BÖLÜM 46

MASTOSİTOZ

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MAST HÜCRESİ

Mast Hücre Fizyolojisi ve Bulunduğu Dokular

Mast hücreleri; kemik iliği ve dalakta bulunan kök hücrelerinden köken alan ama diğer myeloid hücrelerden farklı olarak neredeyse tamamı matürasyonunu çevre dokularda tamamlayan hücrelerdir (1). Kemik iliğinden köken almalarına rağmen burada çok az miktarda bulunurlar ve periferik kanda mast hücreli neoplaziler dışında görülmeleri ve kültürde üretilmeleri zordur. Dış çevre ile temas eden epitelize organlarda ise (deri ve gastrointestinal sistem mukozası gibi) bağ dokusunda perivasküler alanda oldukça fazla sayıda bulunurlar (2). Ayrıca diğer myeloid hücrelerden farklı olarak matürasyonları çoğunlukla yine bu çevre dokularda tamamlarlar. Hatta bazı hayvan çalışmalarında mast hücrelerinin kemik iliği dışında periferik bağ dokularında çoğalabildiği de gösterilmiştir (3, 4). Yine de insanlarda mast hücrelerin kemik iliği kök hücrelerinden çoğaldığı kabul edilir.

Mast hücrelerinin çoğalması ve gelişimi için esas gerekli olan büyüme faktörü kök hücre faktörü veya diğer ismi ile Stem Cell Factor'dür (SCF). Bu büyüme faktörü mast hücre yüzeyinde bulunan CD117 veya KIT isimli transmembran bir protein reseptöre bağlanarak etkinliğini gösterir ve bu etkileşim sonrası mast hücre içerisinde tirozin kinaz yolağı aktive olur. Bunun sonucunda hücre proliferasyonu ve matürasyonu gerçekleşir (5, 6). Mast hücre sayısı ve fonksiyonunu ilgilendiren birçok patoloji bu reseptöre fonksiyon kazandıran KIT mutasyonları sonucu ortaya çıkar ve birçok mast hücre hastalığı tedavisi için bu gen bir hedef oluşturur (5, 7). Ayrıca inflammatuar süreçlerde oluşan sekonder mast hücre artışının sebebi de bağ dokusundaki fibroblastlardan sentezlenen SCF miktarının artışıdır (8, 9).

Mast hücrelerinin normal fizyolojik şartlarda görevleri vücut savunması ile ilişkilidir. Bu görev-

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moterapisi alana kadar bu semptomlar gerilemeyecektir (49-51).

SONUÇ

Mast hücre bozuklukları birçok farklı hastalığı içeren kronik myeloproliferatif hastalık gruplarından birini oluşturur. Bu hastalıklar benzer patogeneze veya genetik özelliklere sahip olsa da prognoz ve tedavi gereksinimi açısından çok geniş bir yelpazeye sahip oldukları unutulmamalıdır. En hafif cilt mastositozundan, mast hücreli lösemiye kadar her bir hasta kendi içinde değerlendirilmeli, gerekli tetkikler ve incelemeler yapılmalı ve hastaların tedavisi bunların üzerine kurulmalıdır. Nadir görülen bu hastalar için alerji ve hematoloji hekimleri gerektiğinde birlikte çalışmalı, gereksiz tedavilerden kaçınmalı ve uygun gördükleri anda tedavi için hastaları tecrübeli merkezlere yönlendirmekten çekinmemelidir. Bütün bunlara ek olarak mastositoz hastalarındaki yapılan yeni genetik modellemeler ve tedavi seçenekleri ile yakın gelecekte bu hastalara çok daha semptomsuz ve progresyonsuz sağkalımları sağlanması beklenilmektedir.

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