Bölüm
39

RESTRİKTİF KARDİYOMİYOPATİ

Uğur TAŞKIN[1]

GİRİŞ

Restriktif kardiyomiyopatiler (RKM) gelişmiş dünyadaki erişkinlerde en az rastlanan primer kalp kası hastalığıdır(3,4). RKM'ler etiyolojilerine göre birincil veya ikincil olarak sınıflandırılır. Birincil RKM, idiyopatik RKM ve endomiyokardiyal fibrozu içerir. Yaygın sekonder formlar arasında infiltratif kardiyomiyopatiler, özellikle amiloidoz ve sarkoidoz; hemokromatozis primer ve sekonder formları; Fabry hastalığı gibi depo bozuklukları ve metastatik kanser ve radyasyona bağlı hastalık vardır.

RKM' lerin belirgin morfolojik ve hemodinamik özellikleri vardır. Kalbin duvar kalınlıkları genellikle normaldir, ancak infiltratif süreçlerle bu artabilir. Ejeksiyon fraksiyonu hastalığın ileri evrelerine kadar normaldir. Miyokard sertliği artmıştır ve buna bağlı olarak, şiddetli diyastolik fonksiyon bozukluğu, yüksek doldurma basınçları ile restriktif patern, normal sol ventrikül (LV) kavite boyutu ve dilate atriyuma neden olur. Nonkompliyan sol ventrikül, hacimdeki küçük artışlara rağmen dolma basınçlarında hızlı yükselme gösterir. Çoğu durumda hem sağ hem de sol ventriküller etkilenir buna bağlı olarak sağ, sol veya biventriküler yetmezliğin belirti ve semptomlarına neden olabilir. Ventriküler dolum basınçları istirahatte yüksektir ve egzersiz sırasında ventriküler nonkompliyans nedeniyle hızla daha da yükselir. Nonkompliyan sol ventrikül hızlı venöz dönüşü inhibe eder ve stroke volümde sınırlı artışlara neden olur. Aşamalı atriyal genişleme, atriyal aritmilere ve ikincil atriyoventriküler yetmezliğin gelişmesine neden olabilir. Eşlik eden atriyal fibrilasyon ile tromboembolik komplikasyonlar belirgin biatriyal genişleme ve zayıf atriyal kasılma nedeniyle görülebilir.

KLİNİK ÖZELLİKLER VE TANISAL DEĞERLENDİRME

Kalp yetersizliği en sık başvuru şeklidir, efor dispnesi tipik bir özelliktir. Egzersiz intoleransı, ventrikülün daha yüksek kalp hızlarında yeterince dolamamasından dolayı sıklıkla mevcuttur. Yorgunluk ve alt ekstremite ödemi de öne çıkan özelliklerdir.

Hepatomegali, assit ve belirgin ayak ödemi, hastalık ilerledikçe ortaya çıkabilir. Mitral ve triküspit yetersizliği sıklıkla mevcuttur. Telekardiyogram genellikle dilate atrium ve değişen derecelerde pulmoner konjesyona sahip normal boyutta bir ventriküler siluet gösterir. Elektrokardiyogramda (EKG) spesifik olmayan repolarizasyon anormallikleri eşliğinde biatrial dilatasyonun göstergesi olan büyük P dalgaları ile sinüs ritmi görülür. Atriyal fibrilasyon nadir değildir.

Ekokardiyografi RKM'de normal sağ ve sol ventrikül ejeksiyon fraksiyonunu ve boşluk çaplarını, biatriyal dilatasyonu ve restriktif diyastolik dolum parametrelerini gösterir. Anormal diyastolik kompliyans, doppler ekokardiyografik bulgularla değerlendirilir. Bu bulgular yükselmiş

[1] Uzman Doktor, Salihli Devlet Hastanesi, ugurtaskins@gmail.com

guların yarısına yakınında geç bir komplikasyon olarak görülür. Kardiyak lezyonların gelişimi, dolaşımdaki serotonin seviyeleri ve bunun başlıca metaboliti olan 5 hidroksiindoleasetik asit ile koreledir. Dilate kardiyomiyopatiye neden olmasının yanı sıra, antrasiklinler endomiyokardiyal fibrozise de neden olabilir. Radyoterapi öyküsü de varsa restriktif kalp yetmezliği riski büyük ölçüde artmaktadır(53).

