BÖLÜM 18

APENDİKS VERMİFORMİS TÜMÖRLERİ

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GİRİŞ

Apendiks neoplazmaları, değişen malign potansiyel sergileyen heterojen bir tümör grubunu içerir. Son yıllarda insidansı bir milyon kişi başına altı vaka ile nadir görülen tümörlerdir.[(1)] Apendiks neoplazmlarının prognozu ağırlıklı olarak tümör tipine ve derecesine bağlıdır ve uzun süreli sağkalım %10 ile %90 arasında değişmektedir.[(2)]

Apendiks neoplazmlarının beş ana histopatolojik alt tipi vardır: non-epitelyal tümörler olan nöroendokrin tümörler (NET), epitelyal tümörler olan müsinöz neoplazmalar, goblet hücreli adenokarsinomlar (GHA), kolonik tip (müsinöz olmayan) adenokarsinomlar ve taşlı yüzük hücreli adenokarsinomlar.

Apendiks neoplazmlarının tanı ve tedavisi, histopatolojik ve moleküler özelliklerine ilişkin anlayışımız geliştikçe giderek daha farklı hale geldi. Örneğin, daha önce "goblet hücreli karsinoidler" olarak adlandırılan neoplazmlar, "goblet hücreli adenokarsinom" olarak yeniden adlandırılmıştır, çünkü bu tümörlerin nöroendokrin bileşeninin daha az önemli olduğu anlaşılmıştır. Ayrıca, apendiksin primer ve metastatik müsinöz neoplazmlarının biyolojilerini daha iyi yansıtmak için daha spesifik evreleme ve derecelendirme sistemleri geliştirilmiştir.

Primer apendiks tümörlerinin apendiks ile sınırlı olduklarında preoperatif olarak teşhis edilmesi zordur. Apendektomi örneklerinin yaklaşık %1'inde bulunan tümörler, sıklıkla akut apandisiti andırır tablo ile karşımıza çıkar.[(3)] İntraoperatif olarak tanı genellikle belirsiz kalır. Cerrah maligniteden şüphelense bile, ilişkili inflamasyon kesin intraoperatif tanıyı engelleyebilir. Sonuç olarak, çoğu apendiks kanseri vakası ameliyat sonrası teşhis edilir.

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AA için sistemik kemoterapi önerilir, ancak eklere özel kılavuzlar yoktur. Üst düzey verilerin olmaması nedeniyle, AA'lı hastalar için öneriler kolon kanseri için tedavi algoritmalarını takip eder. Apendikse sınırlı metastatik olmayan ve lenf nodu tutulumu olmayan hastaların sistemik tedaviye ihtiyacı yoktur. Tamamen rezeke edilmiş lokalize tümörleri olan ve lenf nodu tutulumu olan hastalar, bir floropirimidin/oksaliplatin ikilisi ile adjuvan kemoterapiden fayda görebilir. Metastatik hastalığı olan hastalar, metastatik kolorektal kanser için kullanılanlara benzer sistemik rejimler almalıdır.

SONUÇ

Apendiks tümörleri için kemoterapi seçeneklerindeki nüansları tanımlayan küçük seriler vardır. Çoğu, fluorourasil, platin ve irinotekan kombinasyonu ile rejimleri kullanır. EGFR inhibitörlerinin apendiks KRAS wild tip tümörler üzerinde kolorektal tümörlere göre daha az etkiye sahip olduğu öne sürülmüştür. EGFR inhibitörleri, sol taraflı tümörleri olan hastalarda sağ taraflı tümörlere kıyasla daha fazla fayda göstermiştir ancak apendiks tümörleri genellikle bu çalışmaların dışında tutulmuştur. AA'nın belirli sistemik rejimlere daha olumlu yanıt verebilecek hedeflenebilir yönlerini anlamak için daha fazla çalışmaya ihtiyaç vardır.

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