

BÖLÜM 10

HİPERTROFİK KARDİYOMYOPATİ (HKMP)

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GİRİŞ

Kardiyomyopatiler kalp kasının kanı pompalama yeteneğinin kaybına yol açan hastalıklarıdır. Kardiyomyopatiler, koroner arter akım kısıtlılığı veya kalbin aşırı yüklenmesi ile açıklanamayan ventriküler miyokardın yapısal ve fonksiyonel anormalileri olarak tanımlanır. Tarihsel olarak bu hastalık, kalbin tek tutulan organ olduğu primer ve bir sistemik hastalığın sonucunda olduğu sekonder formları olmak üzere iki alt gruba ayrılmıştır. Hipertrofik kardiyomyopatide diğer kardiyomyopatilerin aksine sol ventrikül (SIV) duvar kalınlığı artmıştır. Daha ayrıntılı bir tanımlama yaparsak, *primer HKM, kalp kasının, kardiyak proteinleri kodlayan genlerin mutasyonu ile ilgili çoğu zaman otozomal dominant geçen, küçük sol ventrikül kavitesi, artmış sistolik ve bozulmuş diyastolik fonksiyonla karakterize primer hipertrofidir*¹. Histopatolojik olarak miyokardiyal hipertrofi miyokardiyal liflerin düzensizliği, artmış bağ dokusu kaybı ve fibrozis ile birliktedir, bu da kalp kasının güç üretimini etkiler. Aslında geleneksel olarak, sol ventrikül çıkım yolunun ostrüksiyona neden olan hiperdinamik sistolik bir bozukluğu olarak bilinse de, HKM’li hastaların büyük bir kısmı normal sistolik fonksiyon gösteren diyastolik kalp yetmezliği semptomları bulunan nonobstrüktif olgulardır. *Bu olgularda myokard adelesinin gevşeme kusurları ve diyastolde sol ventrikülün dolamaması temel problemdir*. Bununla beraber, daha az görülen dinamik tip SIV çıkım yolu darlığına yol açan obstrüktif form, özellikle cerrahi tedavisiyle önem arzeder. Bu hastalık grubuna literatürde, asimetrik septal yapıyı, dinamik darlığı, artmış sistolik fonksiyonları ve hiperkinetik ventrikülleri, genetik ilişkilerini gösterir bir çok isim verilmiştir. Hipertrofik obstrüktif kardiyomyopati (HOKM), idiyopatik hipertrofik subaortik stenoz (İHSS), musküler

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