

BÖLÜM 15

PRİMER KUTANÖZ CD 30+ LENFOPROLİFERATİF HASTALIKLAR

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GİRİŞ

Primer kutanöz CD 30+ lenfoproliferatif hastalıkların ayırt edici özelliği, lezyondan alınan deri biyopsisinde CD30 eksprese eden atipik T lenfositlerin varlığıdır. WHO/EORTC 2018 sınıflandırmasında primer kutanöz CD30+ lenfoproliferatif hastalıklar; kutanöz T hücreli lenfomalar grubuna dahildir. Bu lenfoma grubu, lenfomatoid papülozis, primer kutanöz anaplastik büyük hücreli lenfoma, transforme mikozis fungoides (MF) ve borderline lezyonları içerir (Willemze vd.2019). CD30 pozitifliği sadece lenfoproliferatif hastalıklara özgü değildir. CD 30 boyanması böcek ısırıkları, ilaç reaksiyonları, enfeksiyonlar ve neoplazi gibi birçok hastalıkta görülebildiği için lenfoproliferatif hastalıkların tanısında klinikopatolojik korelasyon önemlidir (Chen vd.2019).

LENFOMATOİD PAPÜLOZİS

İlk kez 1968'de Macaulay tarafından tanımlanan LeP, kendi kendine iyileşen papülodüler veya papülonekrotik deri lezyonları ile karakterize ve atipik T hücrelerinin CD30+ lenfoid proliferasyonu histolojik özelliklerine sahip kronik, tekrarlayan bir hastalıktır (Macaulay, 1968, Martinez-Cabrales, 2020). Herhangi bir yaşta ortaya çıkabilir, en sık 5. dekatta görülür, ırk ayrımı yoktur, yaş ve cinsiyete göre bimodal dağılım mevcut, pediatrik popülasyonda erkek, erişkinlerde ise kadın cinsiyette daha fazla görülür. Yıllık hastalık insidansı milyon kişi başına 1.2-1.9 olarak bildirilmektedir (Wieser vd.,2016, Bekkenk vd., 2000).

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