BÖLÜM 17

Hipertrofik Kardiyomyopati ve Atriyal Fibrilasyon

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GİRİŞ

Hipertrofik kardiyomyopati (HKMP), genel popülasyonda yaklaşık 400 kişiden 1'ini etkileyen, en yaygın kalıtsal kalp hastalığıdır (1). Tanım olarak bakıldığında HKMP; aort darlığı, sistemik hipertansiyon gibi herhangi bir yüklenme durumu yokluğunda ventrikül duvar kalınlığında artış (>15 mm veya pozitif genetik test veya HKMP tanılı aile üyesi varlığında >13 mm) ile karakterizedir (2,3). Atriyal fibrilasyon (AF) ise HKMP hastalarında en sık görülen devamlı aritmidir (4,5) Genel popülasyon ile karşılaştırıldığında HKMP hastaları yaşamları boyunca 4 ila 6 kat fazla AF gelişme riskine sahiptir (6,7).

AF'nin bu hasta grubunda daha sık görülmesine ilave olarak, daha erken yaşlarda ortaya çıkma eğiliminde olduğu gösterilmiştir (8). Sinüs ritmindeki ve AF olan HKMP hastaları karşılaştırıldığında, AF olanlarda 4 kat artmış ölüm riski bulunmaktadır (9). İnme ve tromboembolik olaylar en yaygın komlikasyonlardır ve iskemik strok 8 kat fazla izlenmektedir (8).

PATOFİZYOLOJİ

HKMP'nin patogenezi, sarkomer proteinlerinde yapısal değişikliklere neden olan ve kalbin kas hücresi boyutunun artmasına neden olan mutasyonlar ile açıklanmaktadır (10). Mutasyonlar, kalp kası hücrelerinin yapısal bileşenlerinde fibrozu hızlandıran ve interventriküler septumun kalınlığını artıran hücresel bir düzensizliğe neden olur (11). Mutasyonların neden olduğu bu değişiklikler LVOT obstrüksiyonuna (LVOTO), sistolde LV end sistolik volüm artışına neden olur. Ayrıca, HKMP hastalarında sıklıkla atriyal iskemi ve mikrovasküler disfonksiyona bağlı artan miktarda atriyal fibrozis ile karşılaşılır, bu da atriyal genişlemeye ve fonksiyon bozukluğuna katkıda bulunur (12) Son olarak, genetik faktörler de LA remodelingini modüle ederek AF insidansında rol oynayabilir (13).

Son zamanlarda spesifik sarkomerik gen mutasyonları, HKMP popülasyonunda AF'nin daha erken başlamasıyla ilişkilendirilmiştir (14). Ayrıca, çoğunlukla renin anjiyotensin-al-

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atriyal boyut ve AF tipi, AF nüksünün güçlü öngörücülerinden biridir. Yapılan çalışmalarda uzun dönem başarı oranı %76.7 bulunmuştur. Antiaritmik ilaç tedavisi almadan 1.yılda ve en son takipte başarı oranı %69.45 ve %50.4'tür (38, 40-42).

Eşzamanlı septal miyektomi sırasında cerrahi ablasyon yapılan birçalışmada ise 1 yıl ve 6 yıllık takipte; sırasıyla %96 ± 3.5 ve %80 ± 8.1 oranında aritmi kontrolü sağlanmıştır (43).

Cox-maze-III, Cox-maze IV veya PVI prosedürleri ile kombine edilen septal miyektomi sonuçlarının incelendiği başka bir çalışmada 5 yıllık AF nüksü %48 olarak tespit edilmiştir (44).

SONUÇ

AF, HKMP hastalarında sık görülen bir olaydır ve aynı zamanda olumsuz bir prognoz ile ilişkilidir. Gelişiminin nedenleri, HKMP ve genetik faktörlerle ilişkili tipik anatomik ve hemodinamik değişiklikler de dahil olmak üzere çok faktörlüdür. Klinisyenler, hastalarla yapılan düzenli takipler sırasında bu aritmiye karşı yüksek şüphe duymalı ve dikkatli olmalıdır.

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