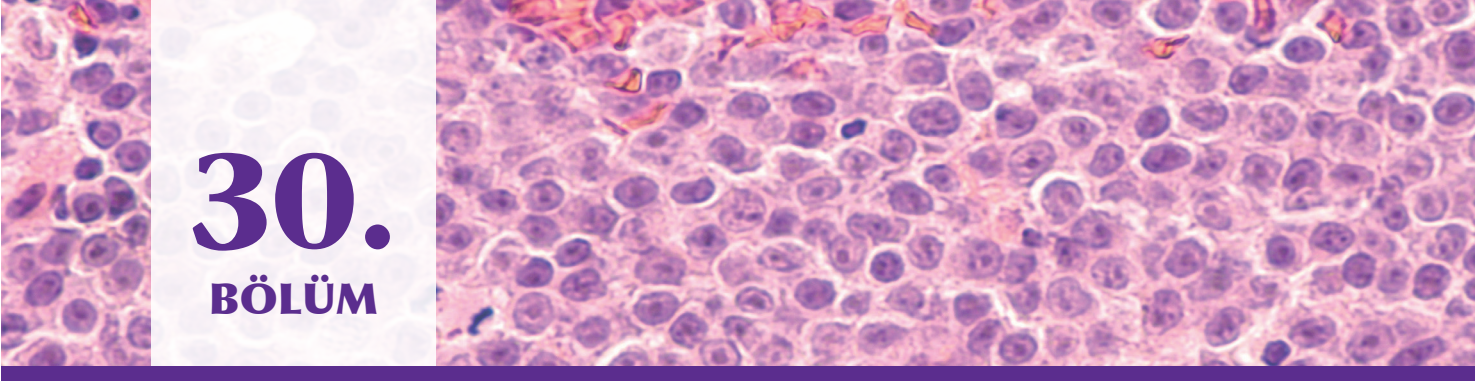

30.
BÖLÜM

İMMÜN YETMEZLİK ZEMİNİNDE OLUŞAN LENFOPROLİFERATİF BOZUKLUKLAR

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GİRİŞ

Konjenital, kazanılmış veya iatrojenik nedenlere bağlı olarak gelişen immün yetmezlik olgularında normal popülasyona oranla lenfoproliferatif gelişme riski daha sıktır. İmmünyetmezlik ilişkili lenfoproliferatif hastalıklar Dünya Sağlık Örgütü hematopoietik ve lenfoid tümör sınıflamasında ayrı bir başlık olarak yer almaktadır. Altta yatan nedene bağlı olarak klinik ve patolojik özellikleri farklılıklar içerir.

Ancak gelişen lenfomaların çoğu B lenfosit kaynaklı olup agresiv histopatolojik özellikler taşımaktadır. Sıklıkla Epstein-Barr virüs (EBV) ile ilişkilidir. İmmünyetmezlikli hastalarda sitotoksik T lenfositler tarafından EBV'ye karşı yanıtın bozukluğu sonucu, EBV enfekte B hücrelerinin çoğalması kontrol dışı hal almaktadır (1-3).

İmmün yetmezlikli hastalarda; onkojenik ve tümör baskılayıcı genlerdeki kalıtımsal veya sonradan kazanılan genetik değişiklikler, kronik antijenik uyarım, onkojenik virüs enfeksiyonları, lenfoproliferatif hastalığa yol açabilen diğer faktörlerdendir (4). Ekstranodal bölge tutulumu diğer lenfomalara oranla progresyon daha hızlı ve tedaviye yanıtları kötüdür.

DSÖ tarafından immünyetmezlik ilişkili lenfoproliferatif hastalıklar 4 ana gruba ayrılmıştır (5).

1. Primer immün bozukluklar ile ilişkili lenfoproliferatif hastalıklar
2. İnsan İmmün Yetmezlik Virüsü (Human immunodeficiency virüs, HİV) Enfeksiyonu ile ilişkili Lenfomalar
3. Nakil sonrası lenfoproliferatif bozukluklar (NSLB)
4. Diğer İatrojenik immün yetmezlik ile ilişkili lenfoproliferatif bozukluklar

1. PRİMER İMMÜN BOZUKLUKLAR İLE İLİŞKİLİ LENFOPROLİFERATİF HASTALIKLAR

Primer immün bozukluk (PİB) ile ilişkili lenfoproliferatif hastalıklar primer immün yetmezliğe veya immün düzenleme bozukluklarına bağlı immün yetmezlik koşullarında ortaya çıkar. 60'tan fazla PİB'in patoloji ve patogenezi heterojen olduğu için bu lenfoproliferatif hastalıklarda tablo oldukça değişkendir. Lenfoproliferatif hastalıklara en sık ilişkiye sahip PİB'ler; Ataksi-Telenjiektazi (AT), Wiskot-Aldrich Sendromu (WAS), Yaygın Değişken İmmün Yetmezlik (YDİY), Şiddetli Kombine İmmün Yetmezlik

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EBV pozitifliği ive DBBHL-dışı lenforproliferatif hastalık ile ilişkili iken >70 hasta yaşı ve DBBHL tipi daha kısa sağ kalım göstergesidir (73).

SONUÇ

Sonuç olarak immün yetmezlik durumu klinik olarak bilinen veye şüphe edilen olgularda ayrıntılı klinik anamnez ile birlikte patolojik değerlendirilmelerinin birlikte yapılması doğru tanı açısından oldukça yardımcı ve gereklidir. İlave edilmesi veya ilave edilmemesi gereken kemoterapotik ajanlar olguların takip ve tedavisinde sağ kalım durumunda büyük rol sahibidirler.

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