BÖLÜM 16

NADİR PANKREAS TÜMÖRLERİ

Esma YETİM[1]

GİRİŞ

Pankreas kanseri, kadın ve erkeklerde kanser ilişkili ölümlerde dünya genelinde 7. sıradadır. 45 yaşından önce nadir olup yaşla birlikte insidansı artar. İnsidansın pik yaptığı yaş erkeklerde 65-69, kadınlarda 75-79'dur (1). Vakaların büyük çoğunluğu vasküler invazyon nedeniyle cerrahiye uygun olmayıp ancak %15-20 vaka rezektabl evrede yakalanabilmektedir (2). Obezite, fiziksel inaktivite, sigara, alkol, yüksek kalori alımı, asetilsalisilik asit ve non-steroid anti-inflamatuar ilaçlar, helikobakter pylori, HBV, HCV, diabetes mellitus, insülin direnci, aile öyküsü, genetik predispoazan faktörler, ABO kan grubu ve kistik fibrozis pankreas kanseri risk faktörleri olarak gösterilmiştir (3, 4). Bu kanserlerin yaklaşık %85-95'ini pankreas duktal adenokarsinomu, %1-5'ini nöroendokrin tümörler oluşturmakta olup geriye kalan %1-2'lik bir kısmını nadir subtipler oluşturmaktadır (3, 5). Bu bölümde pankreasın nadir primer tümörlerini oluşturan pankreatoblastom, pankreas asiner hücreli karsinom, pankreas berrak hücreli karsinom, primer pankreas lenfomaları, primer pankreatik sarkom, primer pankreatik skuamöz hücreli karsinom ve pankreasın sekonder metastatik tümörleri ile ilgili literatürdeki bulguların derlenmesi amaçlandı.

Pankreatoblastom

Yetişkinlerde pankreas kanserlerinin %1'den daha azı pankreatoblastomadır (6). Çocuklarda ise 1. dekatta pankreas kanserlerinin %2' sini oluşturur (7). Ortalama tanı yaşı erişkinde 41 (18-78) olup erkek/kadın oranı 1.2:1'dir (8). Etyoloji bilinmemekte olup çoğu vaka sporadiktir (9). Yetişkinlerde ailesel adenomatöz polipozis ile birlikte tanımlanmış birkaç vaka vardır (10). En yaygın görülen semptom karın ağrısıdır (8). Karında kitle, kilo kaybı, bulantı, sarılık ve ishal ile de prezente olabilmektedir (11). Nadiren bazı hastalar üst gastrointestinal kanama ile prezente olabilir (12). Ca 19-9 ve CRH sekresyonu görülebilir (13). AFP yüksekliği yetişkinlerde sık değildir (14). AFP yüksek saptanması halinde nüks veya progres-

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lezyon olarak tanımlanmıştır ((109, 112). Bu tipte ek olarak anjiografik olarak hipervaskülarite gözlenmektedir (111, 114). Tanısında diğer pankreas kitlelerinde olduğu gibi EUS aracılı biyopsi uygun yöntemdir. Literatürde tanıda endoskopik retrograd kolanjiopankreatografi kullanılan vakalar da bildirilmiştir (114). Ancak mümkün ise kitlenin cerrahi çıkarılarak patolojik tanının kesinleştirilmesi gerekir (109). Labaratuar bulgusu olarak hiperkalsemi ve çeşitli fizik anomaliler bildiren çalışmalar mevcuttur (108). Tedavisi zor olup kemoterapi ve radyoterapiye yanıtı kötüdür (110, 112). Altı hastanın dahil edildiği bir çalışmada ortalama sağ kalım 1-7 ay olarak bildirilmiştir (112).

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