



Bölüm 37

Çocukluk Çağı Yumuşak Doku Sarkomları

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Giriş

Çocukluk çağı yumuşak doku sarkomları mezanşimal hücrelerden kaynaklanan heterojen malign tümörler olup tüm pediatrik kanserlerin %6-8'ini oluştururlar (1, 2). Rabdomyosarkom (RMS) ve rabdomyosarkom dışı yumuşak doku sarkomları olmak üzere iki başlık altında incelenebilir:

Rabdomyosarkom

Epidemiyoloji ve Risk Faktörleri

Çocukluk çağı sarkomların %40'dan sorumludur (1, 2). Erkek-kadın oranı yaklaşık 1.5-1.0'dir (3). Embriyonal RMS'lar 2-6 yaş, alveolar RMS ise 15-19 yaşta pik yapmaktadır (4-6). Tümörün en sık yerleşim yeri sırasıyla, %40 baş boyun bölgesi (parameningeal %25, orbita %9, non-parameningeal %6), %30 genitoüriner, %15 ekstremitelerde, %15 gövdedir (7). Baş boyun bölgesinde en sık embriyonal RMS, genitoüriner bölgede botryoid RMS, ekstremitelerde ve gövde de alveolar RMS gözlenmektedir (8).

RMS'ların nedeni bilinmemekle beraber bazı genetik sendromlarla birlikteliği bulunmaktadır. Nörofibromatozis tip 1, Costello sendromu, Noonan sendromu, pleuropulmonary blastoma, Li-Fraumeni sendromu ve Beckwith-Wiedemann sendromu RMS ile ilişkili bulunan sendromlardır (4).

Biyolojik Karakteristiği ve Patolojisi

RMS çocukluk çağının küçük, yuvarlak, mavi hücreli malignenssidir. Morfolojik olarak RMS hücreleri, farklılaşmamış yuvarlak hücreler, oval hücreler, "kurbağa yavrusu benzeri" hücreler, iç şeklindeki hücreler ve tamamen farklılaşmış rabdomyoblastlar dâhil olmak üzere heterojen şekillere sahiptir (9). RMS'un histolojik alt tipleri, embriyonal (klasik, botryoid), alveolar, spindle/sclerosing, pleomorfik RMS olarak sınıflandırılmaktadır (10). Embriyonal tip, RMS'un en yaygın formu olup, orbita, baş boyun ve genital bölgede sıklıkla tespit edilmektedir (11). Embriyonal RMS, çapraz çizgili kas hücrelerine farklılaşma eğilimi gösteren blastemal mezanşimal hücrelerden oluşmaktadır. Embriyonal RMS'de 11p15

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bariyerler göz önünde bulundurularak değişiklik yapılabilir. Planlanan hedef volüme (PTV), tipik olarak yaklaşık 5 mm olan hastaya özgü bir marj eklemelidir.

Takip

RMS ve RMS dışı sarkomlarda hastalığın takibi ilk 1 yıl için 3 ayda bir anemnez, fizik muayene, laboratuvar çalışmaları, primer tümörün yerleştiği alanın MR'ı ve toraks BT'yi (RMS dışı sarkomlarda 6 ayda bir) içermektedir. İkinci ve üçüncü yıllarda 6 ayda bir, 4-5. yıllarda ise yılda 1 kez olmak üzere yukarıda adı geçen tetkikler tekrarlanmalıdır (39).

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