

PANKREAS NÖROENDOKRİN TÜMÖRLERİ

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GİRİŞ

Pankreas nöroendokrin neoplazmlar (tümörler) (pNET veya Pan-NEN), pankreasın endokrin hücrelerinden kaynaklanan nadir tümörlerdir ve bu nedenle nöroendokrin tümörler (NET'ler) grubuna aittirler. Dünya Sağlık Örgütü (DSÖ) kriterlerine göre pNET'ler, proliferasyon belirteci Ki-67'nin ekspresyon düzeyine göre farklı gruplara ayrılmaktadır. Grade 1 pNET'lerde Ki-67 pozitifliği < %3, Grade 2 pNET'lerde %3-20 ve Grade 3 pNET'lerde >%20'dir (1). Histolojik farklılaşma ve Ki-67 pozitiflik yüzdesinin birleşimine göre, nöroendokrin tümörler (NET'ler) ve nöroendokrin kanserler (NEK) olarak ikiye ayrılır (2,3). NEK'ler tüm NET'lerin yalnızca %10-20'sini oluşturur (3). pNET'ler fonksiyonel (aşırı hormon üreten) ve non-fonksiyonel (aşırı hormon üretimi olmayan) olarak alt bölümlere ayrılabilir (4). Fonksiyonel olmayan pNET'lerin (NF-pNET) prognozu genel olarak daha kötüdür; çünkü pankreas adenokarsinomuna benzer şekilde çok semptoma neden olmadan büyüyüp yayılabilirler, aksine fonksiyonel pNET'ler sıklıkla spesifik hormonların aşırı üretiminin neden olduğu semptomlar nedeniyle kolay teşhis edilir.

EPİDEMİYOLOJİ

pNET'lerin çoğu sporadiktir ancak bazıları, kromozom 11 üzerindeki MEN-1 tümör baskılayıcı gendeki mutasyonların neden olduğu otozomal dominant kalıtsal bir sendrom olan tip 1 çoklu endokrin neoplazi sendromu (MEN-1) gibi genetik mutasyonlarla yakından ilişkilidir. pNET'ler, MEN-1 hastalarının %30-80'inde, von-Hippel Lindau (VHL) hastalığının %15'inde ve Tip 1 Nörofibromatoz'un %10'unda görülür. Tuberoz skleroz (TSC) ile de ilişkilendirilebilir ve pNET'ler %2-9 oranında görülür. Bu sendromlarla ilişkili pNET'ler sıklıkla multifokaldir ve sporadik neoplazmlara kıyasla hastalığın başlangıcı daha erkendir (3,5).

Fonksiyonel pNET'lerin genel prevalansı düşüktür ve yaklaşık 100.000'de 1 olarak bildirilmiştir. Buna karşılık, otopsi çalışmalarında pNET prevalansı %0,5 ila %1,5'tir. Genel pNET insidansı yaşla birlikte artar; altıncı ve yedinci dekatlarda zirve yapar. Amerika Birleşik Devletleri'ndeki SEER veri tabanından elde edilen veriler, mevcut görülme sıklığının 100.000'de 0,8 olduğunu göstermiştir (6).

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