Bölüm
18

B VE T HÜCRELİ PROLENFOSİTİK LÖSEMİ

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B HÜCRELİ PROLENFOSİTİK LÖSEMİ

Epidemiyoloji

B hücreli prolenfositer lösemi (B-PLL), olgun B lenfosit malignitelerinin %1'inden azını oluşturan nadir lenfoproliferatif hastalıklardan biridir. İlk olarak 1970'lerde kronik lenfositer lösemi (KLL) varyantı olduğu düşünülse de Dünya Sağlık Örgütü'nün 2008 sınıflaması ve 2016 revizyonunda ayrı bir olgun B hücre hastalığı olarak tanımlanmıştır (1,2,3).

Medyan tanı yaşı 69 olup erkeklerde kadınlara göre biraz daha sık görülür (1,6/1) (4).

Morfoloji

PERİFERİK KAN: FAB sınıflamasına göre KLL/PLL ayrımında: periferik yaymada prolenfosit oranı %55'in üzerinde oluşu PLL, %10-55 arası prolenfosit oranı varlığı KLL/PLL, %10'un altında prolenfositer hücre varlığı ise KLL olarak tanımlanır (5).

B prolenfositler May-Grünwald Giemsa boyası ile boyandığında belirgin santral nükleoluslu, orta yoğunlukta kromatinli, zayıf bazofilik sitoplazmalı, düzgün sınırlı ve normal lenfositlerin yaklaşık 2 katı büyüklüktedirler (6).

KEMİK İLİĞİ: KLL'den farklı olarak proliferasyon merkezlerinin gözlenmediği, interstisyel ve intertrabeküler dağılımlı nodüler kemik iliği infiltrasyonu şeklindedir.

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Prognoz

Hastalık prognozu kötüdür. Konvansiyonel kemoterapi ile tedavi edilen eski vaka serilerinde medyan toplam sağkalım yaklaşık 7 aydır. Son yıllarda alemtuzumab ve pentostatin gibi yeni ilaçların kullanıma girmesi ile sağkalım iyileşmesi sağlanmaya başlanmıştır.

Sonuç

Prolenfositik lösemi, B hücreli ve T hücreli olmak üzere ikiye ayrılır. Her ikisi de agresif klinik gidişli ve kötü prognozlu lenfoid malignitelerdir. Splenomegali ile beraber yüksek lenfosit sayısının olması benzer özellikleri olmakla beraber her ikisinin biyolojik ve genetik özellikleri oldukça farklıdır. Bir grup hastada değişken sürelerde indolen seyir görülebilmesine rağmen progresyon kaçınılmazdır. Tedavi küratif değildir. Ancak yüksek yanıt oranlarına ulaşılıp remisyon süresi uzatılabilir. T-PLL'de 1. basamak tedavi intravenöz alemtuzumab; B-PLL'de TP53 normal hastalarda kombine kemo-immünoterapi, TP53 delesyon/mutasyonu olan hastalarda alemtuzumab veya BCR inhibitörleridir. Uygun hastalarda allojenik kök hücre nakli düşünülmelidir. T-PLL'de JAK-STAT, B-PLL'de BCR yolağını hedef alan güncel tedaviler gelecekte yeni tedavi yaklaşımları sağlayacak gibi görünmektedir (27).

Anahtar Kelimeler: B hücreli prolenfositik lösemi, T hücreli prolenfositik lösemi

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