Bölüm 7

PROLENFOSİTİK LÖSEMİ

Okan YAYAR[1]

Prolenfositik lösemi(PLL),kan ve kemik iliğinde prolenfosit olarak adlandırılan aktive olmuş lenfosit hakimiyetiile karakterize nadir bir neoplazma grubudur. B veya T hücre kökenli olabilir. PLL, lenfoid lösemilerin %2'sinden azını oluşturur[1].

T HÜCRELİ PROLENFOSİTİK LÖSEMİ(T-PLL)

Giriş

T-PLL;periferikkan,kemikiliği,lenfnodu ve dalağın özellikle tutulduğu nadir bir Thücreneoplazmıdır. Bu hastalıktaki tümör hücreleri,post-timikThücre kökenlidir. Yaşlı yetişkinlerin hastalığı olup ortalama prezentasyon yaşı 61'dir[2]. T-PLL'nin E;K oranı 1,33 olup sıklık hafif bir şekilde erkek eğilimindedir[2]. T-PLL,ataksitelenjiektazi ve Nijmegenbreakage sendromu gibi kalıtımsal genetik bozukluklarda görülebilir.

Klinik

Çoğu vakada B semptomları, belirginlenfositoz (%75 vakada>100.000/microL),hepatosplenomegali(%75),generalizelenfadenopati(%50) saptanır[3]. Ek olarak cilt infiltrasyonu(%25) ve plevral ve peritonealserözeffüzyonlar(%15) görülür[3]. Cilt infiltrasyonları en sıklıkla gövde ve ekstremiteleri tutar. Eritemlimakülopapüler döküntü oluşturur. Ayrıca nodüler cilt tutulumu da görülebilir. T-PLL'de sıklıkla periorbital bölgede purpura ve ödem olarak ortaya çıkan bir yüz tutulumuda görülebilir[4,5]. Santral sinir sistemi tutulumu nadirdir(<%10).

Yaklaşık %10-15 vaka tanı esnasında asemptomatik yani indolent'tir.

Morfoloji

3 morfolojik varyant vardır[1-3]:

1. Tipik(%75): Tümör hücreleri,orta derecede yoğun kromatin ve görünür bir nükleolusa sahip orta büyüklükte lenfoid hücrelerdir. Nukleus yuvarlak veya oval olabilir. Sitoplazma granülsüz,hafifbazofilik olup sitoplazmik çıkıntılar(-kabarcıklar) yaygındır.

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