Bölüm
14

KARDİYOMİYOPATİLER

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GİRİŞ

Kardiyomiyopati; koroner arter hastalığı, kalp kapak hastalığı, hipertansiyon ve konjenital kalp hastalığı yokluğunda, yapısal ve fonksiyonel miyokardiyal işlev bozukluğu ile karakterize kalp kası hastalığıdır (1).

Kardiyomiyopati adlandırması ilk olarak 1957 yılında Brigden tarafından idiyopatik miyokardiyal hastalıkları tanımlamak amacıyla ‘’koroner dışı kardiyomiyopatiler” olarak kullanılmıştır (2). Bu adlandırmadan 23 yıl sonra, 1980 yılında, John F. Goodwin başkanlığında toplanan Dünya Sağlık Örgütü (DSÖ) ilk kardiyomiyopati sınıflamasını yapmıştır. Bu sınıflamada, kardiyomiyopatiler yapısal ve hemodinamik fenotiplerine göre dilate kardiyomiyopati (DKM), hipertrofik kardiyomiyopati (HKM) ve restriktif kardiyomiyopati (RKM) olmak üzere 3 ana sınıfa ayrılmış ve tüm kardiyomiyopatiler ‘’nedeni bilinmeyen kalp kası hastalıkları” olarak tanımlanmıştır (3). DSÖ 1996 yılında yeniden yaptığı yaptığı kardiyomiyopati tanımlamasında, ‘’nedeni bilinmeyen” ibaresini kaldırarak ‘’kardiyak disfonksiyon ile ilişkili miyokard hastalıkları” tanımını kullanmıştır ve ilk kez aritmojenik sağ ventrikül kardiyomiyopatisi sınıflamaya dahil edilmiştir (4,5). Kardiyomiyopatiler, Amerikan Kalp Birliği’nin 2006 yılında yayınladığı uzlaşı raporunda ‘’ventriküler dilatasyon ya da hipertrofi ile karakterize, mekanik (sistolik ya da diyastolik) ve/veya elektriksel işlev bozukluğu ile ilişkili ve çoğunlukla genetik nedenlere bağlı gelişen miyokardiyal hastalık” olarak tanımlanmıştır (6). Bu raporda, kardiyomiyopatiler yalnızca kalp kası tutulumu ile seyreden primer kardiyomiyopatiler ve miyokardiyal tutuluma ek olarak multisistemik tutulum gösteren sekonder kardiyomiyopatiler (Anderson-Fabry hastalığı, amiloidoz, sarko-

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kıntılar ve intertrabeküler derin girintiler lokalizasyon olarak sol ventrikül apeks bölgesi ya da inferolateral duvarın midventriküler segmentlerinde belirgin izlenmelidir (135).

Chin Kriterleri: Diyastol sonu evrede apikal uzun eksen ekokardiyografik görüntülerde sol ventrikül apek ve/veya sol ventrikül serbest duvarında izlenen trabekülasyonlarda tepe-vadi oranını ≤0.5 olması NCKM açısından tanısal kabul edilmektedir (136).

Stöllberger Kriteri: Sol ventrikül kavitesinde en az 4 ve daha fazla sayıda trabekül olmalı ve sol ventrikül apeks bölgesinde en az bir trabekülasyon izlenmelidir. İntertrabeküler boşluklar sol ventrikül kavitesi içinden kanlanmalıdır (137,138).

NCKM tanısında yüksek çözünürlük sağlaması ve kompakte / nonkompakte miyokard katmanlarının daha net saptanabilmesi nedeniyle kardiyak MRI'ın özgüllüğü ve duyarlılığı ekokardiyografiye kıyasla daha yüksektir. Tanısal açıdan kardiyak MRG için farklı tanı kriterleri tanımlanmıştır. Diyastol sonunda elde edilen ölçümlerde nonkompakte miyokard alanı kalınlığının kompakte miyokard alanı kalınlığına oranı 2.3 veya daha fazla olması ya da trabeküle sol ventrikül kitlesinin tüm sol ventrikül kitlesinin >%20'sini oluşturması tanısal kabul edilmektedir (139,140).

Tedavi: NCKM'nin spesifik bir tedavisi yoktur. Hastalığın seyrinde gelişen kalp yetersizliği, tromboembolik komplikasyonlar ve aritmilerin tedavisi NCKM'nin tedavisini oluşturmaktadır. Hipertrofik kardiyomiyopatide olduğu gibi atriyal fibrilasyon atağı olan olguların CHADS2-VASC skorundan bağımsız olarak antikoagüle edilmesi önerilmektedir (119).

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