Bölüm
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HİPERTROFİK KARDİYOMİYOPATİ

Gürkan İMRE[1]

GİRİŞ:TANIM VE GÖRÜLME SIKLIĞI

Hipertrofik kardiyomiyopati (HKMP), hipertrofiye sebep olabilecek kardiyak ya da sistemik bir hastalık olmadan daha çok interventriküler septumu tutan ve dilate olmamış sol ventrikül kavitesi ile karşımıza çıkan primer miyokard hastalığıdır. Başlıca etkilenen sol ventrikül kısmı septum olup, serbest duvar daha az oranda etkilenir. HKMP sol ventrikülün asimetrik hipertrofisi, diffüz veya segmenter sol ventrikül duvar kalınlaşması ile dilate olmamış sol ventrikül kavitesi ile karakterizedir. Hastaların 2/3'ünde septal hipertrofi ve mitral kapağın sistolik öne hareketi belirgin olup sol ventrikül çıkış yolunda değişik derecelerde obstrüksiyon meydana getirerek gradiyent oluşturur. Bunun yanında hastaların 1/3'ünde istirahatte veya eforla sol ventrikül çıkış yolunda obstrüksiyon ve gradiyent artışı saptanmamaktadır(1). Bu hastalık ilk kez 1958 yılında Brock ve Teare tarafından ''Asimetrik hipertrofik kardiyomiyopati" adıyla tanımlanmıştır. Günümüzde sol ventrikül çıkış yolundaki gradiyent varlığına göre obstrüktif ve non-obstrüktif hipertrofikkardiyomiyopati olarak isimlendirilmektedir(2,3,7).

HKMP'li hastaların %40-60' ında etiolojide otozomal dominant geçişli sarkomerik protein gen mutasyonu mevcuttur. Hastaların %25-30'unda herhangi bir neden saptanamazken, %10 hastada ise amiloidozis gibi infiltratif hastalıklar, ilaçlar (özellikle anabolizan steroidler), feokromasitoma ve akromegali gibi endokrin hastalıklar, metabolik hastalıklar, bazı sendromik hastalıklar ile bazı mitokondrial hastalıklar sebep olmaktadır (Tablo-1). En sık görülen genetik geçişli kalp damar hastalığı olarak kabul edilen HKMP'nin prevalansı %0.02- 0.023 arasında olduğu bildirilmektedir. Yani yaklaşık olarak 500 erişkinde bir görülmektedir. Erkeklerde 2 kat daha sık görülmekle beraber kadınlarda genç yaşlarda daha sık görülür ve daha ağır bir seyir göstermektedir.

Belirgin hipertrofinin gelişmesi için belirli bir latent dönem gerektiğinden dolayı ergenlik döneminin sonrasına kadar bu değişim devam etmektedir. Gençlerde genellikle asemptomatik seyretmesi nedeniyle ilk tanı sıklıkla başka sebeplerle yapılan sağlık taramaları veya aile taramaları sırasında konulmaktadır. Daha ileri yaşlarda ise genellikle kardiyovasküler hastalıkların araştırılması sırasında tesadüfen tespit edilmektedir. Genç atletlerde hipertrofik kardiyomiyopati çok nadirdir, buna karşın ani kalp ölümlerinin görüldüğü bu popülasyonda ani ölümlerin 1/3'ünün sebebi hipertrofik kardiyomiyopatidir. Sporcularda daha sıklıkla sol ventrikül kavitesinin de genişlediği sol ventrikül hipertrofisi gelişir(4-8).

Patofizyoloji

Hipertrofik kardiyomiyopatideki temel patofizyolojik değişim fibrozis ve miyozitlerdeki organizasyon kusuru ile karakterizedir. Miyokard dokusunda farklı derecelerde fibroz doku artışı ve skar oluşumu vardır. Bu defektler miyozitlerin elektriksel yapısını bozarak aritmojenik ortam

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Ekokardiyografi, Pacemaker, ICD, Perkütan Septal Ablasyon, Miyektomi

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