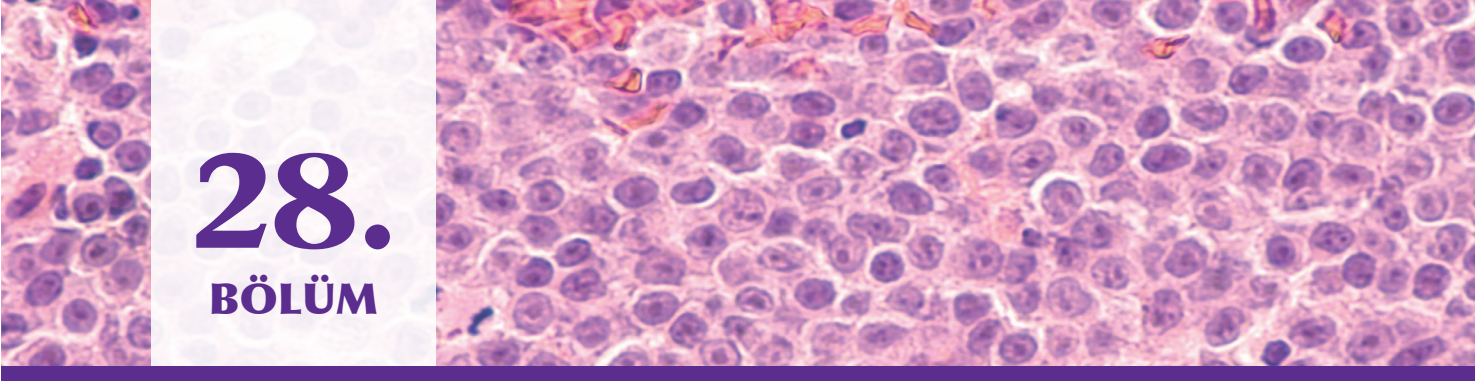

28.
BÖLÜM

# LANGERHANS HÜCRELİ HİSTİYOSİTOZİS

*Gülsüm Şeyma YALÇIN* [1]

## LANGERHANS HÜCRELİ LEZYONLAR

Langerhans hücreleri (LH) deri ve mukozal bölgelerde bulunan özelleşmiş dendritik hücrelerdir (1). İlk olarak 1868 yılında Paul Langerhans Jr. tarafından tanımlanmıştır (2, 3). Aktive oldukları zaman T lenfositlere antijen sunma görevi görür ve lenfatikler aracılığı ile lenf nodlarına göç ederler (1).

Güncel Dünya Sağlık Örgütü sınıflandırmasında Langerhans hücrelerinden köken alan tümörler, sitolojik atipi derecesine ve klinik davranış özelliklerine göre, genel olarak iki ana başlık altında ele alınmaktadır: Langerhans Hücreli Histiyositozis (LHH) ve Langerhans Hücreli Sarkoma (LHS). Bu tümörler tanım olarak Langerhans-tipi hücrelerin klonal neoplastik proliferasyonundan oluşmaktadır. Vücuttaki normal Langerhans hücreleri gibi CD1a, langerin ve S100 proteini eksprese etmekte ve normal Langerhans hücreleri gibi yapısal olarak elektron mikroskopik inceleme ile görülebilen Birbeck granülleri içermektedirler. Birbeck granülleri tenis raketi benzeri morfolojide, 200-400 nm uzunlukta, 33 nm genişlikte, fermuar benzeri görünümde mikroyapılardır (4). İmmünohistokimyasal langerin ekspresyonu Birbeck granülleri varlığı ile koreledir (5).

LHH ile ilgili bazı genetik değişikliklerin kemik iliğindeki prekürsör hücrelerde de tespit edilmesi nedeniyle myeloid bir neoplazi olarak sınıflanması önerisinin (6), revize 2016 "Histiocyte Society" klasifikasyonunda bazı otörlerce uygun görüldüğü belirtilmiştir (7).

LHH klinikopatolojik bir tanıdır. -Özellikle lenf nodundaki- normal reaktif LH'ler görülerek yanlışlıkla LHH tanısı konmaması için, ancak uygun klinik tablo olduğunda LHH tanısı verilmelidir. Ayrıca uygun klinik ve radyolojik bulgular olsa da, kesin tanı histopatolojik ve immünohistokimyasal bulgulara dayanarak verilmelidir (5).

## 1- LANGERHANS HÜCRELİ HİSTİYOSİTOZİS

### Tarihçe

1893 yılında Alfred Hand Jr, egzoftalmus, diabetes insipidus, kafa kemiklerinde lezyonlar ve deride skabies benzeri lezyonlar bulunan bir çocuk hasta tanımlamış ve tüberküloza bağlı olabileceğini düşünmüştür (8, 9). 1915 yılında ise Artur Schüller kafa kemiklerinde lezyonlar ve egzoftalmus izlenen 2 hasta tanımlamıştır. Biri diabetes insipidus izlenen bir çocuk hasta iken, diğeri adipozogenital distrofi belirtileri bulunan bir

[1] Dr. Öğr. Üyesi, Afyonkarahisar Sağlık Bilimleri Üniversitesi Tıp Fakültesi Patoloji AD, drgsu@hotmail.com

### Patogenez

Literatürde bildirilen BRAF V600E mutasyonu tespit edilen LHS olgusu mevcuttur (63).

### Prognoz

LHS'de agresif seyir ve progresif bir gidişat izlenmektedir. Mortalite oranı >%50 olarak belirtilmektedir (4, 57).

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