23. BÖLÜM

REKÜRREN VE METASTATİK MALİGN FEOKROMASİTOMA YÖNETİMİ

Perihan PERKİN[1]

GİRİŞ

Feokromasitomalar adrenal medulladaki kromaffin hücrelerden, paraganglioma olarak adlandırılan ekstraadrenal feokromasitomalar ise paraaortik sempatik ganglionlardan köken alan neoplazilerdir. Nadir görülen feokromasitoma hipertansif hastaların yüzde 0.2'sinden azında saptanmaktadır (1). Her yaşta görülebilmekle birlikte en sık dördüncü ve beşinci dekatta ortaya çıkar, kadın ve erkeklerde eşit oranda görülür (2). Feokromasitoma ve paragangliomalar histopatoloji, epidemiyoloji, moleküler patobiyoloji açısından benzer karakteristik özellikler gösterse de klinik seyir, agresif davranış, metastaz potansiyeli, biyokimyasal bulgular ve kalıtsal genetik sendromlarla ilişkiler açısından farklılıkları mevcuttur. Feokromasitoma çoğunlukla sporadik olsa da vakaların yaklaşık %40'ında von Hippel-Lindau (VHL) sendromu, multiple endokrin neoplazi tip 2 (MEN2), nörofibromatozis tip 1 (NF1) gibi ailesel hastalıklarla birlikte görülebilmektedir.

Feokromasitomalar katekolamin sekrete eden tümörlerdir ve yaklaşık vakaların yarısında katekolamin sekresyonuna bağlı olarak hipertansiyon, epizodik baş ağrısı, terleme, titreme, çarpıntı gibi semptomlar ortaya çıkmaktadır. Hipertansiyon en sık bulgu olsa da %5-15 hastada kan basıncı normal saptanabilir, baş ağrısı semptomatik vakalarda %90 oranında görülmektedir (3). Daha nadir görülen belirtiler arasında ortostatik hipotansiyon, poliüri, polidipsi, kabızlık, insülin direnci, hiperglisemi yer almaktadır (4).

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varlığında EBRT kullanılabilir. Radyofrekans ablasyon, kriyoablasyon, etanol enjeksiyonu gibi lokal ablatif yöntemler kemik, yumuşak doku, karaciğer metastazlarında uygulanabilir. Yaygın karaciğer metastazı varlığında rezeksiyon veya diğer lokal ablatif yöntemlere uygun olmayan hastalarda transarteriyel kemoembolizasyon uygulanabilir. Lokal ablatif tedavilerde de işlem esnasında aşırı katekolamin sekresyonu ve hipertansif kriz olabileceğinden işlem öncesi medikal hazırlık gereklidir.

Iobenguane I-123 tanısal sintigrafisinde feokromasitoma/paragangliomalı hastaların yaklaşık %60'ında tutulum saptanır. Unrezektable, semptomatik, progresif MIBG-pozitif tümörü olan hastalarda lokorejyonel yöntemler uygun değilse, tümör yükü çok fazlaysa, kemik metastazı ön planda değilse sistemik kemoterapiden önce Iobenguane I-131 tedavisi tercih edilmelidir. Ancak en uygun doz ile ilgili ortak bir görüş yoktur.

Hızlı progrese olan veya kemik metastazlarının ön planda olduğu hastalarda MIBG-pozitif olsa bile sistemik kemoterapi öncelikli tercih edilmelidir. Optimal rejim net olmasa da siklofosfamid, vinkristin, dakarbazin kombinasyonu önerilmektedir.

Yakın takip ve uygun antihipertansif tedavi ile sunitinib etkinliği gösterilmiş ve güvenle kullanılabilecek bir ajandır.

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