22. BÖLÜM

MİKST EPİTELYAL STROMAL TÜMÖR AİLESİ

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Erişkin kistik nefroma (EKN) ve mikst epitelyal stromal tümör (MEST), 2016 Dünya Sağlık Örgütü (DSÖ) sınıflamasına göre, klinik, histolojik ve immunohistokimyasal özelliklerinin benzerliği nedeniyle mikst epitelyal stromal tümör ailesi başlığı altına altında toplanmıştır (1).

ERİŞKİN KİSTİK NEFROMA

Epidemiyoloji ve klinik bulgular

Erişkin kistik nefroma, ilk defa 1892 yılında tanımlanan benign böbrek tümörüdür. Kadınlarda daha sık olup kadın/erkek oranı 8/1'dir. Genellikle 50-60 yaşlarında görülür. Sıklıkla insidental olarak saptanır, fakat ağrısız abdominal kitle, yan ağrısı, karın ağrısı ve daha az sıklıkta hematüri ile klinik bulgu verebilir (2, 3, 4).

Histogenez ve moleküler özellikler

EKN, klinik, morfolojik, immünohistokimyasal ve genetik özellikleri ile pediatrik kistik nefromadan farklıdır. Pediatrik kistik nefromaların tamamında görülen Dicer1 mutasyonu EKN'lerde günümüze kadar sadece bir olguda bildirilmiştir (5, 6, 7).

Makroskopi

Tümör tek taraflı ve düzgün sınırlıdır. İki milimetreden kalın olmayan septalar ile birbirinden ayrılan kistlerden oluşur. Genellikle çevre parankimden

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Ayırıcı tanı

MEST ayrıcı tanısında kistik nefroma, multiloküler kistik renal hücreli karsinom yer alır. Malign MEST'te leiomyosarkom, sinoviyal sarkom, sarkomatoid renal hücreli karsinom vardır (26, 18). Morfolojik bulguların yanısıra immunhistokimyasal belirteçler ile ayrım mümkündür.

SONUÇ

Mikst epitelyal stromal tümör ailesi başlığı altında, erişkin kistik nefroma ve mikst epitelyal stromal tümörler mevcuttur. Histopatolojik ve immunohistokimyasal incelemelerin yanı sıra klinik bilgilerin bilinmesi lezyonların tanınmasında, pediatrik kistik nefromadan ayırt edilmesinde ve genetik incelemelerin yapılabilmesi açısından yol gösterici olacaktır.

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