

BÖLÜM 9

MALİGN SEKS KORD - STROMAL TÜMÖRLERDE YÖNETİM

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Seks kord stromal tümörler ,tüm over malgniteleri arasındaski yaklaşık %5-8 oranına sahiptir.^{2,3} Heterojen nadir bir neoplazi grubu olan bu tümörler; seks kordlarından, over matriksinden veya mezenkimden köken alırlar. Granüloza ve teka hücreleri veya leydig ve sertoli hücreleri gibi undiferansiye hücre gruplarının farklı kombinasyonlarından oluşurlar. Bu tümörler hormon üretme potansiyeline sahiptirler ve hormon üreten tümörlerin yaklaşık %90'ı seks kord stromal tümörlerdir. Bu tümörler ,östrojen ve androjenler dahil seks steroidleri üretirler.^{4,5} Olgular genellikle bu hormonların fazlalığı, semptom ve bulguları ile başvururlar.

Maligen seks kord stromal tümörler, epitelyal over neoplazmalarının aksine daha erken tanı alırlar ve tanı konduklarında maligen de olsalar daha düşük derecelidirler.⁶ Bu nedenle platin bazlı kemoterapi, çok az hasta için gereklidir. Tümörün nüks etmesi sıklıkla tedaviye kötü yanıt verir ancak tümörün yavaş ilerlemesi nedeni ile hastalar yıllarca yaşayabilirler. Hastalığın erken tanı alması, tanı esnasında tümörün düşük dereceli olması, cerrahi ile kür sağlanabilmesi nedeni ile prognoz çok iyidir.

EPİDEMİYOLOJİ

Seks kord stromal tümörler over kanserlerinin alt tipleri arasında en az görülenlerdir. Yaşa bağlı sıklık oranı epitelyal over kanserlerinden daha azdır ve maligen germ hücreli tümörlerin yarısı kadardır.⁷ Sebebi tam olarak açıklanamasa da siyah kadınlarda, bu tümörlerin gelişme olasılığının daha fazla olduğu yapılan

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Seks-kord stromal tümörlerden; granüloza hücreli tümörler, fibrosarkomlar, sertoli-leydig hücreli tümörler, annüler tübüllü seks kord tümörleri (Peutz-Jeghers sendromu ile ilişkisiz), steroid hücreli tümörler (orta ve kötü diferansiye) için evreleme gereklidir. Tekoma, fibroma, sklerozan stromal tümör, jinandrobalstom, sertoli-leydig hücreli tümörler (iyi diferansiye), annüler tübüllü seks kord tümörler (Peutz-Jeghers sendromu ile ilişkili) için evreleme gerekli değildir.

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