

26. BÖLÜM

METASTATİK ADRENOKORTİKAL KARSİNOMDA MEDİKAL TEDAVİ YAKLAŞIMLARI

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GİRİŞ

Adrenokortikal kanser (AKK), yılda 1/1.000.000 kişi insidansı ile nadir ve agresif bir malignitedir (1). AKK her yaşta gelişebilse de beş yaşından önce ve yaşamın dördüncü ile beşinci dekadında hastalık iki ayrı pik yapmaktadır (2). AKK kadınlarda daha sıktır (% 55-60). AKK çoğunlukla sporadiktir, ancak multipl endokrin neoplazi tip 1, Li-Fraumeni, Lynch, ailesel adenomatöz polipozis koli ve Beckwith-Wiedeman sendromları gibi kalıtsal tümör sendromlarının bir parçası olabilir (3). AKK kötü prognoza sahiptir, prognozu belirleyen en önemli faktör hastalık evresidir, 5 yıllık sağkalım evre I olan hastalarda %60-80'den evre IV olan hastalarda %13'e kadar değişmektedir (4). Hastaların yaklaşık üçte birinde tanısında uzak metastaz mevcuttur (5). Opere olan AKK tanılı hastaların yaklaşık %25'inde rezidüel hastalık olduğu ve radikal rezeksiyon geçiren hastaların yarısı uzak metastazla nüksetmektedir (6). Metastatik veya nüks adrenokortikal karsinom için küratif bir tedavi yoktur (7).

MEDİKAL TEDAVİLER

Mitotan

Mitotan; inoperabl veya metastatik AKK için anahtar bir tedavidir. Mitotan monoterapisi lokal ileri veya metastatik hastalık durumlarında incelenmiş olup mevcut bilgilerin kanıt değeri düşük ve sonuçlar da oldukça değişkendir, parsi-

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hastanın akciğer metastazlarında regresyon ve karaciğer lezyonlarında progresyon ile mixt yanıt sağlanmıştı (PFS 2.1 ay ve OS 24.9 ay) (39).

Ne yazık ki immünoterapi birçok nedenden ötürü AKK'de sınırlı etkiye sahiptir.

Nivolumab, ipilimumab (ClinicalTrials.gov NCT02834013 ve NCT03333616) ve pembrolizumab (ClinicalTrials.gov NCT02721732) ile kombine tedavi denemeleri devam etmektedir.

SONUÇ

Metastatik AKK'nin prognozu kötüdür ve tedavi seçenekleri sınırlıdır. Birinci basamakta EDP-Mitotan kombinasyon kemoterapisi hala standart tedavidir. İkinci ve üçüncü basamakta kanıtlanmış etkin bir tedavi rejimi maalesef yoktur. Umut vaadedebilecek immünoterapi çalışmaları devam etmektedir. Mevcut tedavilerin çoğu hastalar için hayal kırıklığıyla sonuçlanmaktadır ve bu yüzden daha etkili tedavi seçenekleri için çok kapsamlı klinik çalışmalara ihtiyaç vardır.

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