Bölüm 38

Çocukluk Çağı Kemik Sarkomları

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Giriş

Ewing sarkomu (EWS) ve osteosarkom (OS) pediatrik ve adölesan yaş grubunda en sık görülen iki malign kemik tümörüdür (1,2).

Ewing Sarkom

Epdemiyoloji ve Risk Faktörleri

EWS, çocukluk çağında kemiğin en sık görülen ikinci tümörüdür (3). Daha önceleri extraosseöz EWS ve malign periferal nöroektodermal tümör (PNET) olarak kabul edilip, farklı tedaviler uygulanmakta olan bu tümörler Ewing sarkom ailesel tümörler (EWSFT) başlığı altında toplanmıştır (4). EWSFT'lerinin %87'si kemiğin EWS'udur. EWS genelde 10-15 yaş aralığında, beyazlarda ve erkeklerde daha sık görülmektedir (5, 6). Sekiz yaş öncesi ve 25 yaş sonrası nadir görülmektedir. Yerleşim yeri olarak en sık ekstremite (%53), aksiyal iskelet (%47) de lokalizedir. Ekstremite olarak en sık femurda, aksiyal iskeletten de en sık pelvis kemiklerinde yerleşim göstermektedir (5).

Biyolojik Karakteristiği ve Patoloji

EWS, kemiğin farklılaşmamış mavi yuvarlak hücreli bir tümörüdür. Kemik iliğindeki mezenşimal projenitör veya kök hücrelerden köken alan Ewing hücrelerinden kaynaklandığı düşünülmektedir (7). ESFT'lerin yaklaşık %95'i, kromozom 22 üzerindeki EWS geni ile kromozom 11'deki FLI1 geni [t(11;22) (q24; q12)] veya kromozom 21'deki ERG geni [t(21;22) (q22;q12)] arasında bir translokasyona sahiptir (7, 8). Hastaların %85'den fazlasında, CD99 (MIC2) yüzey antijeni eksprese edilir (4, 9).

İmmunohistokimyasal çalışmalarda, hücre-yüzey glikoprotein p30/32 MIC2 (CD99), vimentin, HBA-71, B2-mikroglobulin pozitifliği görülmektedir. Bu markerler EWSFT'leri diğer küçük yuvarlak mavi hücreli tümörlerden ayırt etmek için kullanılmaktadır (4).

Yayılım Yolları

Tanı anında hastalık %75 lokalizedir. Sadece lokal tedavi uygulananların %80'inde uzak metastaz gelişmektedir. Bu da vakaların çoğunda tanı anında tanımlanamamış mikrometastazların ol-

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olmamasına rağmen, eksternal RT, polikemoterapi ve otolog hematopoietik projenitör hücre desteğiyle birlikte yüksek aktiviteli ^{153}Sm-EDTMP'nin multimodal uygulanabilirliği, rezeke edilemeyen osteosarkom için umut verici gözükmektedir (60).

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