KAYNAKÇA

1. Pereira NL, Grogan M, Dec GW, Spectrum of Restrictive and Infiltrative Cardiomyopathies Part 1 of a 2-Part Series. J Am Coll Cardiol 2018;71:1130–48
2. Pereira NL, Grogan M, Dec GW, Spectrum of Restrictive and Infiltrative Cardiomyopathies Part 2 of a 2-Part Series. J Am Coll Cardiol 2018;71:1149–66
3. Elliott P, Andersson B, Arbustini E, et al. Classification of the cardiomyopathies: a position statement from the European Society Of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J 2008;29:270–6.
4. Kushwaha SS, Fallon JT, Fuster V. Restrictive cardiomyopathy. N Engl J Med 1997;336:267–76.
5. Vaitkus PT, Kussmaul WG. Constrictive pericarditis versus restrictive cardiomyopathy: a reappraisal and update of diagnostic criteria. Am Heart J 1991;122:1431–41.
6. Garcia MJ. Constrictive pericarditis versus restrictive cardiomyopathy? J Am Coll Cardiol 2016;67:2061–76.
7. Gupta A, Singh Gulati G, Seth S, et al. Cardiac MRI in restrictive cardiomyopathy. Clin Radiol 2012;67:95–105.
8. Iles L, Pfluger H, Phrommintikul A, et al. Evaluation of diffuse myocardial fibrosis in heart failure with cardiac magnetic resonance contrastenhanced T1 mapping. J Am Coll Cardiol 2008; 52:1574–80.
9. Boynton SJ, Geske JB, Dispenzieri A, et al. LGE provides incremental prognostic information over serum biomarkers in AL cardiac amyloidosis. J Am Coll Cardiol Img 2016;9:680–6.
10. Geske JB, Anavekar NS, Nishimura RA, et al. Differentiation of constriction and restriction: complex cardiovascular hemodynamics. J Am Coll Cardiol 2016;68:2329–47.
11. Cooper LT, Baughman KL, Feldman AM, et al. The role of endomyocardial biopsy in the management of cardiovascular disease: a scientific statement from the American Heart Association, the American College of Cardiology, and the European Society of Cardiology. Circulation 2007; 116:2216–33.
12. Manual of Cardiovascular Medicine- Eric J.Topol, Lippincott Williams & Wilkins, 2004
13. Braunwald's Heart Disease - A Textbook of Cardiovascular Medicine, Eugene Braunwald, W.B. Saunders Company; 6th Edition.
14. Rajagopalan N, Garcia MJ, Rodriguez L, et al. Comparison of new Doppler echocardiographic methods to differentiate constrictive pericardial heart disease and restrictive cardiomyopathy. Am J Cardiol 2001;87:86–94.
15. Ling LH, Oh JK, Tei C, et al. Pericardial thickness measured with transesophageal echocardiography: feasibility and potential clinical usefulness. J Am Coll Cardiol 1997;29:1317–23.
16. Hurrell DG, Nishimura RA, Higano ST, et al. Value of dynamic respiratory changes in left and right ventricular pressures for the diagnosis of constrictive pericarditis. Circulation 1996;93: 2007–13.
17. Hatle LK, Appleton CP, Popp RL. Differentiation of constrictive pericarditis and restrictive cardiomyopathy by Doppler echocardiography. Circulation 1989;79:357–70.
18. Welch TD, Ling LH, Espinosa RE, et al. Echocardiographic diagnosis of constrictive pericarditis: Mayo Clinic criteria. Circ Cardiovasc Imaging 2014;7:526–34.
19. Ha JW, Oh JK, Ling LH, et al. Annulus paradoxus: transmitral flow velocity to mitral annular velocity ratio is inversely proportional to pulmonary capillary wedge pressure in patients with constrictive pericarditis. Circulation 2001;104:976–8.
20. Masui T, Finck S, Higgins CB. Constrictive pericarditis and restrictive cardiomyopathy: evaluation with MR imaging. Radiology 1992;182: 369–73.
21. Kojima S, Yamada N, Goto Y. Diagnosis of constrictive pericarditis by tagged cine magnetic resonance imaging. N Engl J Med 1999;341:373–4.
22. Bogabathina H, Biederman RW. Lack of slippage by cardiovascular magnetic resonance imaging is sine qua non for constrictive pericarditis. Circulation 2011;123:e418–9.
23. Longhi S, Quarta CC, Milandri A. Atrial fibrillation in amyloidotic cardiomyopathy: prevalence, incidence, risk factors and prognostic role. *Amyloid*. 2015;22:147–155. doi: 10.3109/13506129.2015.1028616.
24. Quarta CC, Solomon SD, Uraizee I, et al. Left ventricular structure and function in transthyretin-related versus light-chain cardiacamyloidosis. *Circulation*.2014;129:1840–1849. doi: 10.1161/CIRCULATIONAHA.113.006242.
25. Muchtar E, Derudas D, Mauermann M, et al. Systemic immunoglobulin light chain amyloidosis-associated myopathy: presentation, diagnostic pitfalls, and outcome. *Mayo Clin Proc*. 2016;91:1354–1361. doi: 10.1016/j.mayocp.2016.06.027.
26. Muchtar E, Dean DS, Dispenzieri A, et al. Prevalence and predictors of thyroid functional abnormalities in newly diagnosed al amyloidosis. *J Intern Med*. 2017;281:611–619. doi: 10.1111/joim.12617.
27. Dubrey SW, Bilazarian S, LaValley M, et al. Signal-averaged electrocardiography in patients with AL (primary) amyloidosis. *Am Heart J*. 1997;134:994–1001.
28. Lin G, Dispenzieri A, Kyle R, et al. Implantable cardioverter defibrillators in patients with cardiac amyloidosis. *J Cardiovasc Electrophysiol*. 2013;24:793–798. doi: 10.1111/jce.12123.
29. Baughman RP, Lower EE, du Bois RM. Sarcoidosis. *Lancet*. 2003;361:1111–1118. doi: 10.1016/S0140-6736(03)12888-7.
30. Martusewicz-Boros MM, Boros PW, Wiatr E, et al. Cardiac sarcoidosis: is it more common in men? *Lung*. 2016;194:61–66. doi: 10.1007/s00408-015-9805-8.

