Bölüm
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TİMUSUN NÖROENDOKRİN TÜMÖRLERİNDE CERRAHİ YAKLAŞIM

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GİRİŞ

Timüs tümörleri anterior mediastenin en sık görülen tümörleridir. Timusun epitelyal tümörleri üç gruba ayrılır. Bunlar, timoma, timik karsinomlar ve timusun nöroendokrin tümörlerdir (TNET). Bu bölümün konusu olan TNET' ler, timik tümörler içerisinde nadir görülen neoplazmalardır ve tüm timik tümörlerin % 5' ini oluştururlar. İlk defa 1972 yılında Rosai ve Higa tarafında tarif edilmiştir (1). Yaklaşık oranı 0.001/100.000'dir. Erkeklerde kadınlardan üç kat daha fazla görülmektedir (2). Nöroendokrin tümörler vücudun birçok organında tanımlanmıştır. TNET' ler tüm nöroendokrin tümörlerin % 0,4' ünü oluşturur ve sağ kalımları timüs dışında gelişen nöroendokrin tümörlerden (NET) daha kötüdür. Paraneoplastik sendromların eşlik etme olasılığı daha fazladır. TNET' lerde komplet tümör rezeksiyonundan sonra bile lokal nüks ve uzak metastaz olasılığı yüksektir ve 5 yıllık sağ kalımlar % 30 - % 70 arasında bildirilmiştir (3-4-5). TNET' lerin standart tedavisi rezektabl tümörler için cerrahidir. Unrezektabl tümörlerde neoadjuvan kemoterapi (KT) ve/veya radyoterapi (RT) rezeksiyon şansını arttırmak için kullanılır. İmmünoterapi son yıllarda unrezektabl tümörlerde kullanılan bir tedavi seçeneğidir.

SINIFLANDIRMA

Timik tümörlerde ilk sınıflandırma 1999 yılında dünya sağlık örgütü (DSÖ) tarafından yayınlanmış, sonrasında 2004 ve 2015 yıllarında revize edilmiştir (6). Timik karsinomlar, iyi prognozdan kötü prognoza doğru sınıflandırılmıştır. TNET' ler önceleri timik karsinomların bir varyantı olarak görülmüşse de farklı histopa-

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bazı çalışmalarda prognostik faktör olarak görülmese de, özellikle postoperatif RT' in sağ kalıma etkisi birçok çalışmada ortaya konmuştur (64-65). Paraneoplasitk sendrom varlığı prognostik belirteç olmadığını gösteren çalışmalar mevcutsa da, daha geniş serilerde kötü prognostik faktör olduğu görülmüştür. TNET ile cushing sendrom veya MEN-1 birlikteliğinin 5 yıllık sağ kalımı % 65' den % 35'e düşürdüğünü gösteren çalışmalar mevcuttur (66). Nodal tutulumun kötü prognostik faktör olduğu birçok çalışmayla ortaya konmuştur. Bu yüzden doğru sağ kalım analizleri için peroperatif lenf nodu örneklemesi yapılmalıdır (67-68). Nüks gelişen olgularda reoperasyon yapılması sağ kalımı arttırmaktadır (69). Yüksek rekürens ve uzak metastaz ihtimallerinden dolayı, TNET hastalarının yakın takibi önemlidir. Önerilen, ilk 3 yıl her altı ayda bir defa BT ile tetkik edilmesidir.

Sonuç

TNET' ler, diğer timik tümörlerden daha az görülen, tanı anında daha ileri evrelerde tespit edilen agresif tümörlerdir. Hastalar lokal bası bulguları ile başvururlar. Tanıda en etkili yöntem TTİA' dır. Radyolojik olarak kontrastlı toraks BT yeterli değerlendirmeyi sağlamaktadır. Erken evrede tedavi cerrahi iken, lokal ileri evre tümörlerde multidisipliner tedaviler gerekmektedir. En sık kullanılan cerrahi yaklaşım büyük tümör çaplarından dolayı median sternotomidir. Rezeksiyonu zor olan tümörlerde, neoajuvan KT ve/veya RT ile tümör boyutu küçültülüp rezektabl sınırlara getirilebilir. R0 rezeksiyonda dahi, lokal ileri evre tümörlerde PORT önerilmektedir. R0 rezeksiyon ve hastalığın evresi en önemli prognostik faktörlerdir. Unrezektabl ve metastatik tümörlerde immünoterapi umut verici sonuçlar sağlamaktadır.

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