Bölüm 7

PRİMER MERKEZİ SİNİR SİSTEMİ LENFOMALARI

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GİRİŞ

Primer merkezi sinir sistemi lenfomaları (PMSSL), diffüz büyük B-hücreli lenfomaların (YBBHL) sistemik bulgusu olmayan nadir bir alt grubu olup, santral sinir sistemi (SSS) (beyin, gözler, leptomeninksler ve omurilik) tutulumlu oldukça agresif bir ekstranodal non-Hodgkin lenfomanın (NHL) varyantıdır (1-3).

PMSSL yetişkinlerde primer SSS tümörlerinin %2-4'ünden, NHL'lerin ise yaklaşık %1'inden sorumludur (4-6). PMSSL, immünokompetan yaşlı hastalarda artan görülme sıklığıyla birlikte immünsüprese popülasyonlarda immünokompetan popülasyonlara göre daha yüksek pravalansa sahip olup, hastalığın heterojenliği nedeniyle farklı terapötik stratejiler ve değişen klinik sonuçlar söz konusudur (7-9).

Yüksek doz metotreksat (HD-MTX) bazlı polikemoterapi temel birinci basamak tedavi olmakla birlikte, tam yanıt başarı oranlarının düşük olması, seçilmiş hastalarda düşük doz tam beyin radyoterapisi (dr-TBRT) veya yüksek doz kemoterapi (HDC-ASCT) ve otolog kök hücre transplantasyonu ile konsolidasyon tedavisini gerektirmektedir (10).

Terapötik yaklaşımlardaki ilerlemeler özellikle genç ve performans statüsü iyi olan hastalarda uzun süreli remisyon oranlarında iyileşme sağlamış olmakla birlikte, tedavi sonuçları PMSSL için tatminkâr düzeylerde olmayıp (30-60 aylık medyan genel sağkalım süresi ve %30'luk 5 yıllık sağkalım oranı), prognoz SSS-dışı lenfomalara göre de daha düşüktür (11-18).

Dolayısıyla PMSSL nöroonkoloji pratiğinde yönetimi zorlu hastalıklar arasında kabul edilmekte ve yanı sıra kanıta dayalı standart bir tedavi yaklaşımının olmaması bazında tedavi önerileri retrospektif olgu serileri ve sınırlı sayıda büyük ölçekli prospektif çalışmalara dayanmaktadır (2,3).

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- Yaşlı popülasyonunda ve HIV pozitif hastaların sağ kalmalarındaki artışa rağmen, immünokompetan yaşlı popülasyonda görülme sıklığını artıran potansiyel nedenlerin yanı sıra HIV-pozitif hastalarda agresif tedavi rejimlerinin kullanımı hakkında sınırlı bilgi

SONUÇ

HD-MTX-bazlı polikemoterapi, dr-TBRT ve HDC-ASCT ile konsolidasyon tedavisi ve yeni tedavi yaklaşımları yanıt oranlarında iyileşme sağlamış olmakla birlikte, PMSSL hastalarında tedavi sonuçları yüksek relaps ve düşük uzun-süreli sağkalım oranları bazında henüz tatmin edici düzeyde değildir. PMSSL nöroonkoloji pratiğinde kanıta-dayalı standart bir tedavi yaklaşımının da olmamasından hareketle yönetimi zorlu hastalıklar arasında kabul edilmektedir. Cerrahi tedavi, radyasyon ve intratekal tedavinin rolüne dair mevcut görüş ayrılıkları temelinde minimal erken mortalite ve sınırlı uzun-süreli nörotoksisite ile ilişkili optimal bir tedavi rejimine yönelik bir uzlaşının olmaması temel zorluk olarak görünmektedir. Dolayısıyla, klinisyenlere indüksiyon kemoterapisi ve konsolidasyon stratejilerinin optimal doz veya kombinasyonlarına yönelik daha fazla kanıt sağlamak için prospektif randomize klinik çalışmalara gereksinim olup, PMSSL'nin moleküler özelliklerinin yanı sıra nöro-görüntüleme ve biyobelirteçlerin daha iyi anlaşılması, mevcut tedavi seçeneklerini genişleterek, PMSSL'li hastalarda hedef ajanlarla bireyselleştirilmiş tedavilerin uygulanmasını ve böylelikle uzun-süreli sağkalım oranında artışı sağlayabilir.

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