



BÖLÜM 5

BÖBREĞİN MEZENŞİM KÖKENLİ TÜMÖRLERİ

5.1. YETİŞKİN BÖBREĞİNDE MEZENŞİM KÖKENLİ TÜMÖRLER

5.1.1. KLASİK ANJİOMYOLİPOMA/ PECOMA

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5.1.1.1. Tanım

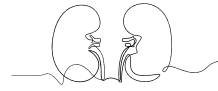
- ▶ Çeşitli oranlarda adipöz doku, içsi ve epiteloïd düz kas hücreleri ve kalın duvarlı kan damarlarından oluşan myomelanositik farklılaşma gösteren benign mezenkimal tümördür (1).

5.1.1.2. Köken/Patogenez

- ▶ 1991 yılında, perivasküler proliferasyon gösteren epiteloïd hücrelerle karakterli neoplazilerde, HMB45 immünreaktivitesi gösterilmiştir. Anjiomyolipom (AML), lenfanji-oleiomyomatozis, akciğerin ve pankreasın berrak hücreli tümöründen oluşan bu tümörlere perivasküler epiteloïd hücreli tümör (PEComa) ismi verilmiştir (2-4).
- ▶ Uzun bir süre hamartoma olarak tanımlanmıştır. 1990'lı yıllarda X kromozomunda rastgele olmayan inaktivasyon ile karakterli klonalitenin gösterilmesi ile neoplazi olarak kabul edilmiştir (5).
- ▶ 16p kromozomunda (TSC2) heterozigote kaybı ve sık gen mutasyonu keşfedilmiştir (6-8).

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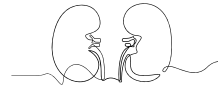
- ▶ İntraglomerüler lezyonlar Katepsin K, parvalbumin ve STING immünohistokimyasal pozitifliği ile tanınabilir (4).
- ▶ Kistik alanlar içeren AML ise fibrotik duvar, küçük perivasküler epiteloïd hücrelerin varlığı ve HMB45 pozitifliği işe yarar. Perivasküler epiteloïd hücreler bazen makrofaj kümesi ile karışabilir. Bu durumda her iki hücre tipinde de pozitif olan KatepsinK işe yaramaz. Kistleri döşeyen epitelyal hücreler keratin, PAX2 ve PAX8 ifade ederler. Stromal hücreler ise HMB45, Melan-A, KatepsinK, CD10, ER ve PR pozitifdir. Kist dışı solid bileşen ise HMB45, ER, PR, SMA ve Desmin pozitifdir (30,31). Bu tümörler mikst epitelyal ve stromal tümör, kistik nefroma (CD10, ER, PR pozitif, HMB45, Melan-A, KatepsinK negatif) ve daha nadiren düşük malignite potansiyelli multiloküler kistik berrak hücreli renal hücreli neoplazi ile karışabilir (4).
- ▶ Onkositom benzeri AML; nadir eozinofilik kistik böbrek hücreli karsinom, t (6;11) böbrek hücreli karsinom, onkositom ve kromofob böbrek hücreli karsinomun eozinofilik alt tipi ile ayrılmalıdır. HMB45 ve KatepsinK pozitifliği ile keratin negatifliği AML tanısını koydurur. Aynı immunhistokimyasal panel eozinofilik ve solid kistik böbrek hücreli karsinomlarda işe yarayabilir; ancak vakaların bir kısmında KatepsinK pozitifliği bildirilmiştir (4).

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