37.
BÖLÜM

KARSİNOİD SENDROM KLİNİK VE BİYOKİMYASAL ÖZELLİKLERİ

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GİRİŞ

Karsinoid sendrom, sindirim sistemi ile akciğerlerin bazı iyi diferansiye nöroendokrin tümörlerinin (NET) çeşitli biyoaktif maddeler salgılaması sonucu oluşan semptomlar kümesine verilen isimdir (1). Karsinoid sendrom oluşumundan sorumlu başlıca biyoaktif maddeler Tablo 1'de gösterilmiştir. NET'ler gastrointestinal sistemin herhangi bir yerinde, akciğerlerde veya vücudun faklı yerlerinde ortaya çıkabilir. NET'lerin görülme sıklığı 100.000 kişi başına 2.7 iken, karsinoid sendrom görülme sıklığı 100.000 kişi başına 0.27'dir (2). Karsinoid sendrom özellikle karaciğer metastazı olan hastalarda daha yaygın görülmektedir. Normalde karaciğer, portal dolaşıma salgılanan biyoaktif ürünleri inaktive etmekten sorumludur. Karaciğer metastazı olan olgularda bu fonksiyon bozulmakta ve tümörden salgılanan biyoaktif ürünler aktif halde dolaşıma geçerek karsinoid sendroma neden olmaktadır (3). Karsinoid sendrom erkek ve kadınlarda eşit oranda görülürken, Afrikalı ve Amerikalılarda diğer etnik gruplardan daha fazla görülmektedir (1,2).

Karsinoid sendrom vakalarının büyük çoğunluğu orta bağırsaktan (jejunum, ileum ve çekum) kaynaklanan metastatik tümörlerle ilişkili olup az bir kısmı akciğer, distal kolon veya rektumdan (ön bağırsak ve arka bağırsak kökenli) çıkan bir NET'le ilişkilidir (4,5). Mide ve akciğer NET'leri atipik karsinoid sendromlarla ilişkili olabilir (6–8). Pankreas NET'lerinin yaklaşık % 1'i karsinoid sendroma ne-

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SONUÇ

Karsinoid sendrom, nöroendokrin tümörlerden serotonin ve diğer maddelerin salınması sonucunda gelişen, kutaneöz, gastrointestinal, kardiovasküler ve respiratuar bulguları olan klinik bir antitedir. Genellikle karaciğer metastazı olan orta bağırsak (jejunum, ileum ve çekum) kaynaklı NET'lerde daha sık görülür. Karsinoid sendromun tipik klinik semptomları epizodik flushing ve ishaldir. Atipik belirti ve semptomlar arasında wheezing, karın ağrısı, kalp kapak hastalığı, telenjektazi, pellagra ve fibrosiz gelişimine bağlı komplikasyonlar (üreteral obstrüksiyon, intestinal obstrüksiyon, intestinal iskemi) yer alır. Bu semptomlara genellikle serotonin, histamin, kallikrein, prostaglandinler ve taşikininlerin salınması neden olur. Karsinoid sendrom tanısında en yararlı başlangıç testi, serotonin metabolizmasının son ürünü olan 5-HİAA'nın 24 saatlik idrarla atılımını ölçmektir. Kromogranin A düzeyinin karsinoid sendrom tanısı için tarama testi olarak kullanılması önerilmemektedir. Kromogranin A düzeyinin tanısı olan hastalarda hastalık progresyonunu, tedaviye yanıtı veya cerrahi rezeksiyondan sonra nüksü değerlendirmede kullanılması önerilmektedir.

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