

Bölüm 27

HİPEREOZİNOFİLİK SENDROM VE KARDİYOVASKÜLER TUTULUM

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GİRİŞ

Eozinofiller, doğal bağışıklık sisteminin hücreleri olup paraziter infeksiyonlara karşı savunmada önemli rol oynarlar. Hipereozinofili; bir ay ara ile en az iki farklı ölçümde periferik kanda eozinofil sayısının $> 1,500/\mu\text{L}$ olması ya da dokuda hipereozinofilinin gösterilmesi olarak tanımlanmaktadır. Hipereozinofilik sendrom (HES) ise etiyojiden bağımsız olarak persiste eden hipereozinofiliye, hedef organ hasarının eşlik etmesidir.(1)

Hipereozinofilik sendromlar, etiyojilerine göre sınıflandırılırlar:(2)

- Primer HES: Eozinofil serisinde klonal proliferasyon ile seyreden neoplastik bir tablodur.
- Sekonder HES: Reaktif HES olarak da bilinir. Diğer hücrelerden üretilen interlökin 5 (IL-5) gibi sitokinlere reaktif olarak eozinofili gelişmesidir. Advers ilaç reaksiyonları, lenfoma, paraziter infeksiyonlar, bağ doku hastalıkları ve bazı solid tümörlere eşlik edebilir. STAT-3 gen fonksiyon kazanımı mutasyonu sonucu T hücrelerinden salınan sitokinlere reaktif gelişen HES “lenfositik varyant HES” olarak tanımlanmaktadır.
- Ailesel HES: Nadir, otozomal dominant geçişli bir hastalıktır. Eozinofil sayısı yüksek olmakla birlikte fonksiyonlarında azalma mevcut olduğu için HES kliniği genellikle siliktir. Vakaların çoğunda IL-5 regülasyonundan sorumlu 5q31-5q33 lokusunda mutasyon mevcuttur.

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devam ettiği sürece ve ventriküler trombüs sebat ettiği müddetçe antikoagulan tedaviye devam edilmelidir.(12)

Sonuç

Hipereozinofilik sendrom persistan hipereozinofiliye hedef organ hasarının eşlik ettiği klinik tablodur. Kardiyovasküler tutulum hipereozinofilik sendromda önemli bir morbidite ve mortalite nedenidir. Kardiyovasküler sistem tutulumu kardiyak tutulum ve tromboembolik olaylar ile prezente olabilmektedir. Kardiyoloji uzmanları HES vakalarında kardiyovasküler bulguların olabileceğinin farkında olmalıdır ve HES olguları Kardiyoloji uzmanları ile birlikte takip edilmelidir.

KAYNAKLAR

1. Valent P, Gleich GJ, Reiter A, et al. Pathogenesis and classification of eosinophil disorders: a review of recent developments in the field. *Expert Rev Hematol.* 2012; 5(2):157-76.
2. Dispenza MC, Bochner BS. Diagnosis and Novel Approaches to the Treatment of Hypereosinophilic Syndromes. *Curr Hematol Malig Rep.* 2018 Jun;13(3):191-201. doi: 10.1007/s11899-018-0448-8.
3. Crane MM, Chang CM, Kobayashi MG, et al. Incidence of myeloproliferative hypereosinophilic syndrome in the United States and an estimate of all hypereosinophilic syndrome incidence. *J Allergy Clin Immunol* 2010; 126:179.
4. Weller PF, Bubley GJ. The idiopathic hypereosinophilic syndrome. *Blood* 1994; 83:2759.
5. Ogbogu PU, Bochner BS, Butterfield JH, et al. Hypereosinophilic syndrome: a multicenter, retrospective analysis of clinical characteristics and response to therapy. *J Allergy Clin Immunol* 2009; 124:1319.
6. Legrand F, Renneville A, Macintyre E, et al. The Spectrum of FIP1L1-PDGFR α -Associated Chronic Eosinophilic Leukemia: New Insights Based on a Survey of 44 Cases. *Medicine (Baltimore)* 2013.
7. Parrillo JE, Borer JS, Henry WL, et al. The cardiovascular manifestations of the hypereosinophilic syndrome. Prospective study of 26 patients, with review of the literature. *Am J Med Oct;1979 67(4):572-582.*
8. D'Souza MG, Swistel DG, Castro JL, et al. Hypereosinophilic thrombus causing aortic stenosis and myocardial infarction. *Ann Thorac Surg Nov;2003 76(5):1725-1726.*
9. Cools J, DeAngelo DJ, Gotlib J, et al. A tyrosine kinase created by fusion of the PDGFR α and FIP1L1 genes as a therapeutic target of imatinib in idiopathic hypereosinophilic syndrome. *N Engl J Med Mar 27;2003 348(13):1201-1214.*
10. Young JD, Peterson CG, Venge P, et al. Mechanism of membrane damage mediated by human eosinophil cationic protein. *Nature Jun 5-11;1986 321(6070):613-616.*
11. Tai PC, Ackerman SJ, Spry CJ, et al. Deposits of eosinophil granule proteins in cardiac tissues of patients with eosinophilic endomyocardial disease. *Lancet Mar21;1987 1(8534):643-647.*

