-381.
41. Mirault T, Lambert M, Puech P, et al. Malignant retroperitoneal fibrosis: MRI characteristics in 50 patients. Medicine (Baltimore) 91: 242–250, 2012.
42. Vaglio A, Greco P, Versari A, et al: Post-treatment residual tissue in idiopathic retroperitoneal fibrosis: active residual disease or silent "scar"? A study using ^{18}F-fluorodeoxyglucose positron emission tomography. Clin Exp Rheumatol 23: 231–234, 2005.
43. Salvarani C, Pipitone N, Versari A, et al: Positron emission tomography (PET): evaluation of chronic periaortitis. Arthritis Rheum 53: 298–303, 2005.
44. Cronin CG, Lohan DG, Blake MA, et al: Retroperitoneal fibrosis: a review of clinical features and imaging findings. AJR Am J Roentgenol 191: 423–431, 2008.
45. Abe H, Morikawa T, Araki A, et al: IgG4- related periureteral fibrosis presenting as a unilateral ureteral mass. Pathol Res Pract 207: 712–714, 2011.
46. Ellimoottil C, Hart S, Mehta V, et al: Localized perirenal retroperitoneal fibrosis. Urology 81: e27–e28, 2013.
47. Van Bommel EF, Pelkmans LG, van Damme H et al: Long-term safety and efficacy of a tamoxifen-based treatment strategy for idiopathic retroperitoneal fibrosis. Eur J Intern Med 24: 444–450, 2013.
48. Pipitone N, Versari A, Vaglio A, et al: Role of ^{18}F-fluorodeoxyglucose positron emission tomography in the workup of retroperitoneal fibrosis. Clin Exp Rheumatol 29 [Suppl 64]: S72–S78, 2011.

49. Greco P, Vaglio A, Corradi D, et al: Tuberculosis as a trigger of retroperitoneal fibrosis. Clin Infect Dis 41: e72–e75, 2005.
50. Akhan SE, Dogan Y, Akhan S, et al : Pelvic actinomycosis mimicking ovarian malignancy: three cases. Eur J Gynaecol Oncol 29: 294–297, 2008.
51. Couderc M, Mathieu S, Dubost JJ, et al : Retroperitoneal fibrosis during etanercept therapy for rheumatoid arthritis. J Rheumatol 40: 1931–1933, 2013.
52. Palmisano A, Urban ML, Corradi D, et al : Chronic periaortitis with thoracic aorta and epiaortic artery involvement: a systemic large vessel vasculitis? Rheumatology (Oxford) 54: 2004–2009, 2015.
53. Mertens S, Zeegers AG, Wertheimer PA, et al: Efficacy and complications of urinary drainage proce- dures in idiopathic retroperitoneal fibrosis complicated by extrinsic ureteral obstruction. Int J Urol 21: 283–288, 2014.
54. van Bommel EF, Siemes C, Hak LE, et al : Long-term renal and patient outcome in idiopathic retroperitoneal fibrosis treated with prednisone. Am J Kidney Dis 49: 615–625, 2007.
55. van Bommel EF, Hendriksz TR, Huiskes AW, et al: Brief communication: tamoxifen therapy for nonmalignant retroperitoneal fibrosis. Ann Intern Med 144: 101–106, 2006.
56. Paz Z, Shoenfeld Y. Antifibrosis: to reverse the irreversible. Clin Rev Allergy Immunol 2010;38(2–3):276–86.
57. Scheel PJ Jr, Feeley N, Sozio SM: Combined prednisone and mycophenolate mofetil treatment for retroperitoneal fibrosis: a case series. Ann Intern Med 154: 31–36, 2011.
58. Binder M, Uhl M, Wiech T, et al: Cyclophosphamide is a highly effective and safe induction therapy in chronic periaortitis: a long-term follow-up of 35 patients with chronic periaortitis. Ann Rheum Dis 71: 311– 312, 2012.
59. Marcolongo R, Tavolini IM, Laveder F, et al: Immunosuppressive therapy for idiopathic retroperitoneal fibrosis: a retrospective analysis of 26 cases. Am J Med 116: 194–197, 2004.
60. Wallwork R, Wallace Z, Perugino C, et al. Rituximab for idiopathic and IgG4 -related retroperitoneal fibrosis. Medicine (2018) 97:42.
61. Alberici F, Palmisano A, Urban ML, et al : Methotrexate plus prednisone in patients with relapsing idiopathic retroperitoneal fibrosis. Ann Rheum Dis 72: 1584– 1586, 2013.
62. Maritati F, Corradi D, Versari A, et al: Rituximab therapy for chronic periaortitis. Ann Rheum Dis 71: 1262–1264, 2012.