BÖLÜM 37

NÖROENDOKRİN TÜMÖRLER

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GİRİŞ

Nöroendokrin tümörler (NET) vücudun herhangi bir bölgesindeki nöroendokrin sistemden köken alan bir tümör grubudur. En sık gastrointestinal sistemde görülür. Obendorfer, ilk olarak 1907'de gastrointestinal sistemin tipik adenokarsinomlarına göre daha benign karakterde olduklarını belirtmek amacıyla 'karsinoid' terimini kullanmış ve NET'ler karsinoid tümörler olarak da adlandırılmıştır (1). Gastrointestinal (GI) sistemin NET'leri gastrointestinal nöroendokrin tümörler (GI-NET) ve pankreatik nöroendokrin tümörleri (pNET) içermektedir. GI-NET'ler tüm karsinoid tümörlerin yaklaşık %70'ini oluşturur. Diğerlerinin çoğu solunum sisteminde bulunur. GI-NET'ler en sık ince bağırsak, rektum, apendiks ve midede görülür. İnce bağırsakta, ileum en yaygın bölgedir, bunu duodenum ve jejunum takip eder. pNET'ler, tümörden salınan hormonun neden olduğu bir klinik sendromla ilişkili ise fonksiyonel veya hormon üretimine neden olmayan non-fonksiyonel tümörlerden oluşur.

EPİDEMİYOLOJİ

GI-NET'lerin genel insidansı 25-50/10^6, klinik olarak anlamlı GI-NET insidansı 7-13/10^6, otopside herhangi bir GI-NET görülme insidansı 84/10^6'tür ve bunların çoğu klinik olarak sessizdir. GI-NET insidansı son 30 yılda önemli ölçüde artmıştır. Bu artışın ana nedeninin endoskopik prosedürlerin daha fazla kullanılması ve daha iyi görüntüleme teknikleriyle ilişkili olduğu düşünülmektedir. pNET'ler pankreastan kaynaklanan tümörlerin %1-10'unu oluşturur. Fonksiyonel pNET'lerin genel prevalansı düşüktür; yaklaşık 10/10^6 olduğu bildirilmekle birlikte insidans değişkendir. En sık 0.5-3/10^6 ile insülinoma ve gastrinomalar görülmektedir (2).

SINIFLANDIRMA VE PATOGENEZ

Nöroendokrin tümörler; histopatolojilerine, anatomik kökenine ve fonksiyonel olarak aktif olup olmadıklarına göre değerlendirilmiş ve sınıflandırılmıştır. GI-NET'ler anatomik ve embriyolojik kökenine göre üç grupta incelenmiştir; Foregut; (Akciğer, mide, duodenum), Midgut; (ince barsak, appendiks, sağ kolon), Hindgut; (distal kolon, rektum).

2000 yılında Dünya Sağlık Örgütü (DSÖ), NET'leri anatomik bölgesine bakılmaksızın üç histolojik kategoride tanımlamıştır. Bunlar, 1; iyi diferansiye nöroendokrin tümörler veya karsinomlar, 2; kötü diferansiye endokrin tümörler ve 3; mix ekzokrin-endokrin tümörler olarak sınıflandırıldı.

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larda ilk tercih endoskopik rezeksiyon olmalıdır. Endoskopik rezeksiyon için uygun olmayan vakalarda tedavinin temeli mümkünse küratif amaçlı cerrahidir. Cerrahinin mümkün olmadığı hastalarda ise oktreotid, lanreotid, everolimus ve sunitinib gibi medikal tedavi seçenekleri de bulunmaktadır.

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