12. Ogbogu PU, Rosing DR, Horne MK 3rd. Cardiovascular manifestations of hypereosinophilic syndromes. *Immunol Allergy Clin North Am*. 2007 Aug;27(3):457-75. doi: 10.1016/j.iac.2007.07.001.
13. Salanitri GC. Endomyocardial fibrosis and intracardiac thrombus occurring in idiopathic hypereosinophilic syndrome. *AJR Am J Roentgenol* May;2005 184(5):1432-1433.
14. Hendren WG, Jones EL, Smith MD. Aortic and mitral valve replacement in idiopathic hypereosinophilic syndrome. *Ann Thorac Surg* Nov;1988 46(5):570-571.
15. Subhash HS, Asishkumar M, Jonathan M. Unusual cardiac manifestation of hypereosinophilic syndrome. *Postgrad Med J* Aug;2002 78(922):490-491.
16. Ommen SR, Seward JB, Tajik AJ. Clinical and echocardiographic features of hypereosinophilic syndromes. *Am J Cardiol* Jul 1;2000 86(1):110-113.
17. Shah R, Ananthasubramaniam K. Evaluation of cardiac involvement in hypereosinophilic syndrome: complementary roles of transthoracic, transesophageal, and contrast echocardiography. *Echocardiography* Sep;2006 23(8):689-691.
18. Syed IS, Martinez MW, Feng DL, et al. Cardiac magnetic resonance imaging of eosinophilic endomyocardial disease. *Int J Cardiol*. Mar 29;2007
19. Wagner A, Mahrholdt H, Holly TA, et al. Contrast-enhanced MRI and routine single photon emission computed tomography (SPECT) perfusion imaging for detection of subendocardial myocardial infarcts: an imaging study. *Lancet* Feb 1;2003 361(9355):374-379.
20. Fauci AS, Harley JB, Roberts WC et al. NIH conference. The idiopathic hypereosinophilic syndrome. Clinical, pathophysiologic, and therapeutic considerations. *Ann Intern Med* Jul;1982 97(1):78-92.
21. Klion AD, Robyn J, Akin C, et al. Molecular remission and reversal of myelofibrosis in response to imatinib mesylate treatment in patients with the myeloproliferative variant of hypereosinophilic syndrome. *Blood* Jan 15;2004 103(2):473-478.
22. Pardanani A, Reeder T, Porrata LF, et al. Imatinib therapy for hypereosinophilic syndrome and other eosinophilic disorders. *Blood* May 1;2003 101(9):3391-3397.
23. Hunt SA, Abraham WT, Chin MH, et al. ACC/AHA 2005 Guideline Update for the Diagnosis and Management of Chronic Heart Failure in the Adult: a report of the American College of Cardiology/ American Heart Association Task Force on Practice Guidelines (Writing Committee to Update the 2001 Guidelines for the Evaluation and Management of Heart Failure): developed in collaboration with the American College of Chest Physicians and the International Society for Heart and Lung Transplantation: endorsed by the Heart Rhythm Society. *Circulation* Sep 20;2005 112(12):e154-235.
24. Wilson W, Taubert KA, Gewitz M. Quality of Care and Outcomes Research Interdisciplinary Working Group Prevention of Infective Endocarditis. Guidelines From the American Heart Association. A Guideline From the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Circulation 19 Apr 2007
25. Fuzellier JF, Chapoutot L, Torossian PF, et al. Mitral valve replacement in idiopathic eosinophilic endocarditis without peripheral eosinophilia. *J Card Surg* Sep-Oct;2005 20(5):472-474.
26. Watanabe K, Tournilhac O, Camilleri LF. Recurrent thrombosis of prosthetic mitral valve in idiopathic hypereosinophilic syndrome. *J Heart Valve Dis* May;2002 11(3):447-449.

27. Radford DJ, Garlick RB, Pohlner PG. Multiple valvar replacements for hypereosinophilic syndrome. *Cardiol Young* Jan;2002 12(1):67-70.
28. Korczyk D, Taylor G, McAlister H, et al. Heart transplantation in a patient with endomyocardial fibrosis due to hypereosinophilic syndrome. *Transplantation* Feb 27;2007 83(4):514-516.