

BÖLÜM 16

DİĞER KUTANÖZ T HÜCRELİ LENFOMALAR

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GİRİŞ

Mikozis fungoides (MF), Sezary sendromu (SS) ve primer kutanöz CD30+ lenfoproliferatif hastalıklar dışında kalan diğer kutanöz T-hücreli lenfomalar (KTHL) oldukça nadir görülmekte olup bu grupta yer alan hastalıklar ile ilgili bilgiler günümüzde halen sınırlıdır. Hem klinik ve hem de histopatolojik olarak ayırımalarının zor olması nedeniyle kesin tanı için ayrıntılı immunhistokimyasal belirteçlerin yanı sıra moleküler yöntemlerin kullanımına da ihtiyaç duyulmaktadır. Bu bölümde MF, SS ve primer kutanöz CD30+ lenfoproliferatif hastalıklar dışında kalan diğer KTHL'lerden güncel literatür eşliğinde ayrıntılı olarak bahsedilmiştir.

YETİŞKİN T HÜCRELİ LÖSEMI/LENFOMA

Yetişkin T hücreli lösemi/lenfoma (Adult T-cell leukemia lymphoma; ATLL), etyolojik olarak insan T hücreli lösemi virüsü 1 (HTLV-1) ile ilişkili agresif seyirli, matür bir T hücre malignitesidir. ATLL, HTLV-1 taşıyıcılarının yaklaşık %5'inde ortaya çıkar. Bu nedenle Güneybatı Japonya, Karayip adaları, Güney Amerika ve Orta Afrika'nın bazı bölgeleri gibi HTLV-1 prevalansının yüksek olduğu popülasyonlarda endemiktir (Wolf 2022, Willemze 2005, Hleihel 2021, Cook 2019). Hastalar genellikle yaşamın beşinci veya altıncı dekadındaki erkeklerdir (Malpica 2021, Cerroni 2020, Miyashura 2019). ATLL klasik olarak dört alt tipe

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