

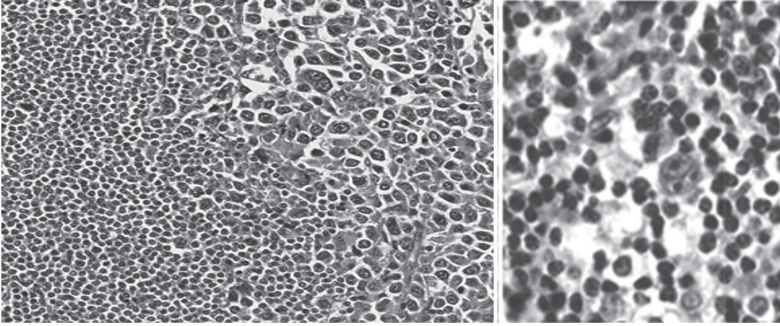
Bölüm 13

RİCHTER TRANSFORMASYONU TANI, KLİNİK VE TEDAVİ

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GİRİŞ

Richter transformasyonu (RT), Maurice Richter tarafından 1928 yılında tanımlanmıştır. Hastalarda daha önceden altta yatan kronik lenfositik lösemi / küçük lenfositik lenfoma (KLL / SLL) zemininde agresif büyük hücreli lenfoma gelişimi olarak tanımlanmıştır. Richter transformasyonu gelişen hastalarda en sık görülen histolojik alt tip Diffüz büyük B hücreli lenfomadır (DBBHL). Ayrıca daha az sıklıkta Hodgkin lenfoma ve T hücreli lenfoma gelişimi olarakta karşımıza çıkabilmektedir (Şekil 1).



Şekil 1. A) Richter transformasyonu gelişen kronik lenfositik lösemili (KLL) bir hastanın lenf düğümü. Sol taraftaki küçük lenfositler önceden var olan KLL'yi temsil eder, sağdaki büyük B hücreleri ile zıtlık gösterir ve dağınık bir büyük B hücreli lenfomaya dönüşümü temsil eder. **B)** Richter transformasyonu sonucu Hodgkin lenfoma gelişimi
Warnke RA, Weiss LM, Chan JK, et al. Tumors of the lymph nodes and spleen. Atlas of tumor pathology (electronic fascicle), Third series, fascicle 14, 1995, Washington, DC. Armed Forces Institute of Pathology den alınmıştır.

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Hodgkin lenfoma (HL) varyantı için kombinasyon kemoterapisi (örn ABVD) kullanımı önerilmektedir. Bu hastalarda tam remisyona ulaşan hastaların ilerlemeye kadar gözlenir. Bir tam remisyona ulaşamayan hastalarda eğer uygunsa, ilerleyen süreçte komplet remisyona ulaşan hastalarda miyeloablative olmayan allojenik HKHT'yi yapma planı ile refrakter HL için kullanılan seri rejimler kullanılabilir.

RT için tedavi sonuçları hakkındaki sınırlı veriler göz önüne alındığında, RT'li hastaların uygun şekilde tasarlanmış bir klinik çalışmaya katılmaları sağlanabilir.

DBBHL histolojik alt tipi RT olan hastalarda hedefe yönelik tedaviler inceleme aşamasındadır.

İbrutinib, pembrolizumab, nivolumab, venetoclax gibi ajanlar bazı çalışmalarda az sayıda hastada kullanılmış ve değişken yanıtlar elde edilmiştir. İlk çalışmalar, hedefe yönelik tedavilere cevap vermiş olsa da, bu ajanları RT hastalarında kullanmanın en iyi yolunu belirlemek için daha fazla çalışmaya ihtiyaç vardır.

Anahtar Kelimeler: Richter transformasyonu, Kronik lenfositik lösemi, Diffüz büyük B hücreli lenfoma,

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