Bölüm 9

HİSTİOSİTOZLAR

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GİRİŞ

Histiositler langerhans hücreleri (LH), monosit-maktofaj sistemi, dermal ve interstisyel dendritleri içeren dokularda yerleşik beyaz kan hücrelerini kapsar (1). Bu hücrelerin etkilenen dokulardaki infiltrasyon ve birikimini tanımlayan proliferatif bozukluklar histiositozlar olarak adlandırılmaktadır. Histiositozlar pek çok farklı organda tutulum oluşturabilmektedir. Önceleri eosinofilik granulom, Histiositoz-X olarak adlandırılan hastalığa 1970'te Nezelof tarafından langerin (CD207) ile ilişkili intrasitoplazmik Birbeck granüllerininin saptanması ile langerhans hücreli histiositoz (LCH) adı verilmiştir (2).

Dünya Sağlık Örgütü (WHO) hematopoietik ve lenfoid tümörler klasifikasyonuna göre bu grup hastalıklar 3'e ayrılır (3):

1. Dendritik Hücre Bozuklukları: Langerhans hücreli histiositoz (LCH), juvenil ksantogranuloma (JXG), dendritik fenotipli soliter histiositoma ve Erdheim-Chester hastalığı (ECD) bu gruba dahildir.
2. Makrofaj İlişkili Bozukluklar: Primer ve sekonder hemofagositik sendromlar, makrofaj fenotipli soliter histiositoma ve Rosai-Dorfman hastalığı (RDD) bu gruba dahil edilmiştir.
3. Malign Histiositik Bozukluklar: Bu grupta monosit ilişkili lösemiler (akut monositik lösemi, akut myelomonositik lösemi), ekstrameduller monositik tümör ve dendrit veya makrofaj ilişkili histiositik sarkom yer alır.

Histiocyte Society 2016'da histoistozları 5 ana başlıkta sınıflandırmıştır (4).

LANGERHANS HÜCRELİ HİSTİOSİTOZ

Langerhans hücreleri (LH) deri ve mukozanın özelleşmiş dendritik hücreleridir. LCH'da dokuları infiltre eden hücreler morfolojik ve immunfenotipik olarak LH ile benzerlik gösterdiğinden hastalığa LCH adı verilmiştir. LCH tüm organlarda infiltrasyon oluşturabilen, daha sık olarak tek veya çok sayıda litik kemik lezyonları ile karakterize bir dendritik hücre hastalığıdır. Tam olarak insidansı

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Sıklıkla hastalık kendini sınırlar; fakat atak-remisyon dönemleri yıllar sürebilir. Klinik olarak hastalar 1-sadece LAP ve spontan remisyon ile giden hastalar, 2-yaygın nodal hastalık ve immunolojik anomaliler ile giden hastalar (67) 3-yaygın nodal ve ekstranodal hastalık, uzamış klinik ve relaps-remisyon ile giden hastalar olarak sınıflandırılabilirler (68).

Kutanöz RDD ise sistemik RDD farklı bir hastalık olup deriye sınırlıdır; benign gidişli hastalıkta kozmetik ya da relaps olgularda tedavi ihtiyacı olabilir (69).

Tedavi:

Hastaların çoğunda tedavi gerekmez. Ekstranodal vital organ tutulumu veya hayatı tehdit eden nodal olan hastalarda tedavi gerekebilir. Obstruksiyona sebep olan hastalarda cerrahi seçenek olabilir (70). Sistemik steroid tedavisi LN boyutlarını küçültse de tedavi sonrası rekürrens gelişebilir. Deri ve LN tutulumu olan bir hastada uzun süreli tedavinin yararı gösterilmiştir. Histiocyte Society vital organ tutulumu olmayan hastalarda tedavi önermemektedir. LN tutumu olan olgularda öncelikle düşük doz prednison tedavisini, vital organ basısı olan vakalarda ise cerrahi ve yüksek doz steroid tedavisini önermektedir. Cerrahi kontrendike olan vakalarda radioterapi denenebilir. Yanıtsız, hayatı tehdit eden ve sık tekrarlayan vakalarda kladribinin efektif olduğunu bildiren yayınlar mevcuttur (70-71) .

JUVENİL KSANTOGRANULOMA (JXG)

Bir başka kendini sınırlayan non langerhans histiositoz olan JXG genellikle infantil dönem-çocuklukta multiple deri lezyonları ile ortaya çıkar. Tutulum en sık baş, boyun, gövdede küçük, sert makulopapuler döküntüler şeklindedir (72). Derin yumuşak doku ve organ tutulumu olabilmekle birlikte sistemik tutulum nadirdir. Deri tutulumu kendini sınırladığından nadiren tedavi gerektirir. Büyük abdominal kitleler, MSS ve kemik iliği tutulumu olanlarda LCH benzeri tedaviler önerilmektedir. Erişkinde ortaya çıkan hastalık daha komplike olma eğilimi gösterir ve spontan düzelme beklenmez.

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