

BÖLÜM 13

MEDULLOBLASTOM

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GİRİŞ

Medulloblastom santral sinir sisteminin (SSS) embriyonel tümörleri içerisinde yer almaktadır ve çocukluk çağının en sık görülen malign beyin tümörüdür ve esas olarak serebellumda görülür. Hastalık 40 yaşından sonra nadir olarak görülür. Ancak sıklığı düşük olmasına karşın ileri yaş hastalarda aynı zamanda risk altındadır. Hastalık ya serebellar hemisferlerde ya da vermiste lokalize olabilmektedir ve 4. ventrikül tutulumu olabilir. Bu yüzden obstrüktif hidrosefali tümörün dördüncü ventriküle olan yakınlığından dolayı nispeten yaygındır.

Diğer SSS embriyonel tümörleri gibi agresif tümör özelliklerine sahiptir. Genellikle derece 4 tümörlerdir.

Bu bölümde medulloblastomların epidemiyolojisi, klinik özellikleri, tanısı, yetişkin ve çocuklardaki hastalığın risk sınıflandırması, histopatolojisi, moleküler patogenezi, tedavisi, prognozu ve geç dönem komplikasyonlarından bahsedilmektedir.

EPİDEMİYOLOJİ

Medulloblastomlar sıklıkla çocukluk çağında görülmektedir. ABD’de her yıl yaklaşık 500 çocuğa medulloblastom tanısı konulmaktadır (1). Çocuklarda yetişkinlere nazaran 10 kat daha sık görülmektedir(2). 19 yaşından küçük çocuklarda merkezi sinir sisteminin tüm primer malign tümörlerinin yaklaşık yüzde 10 ila 20’sini oluşturan çocukluk çağının en yaygın malign beyin tümörüdür (1). Pik insidansı ise 5 ve 9 yaş arasındadır. Vakaların yaklaşık yüzde 70’i 20 yaşından önce tanı almaktadır. İnsidans 20 ila 24 yaş arasında hafif bir şekilde artış göstermekle birlikte hastalığın embriyonel orjinli olması ile tutarlı bir şekilde 4. Dekat sonrası nadiren görülmektedir.

Yetişkinlerde görülme insidansı milyonda 0,6’dır. Ancak literatürde hastalığın zaman içinde insidans hızı konusunda net bir fikir birliği bulunmamaktadır.

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