BÖLÜM 25

NADİR BÖBREK TÜMÖRLERİ

Tanju KAPAĞAN[1]

GİRİŞ

Böbrek kanserleri, tüm kanserlerin yaklaşık %2'sini oluşturmaktadır (1). Erişkinlerde altıncı ve yedinci dekatlarda ve erkeklerde kadınlara göre daha sık görülmektedir (2). Dünya Sağlık Örgütü-Uluslararası Kanser Araştırma Ajansı'na göre, 2025 yılında dünyada 487.500'den fazla yeni böbrek kanseri vakasının görüleceği tahmin edilmektedir (3). Böbrek hücreli kanser (BHK); malign böbrek tümörlerinin yaklaşık %90'ını oluşturur. BHK daha çok berrak hücreli karsinom ve papiller hücreli karsinom alt tiplerinden oluşmaktadır. BHK'lerin nadir görülen diğer alt tipleri ise kromofob hücreli karsinom, toplayıcı (bellini) duktus hücreli karsinom, multiloküler kistik hücreli karsinom, medüller hücreli karsinom, müsinöz tübüler ve iğsi hücreli karsinom, translokasyon ilişkili karsinom, nöroblastom ile ilişkili karsinom ve sınıflandırılmamış karsinomlardır (4, 5). BHK'ler dışında soliter fibröz tümör, epiteloid anjiomyolipom, leiomyosarkom, ewing sarkomu, anjiosarkom, rabdomyosarkom, nöroendokrin tümörler de böbreğin nadir görülen diğer tümörleri olarak bilinmektedir (6). Bu derleme ile nadir görülen bu tümörlerin güncel literatür bilgisi ışığında yeniden gözden geçirilmesi amaçlanmıştır.

Nadir Görülen Böbrek Hücreli Kanser (BHK) Alt Tipleri

I-Kromofob BHK

Kromofob böbrek hücreli karsinom (KBHK), böbrek korteks distal kıvrımlı tübülüs hücrelerinden köken alıp tüm BHK'ler içerisinde %5 oranında görülen histolojik alt tiptir (7-9). Erişkinlerde beşinci ve altıncı dekatta, kadınlarda erkeklere göre daha sık görülmektedir (10). Genel olarak hastalar kliniklere karın veya yan ağrısı, abdominal kitle, ateş, kaşeksi, yorgunluk ve kilo kaybı şikayetleri ile başvurmaktadırlar (11). Bu tümör tipi genellikle sporadik olup iyi bir prognoza sahiptir. Bu tümörün ayrıcı tanısına benign bir tümör olan onkositoma başta olmak üzere berrak, papiller, toplayıcı kanal ve düşük malignite potansiyelli mul-

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kontrast tutan, sık olmamakla birlikte kalsifikasyon veya kistik komponent içeren solid kitleler olarak görülmektedirler (119). Metastatik hastalarda evreleme amaçlı veya hastalık yayılımını değerlendirmek amaçlı gallium temelli PET/BT veya oktreotid sintigrafisi çekilebilir (120, 121). Hastalığın kesin tanısı, biyopsi ile elde edilen hematoksilen- eozin ile boyalı lamlardan İHK olarak sinaptofizin, kromogranin-A, CD56 gibi antikorların gösterilmesine dayanır (122). Temel tedavi yaklaşımı, rezektabl hastalarda kitlenin cerrahi rezeksiyon ile total olarak çıkartılmasıdır. Unrezektabl veya metastatik hastalarda ise genellikle iki veya daha fazla ilacın kombine olarak kullanıldığı karboplatin, sisplatin, doksorubisin, etoposid, ifosfamid, irinotekan, paklitaksel, topotekan gibi kemoterapi ajanları tercih edilmektedir (123).

SONUÇ

Sonuç olarak belirttiğimiz böbrek kaynaklı tümör alt tiplerinin sık görülmediği bilinmektedir. Literatürde bu tümörler ile ilgili daha çok olgu bildirimleri veya retrospektif olarak değerlendirilen küçük hasta serileri bulunmaktadır. Bu tümörler nadir görüldüklerinden ayırıcı tanıda hemen akla gelmeyip sıklıkla gözden kaçabilmektedirler. Bu derlemede nadir görülen bu tümörlere ait başlangıç semptomlar, radyolojik bulgular, histopatolojik-İHK özellikler ve tedavilerindeki güncel yaklaşım vurgulanmıştır. Bu tümörlerden bazıları düşük dereceli olup daha selim seyrederken bazıları ise daha yüksek dereceli olup daha agresif seyretmektedir. Bu nedenle bu tümörlerin klinik özelliklerinin daha iyi anlaşılıp hastalığın erken teşhis edilmesi hastaların tedavi ve yaşam kalitesi açısından büyük önem taşımaktadır.

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