31. Diagnostic standard and guidelines for sarcoidosis. *Jpn J Sarcoidosis Granulomatous Disord*. 2007;27:89–102.
32. Birnie DH, Sauer WH, Bogun F, et al. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis.
33. Kandolin R, Lehtonen J, Kupari M. Cardiac sarcoidosis and giant cell myocarditis as causes of atrioventricular block in young and middle-aged adults. *Circ Arrhythm Electrophysiol*. 2011;4:303–309. doi: 10.1161/ CIRCEP.110.959254.
34. Suzuki T, Kanda T, Kubota S, et al. Holter monitoring as a noninvasive indicator of cardiac involvement in sarcoidosis. *Chest*. 1994;106:1021–1024.
35. Mehta D, Lubitz SA, Frankel Z, et al. Cardiac involvement in patients with sarcoidosis: diagnostic and prognostic value of outpatient testing. *Chest*. 2008;133:1426–1435. doi: 10.1378/chest.07-2784.
36. Yazaki Y, Isobe M, Hiroe M, et al. Prognostic determinants of long-term survival in Japanese patients with cardiac sarcoidosis treated with prednisone. *Am J Cardiol*. 2001;88:1006–1010.
37. Murphy CJ, Oudit GY. Iron-overload cardiomyopathy: pathophysiology, diagnosis, and treatment. J Card Fail 2010;16:888–900.
38. Gujja P, Rosing DR, Tripodi DJ, et al. Iron overload cardiomyopathy: beter understanding of an increasing disorder. J Am Coll Cardiol 2010;56:1001–12.
39. Kremastinos DT, Farmakis D. Iron overload cardiomyopathy in clinical practice. Circulation 2011;124:2253–63.
40. Spirito P, Lupi G, Melevendi C, et al. Restrictive diastolic abnormalities identified by Doppler echocardiography in patients with thalassemia major. Circulation 1990;82:88–94.
41. Lubitz SA, Goldbarg SH, Mehta D. Sudden cardiac death in infiltrative cardiomyopathies: sarcoidosis, scleroderma, amyloidosis, hemachromatosis. Prog Cardiovasc Dis 2008;51:58–73.
42. Kremastinos DT, Tsiapras DP, Tsetsos GA, et al. Left ventricular diastolic Doppler characteristics in beta-thalassemia major. Circulation 1993;88: 1127–35. Pereira et al. JACC VOL. 71, NO. 10, 2018 Restrictive Cardiomyopathies: Part 2 MARCH 13, 2018:1149–66
43. Palka P, Macdonald G, Lange A, et al. The role of Doppler left ventricular filling indexes and Doppler tissue echocardiography in the assessment of cardiac involvement in hereditary hemochromatosis. J Am Soc Echocardiogr 2002; 15:884–90.
44. Kremastinos DT. Heart failure in betathalassemia. Congest Heart Fail 2001;7:312–4.
45. Cheng H, Zhao S, Jiang S, et al. The relative atrial volume ratio and late gadolinium enhancement provide additive information to differentiate constrictive pericarditis from restrictive cardiomyopathy. J Cardiovasc Magn Reson 2011;13:15.
46. Wu JC, Ho CY, Skali H, et al. Cardiovascular manifestations of Fabry disease: relationships between left ventricular hypertrophy, disease severity, and alpha-galactosidase A activity. Eur Heart J 2010;31:1088–97.
47. Weidemann F, Breunig F, Beer M, et al. Improvement of cardiac function during enzyme replacement therapy in patients with Fabry disease: a prospective strain rate imaging study. Circulation 2003;108:1299–301.
48. Mehta A, Beck M, Elliott P, et al. Enzyme replacement therapy with agalsidase alfa in patients with Fabry's disease: an analysis of registry data. Lancet 2009;374:1986–96.
49. Ortiz A, Abiose A, Bichet DG, et al. Time to treatment benefit for adult patients with Fabry disease receiving agalsidase beta: data from the Fabry Registry. J Med Genet 2016;53:495–502.
50. Tharakan J, Bohora S. Current perspective on endomyocardial fibrosis. Curr Sci 2009;97: 405–10.
51. Topilsky Y, Pereira NL, Shah DK, et al. Left ventricular assist device therapy in patients with restrictive and hypertrophic cardiomyopathy. Circ Heart Fail 2011;4:266–75.
52. Fathi AT, Dec GW Jr., Richter JM, et al. Case records of the Massachusetts General Hospital. Case 7-2014. A 27-year-old man with diarrhea, fatigue, and eosinophilia. N Engl J Med 2014;370: 861–72.
53. Bhattacharyya S, Davar J, Dreyfus G, et al. Carcinoid heart disease. Circulation 2007;116: 2860–5.