

Bölüm 41

DİĞER KARDİYAK İNFİLRATİF VE SPESİFİK KARDİYOMİYOPATİLER

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GİRİŞ

Kardiyomiyopati 1980 yılında WHO (World Health Organization) tarafından nedeni bilinen spesifik kalp hastalıklarından ayırmak amacıyla, nedeni bilinmeyen kalp kası hastalıkları olarak tanımlamıştır (1). 1995 yılında ise bu tanım kardiyak disfonksiyonla ilişkili miyokard hastalıkları olarak düzenlenmiştir. Bu raporda WHO/ISFC (International society and federation of cardiology) etyoloji ve dominant patofizyolojiye göre kardiyomiyopatileri beş gruba sınıflamıştır (2):

- 1) Dilate kardiyomiyopati (DKM)
- 2) Hipertrofik kardiyomiyopati (HKM)
- 3) Restriktif kardiyomiyopati (RKM)
- 4) Aritmojenik sağ ventrikül kardiyomiyopati/displazisi (ARVK/D)
- 5) Sınıflandırılmayan kardiyomiyopatiler

Kardiyomiyopatiler primer yani kalbe sınırlı olabilecekleri gibi, sekonder yani kalp tutulumu olan sistemik bir hastalığın parçası da olabilirler (3).

Dilate, hipertrofik ve restriktif kardiyomiyopatiler önceki bölümlerde anlatılmış olup bu konu başlığı içerisinde öncelikle aritmojenik kardiyomiyopati ve sonrasında sınıflandırılmayan, diğer spesifik ve infiltratif kardiyomiyopatilerden bahsedilecektir.

ARİTMOJENİK SAĞ VENTRİKÜLER KARDİYOMİYOPATI/ DİSPLAZİSİ (ARVK/D)

Sağ ventrikül miyokardının progresif olarak fibröz ve yağ doku değişimi ile ilerleyen, öncelikle tipik olarak bölgesel başlayıp sonrasında yaygın sağ ve bazen sol ventrikül tutulumunun izlenebildiği bir kardiyomiyopatidir (4). Biventriküler tutulum da gözlenebilir. Bu nedenle aritmojenik kardiyomiyopati ifadesinin daha uygun olduğu düşünülmüştür (5). İlk kez 1736 yılında tanımlanmış olup prevalansı 1/2500 - 1/5000 aralığındadır. Otozomal dominant geçişin izlendiği ailesel form yaygın olsa da resesif formları (Naxos hastalığı ve carvajal sendromu gibi) da tanımlanmıştır. Özellikle genç hastalarda aritmi ve ani kardiyak ölüm sık olarak görülmektedir (6).

Patolojik olarak miyokardiyal atrofi, fibroadipoz doku değişimi, fibrozis ve etkilenen duvarın incilmesi, kavitenin dilatasyonu ve anevrizma oluşumu ile karakterizedir. Yağ infiltrasyonu ARVK/D için patognomonik olmayıp histolojik bulgular erken evrede bulunmayabilir. Tutulumun en belirgin olduğu yer 'triangle of displasia' olarak bilinen SaV(Sağ ventrikül) çıkış yolu, triküspit kapağın altındaki alan ve SaV apeksini kapsayan alandır. Bu alandaki anormallikler sinüs ritmi sırasındaki tipik EKG görünümü ve LBBB ve superior aks ile karakterize tipik monomorfik VT'den

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Kardiyomiyopatilerin tanımlanması ve özelliklerinin belirlenmesi konusunda yıllar içinde önemli gelişmeler kaydedilmiştir. Fakat kardiyomiyopatilerin genetik temelleri henüz tam olarak bilinmemektedir. Konu ile ilgili gelecekteki çalışmalarla kardiyomiyopatiler ile ilgili bilinmeyenler aydınlatılabilecektir. Mevcut durumda bir kardiyoloğun görevi mevcut veriler ve bilgiler eşliğinde kardiyomiyopatiyi tanımlayabilmek ve sonrasında eşlik edebilecek durumları araştırmaktır.

Anahtar Kelimeler: Kardiyomiyopati, kardiyak infiltrasyon, kalp yetmezliği

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