BÖLÜM 14

ADRENOKORTİKAL KARSİNOMLAR

Özlem AYDIN İSAK[1]

GİRİŞ

Adrenokortikal karsinomlar (AKK), nadir görülen, sıklıkla agresif seyreden tümörlerdir. Bazen fonksiyonel olup cushing sendromuna veya virilizasyona sebep olarak, bazen de fonksiyonel olmayan batın içi kitle ile veya tesadüfi bir bulgu ile ortaya çıkarlar.

Epidemiyoloji

İnsidansı ABD' de milyonda 0,72 iken dünya çapında milyonda 0,5-2'dir(1). İlginç bir şekilde, bir dizi çevresel ve genetik risk faktörünün tanımlandığı Güney ve Güneydoğu Brezilya'daki çocuklarda, insidans yaklaşık 10-15 kat daha yüksektir(2). AKK, iki modlu bir yaş dağılımı gösterir, 5 yaş altı çocuklarda ve 40-60 yaş erişkinde sıklıkla görülür. Çocukluk kanserlerinin %1,3' ünü, yetişkin kanserlerini %0,02-0,2'sini oluşturur. Kadınlarda, erkeklere kıyasla %1,5-2,5 kat fazla görülür. Yetişkinlerde ACC ile ilişkili durumlar arasında Li-Fraumeni sendromu, Ailesel Adenomatoz Polipozis (FAP), Multiple Endokrin Neoplazi tip I ve Lynch sendromu sayılabilir (1, 3, 4).

Klinik

AKK, üç farklı klinik durumla karşımıza gelebilir. Hastaların yaklaşık %40-60'ı artmış hormon salınımına bağlı semptomlarla, üçte biri spesifik olmayan semptomlarla ve %20-30' u başka nedenlerle yapılan görüntülemeler sonucunda tesadüfen tanı alır (4, 5).

Hormon salınımına bağlı semptomlar hiperkortizolemi ve hiperandrojenemiye bağlıdır. Hiperkortizolemi pletore, diabetes mellitus, osteoporoz ve kas atrofisine neden olabilir. Eş zamanlı olarak glukokortikoid aracılı mineralokortikoid reseptör aktivasyonu, hipokalemi ve hipertansiyon ile kendini gösterebilir. Hiperkortizolemi ile prezente olanlarda genellikler hızla ilerleyen ve belirgin kas güçsüzlüğü de eşlik edebilir. Hiperandrojenemi erkek tipi kellik, virilizasyon, hir-

[1] Uzm. Dr., Düzce Atatürk Devlet Hastanesi, Tıbbi Onkoloji Kliniği, ozlemaydin_87@hotmail.com

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