Bölüm 21

MASTOSİTOZ

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GİRİŞ

Mastositöz , neoplastik mast hücrelerinin deri ve/veya birden çok organda birikmesi ile karakterize heterojen , nadir bir hastalık grubudur. Sistemik mastositöz (SM) da neoplastik mast hücreleri fokal ve/veya diffüz olarak kemik iliği , dalak, karaciğer ve gastrointestinal sistemde tutabilir. SM'da hemen hemen bütün hastalarda kemik iliği tutulumu mevcut. Cilt tutulumu genelde indolent mastositöz (ISM) , daha az olarak SM'da ve nadiren mast hücreli lösemi de görülür.

Mastositöz ilk kez 1869' da Nettleship ve Tay tarafından bir deri hastalığı olarak tarif edildi. 1949 ' da ilk SM vakası bildirildi ve gelecek yıllarda farklı semptomlar, özellikleri tanımlanarak alt grupları belirlendi. SM ' da prognoz çok değişkendir. 2001 yılında ilk kez Dünya Sağlık Örgütü (DSÖ) tarafından mastositoz temel sınıflaması yapıldı, 2016'da yeni prognostik parametreler ve etkili tedavi seçenekleri ile sınıflaması güncellendi(Tablo-1)

KUTANÖZ MASTOSİTOZ

2016 ' da kutaneoz mastositoz makulapapuler KM (ürtikeria pigemnetoza) (Shawrtz & arkadaşları, 2016), diffüz KM(Akın & arkadaşları, 2002), derinin mastositoması olarak tanımlandı. Serum triptaz düzeyi genellikle 20 ng/mL 'den düşüktür. KM genelde çocukluk yaşlarında tanı alınır ve iyi prognozludur, çoğu hastalarda puberte sonrası deri lezyonları kaybolur, o nedenle KM ' ye bağlı cilt tutulumların diğer cilt hastalıklarından ayırımı önemlidir(Caplan & arkadaşları, 1963).

Erişkinlerde ise hastalık persiste eder ve sistemik mastositoza dönüşür. Bazı kutanöz mastositozlu olgularda deri lezyonları yaygındır ve tedavi gerektirir. Tedavide antihistaminikler, cilt bakımı ve psoralen-UV A (PUVA) uygulanabilir. Ağır olgularda topikal kortikosteroidler kullanılabilir.

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2-İndüksiyon terapi

Akselere veya blastik faz KNL' de standart indüksiyon tedavisi ile(antrasiklin ve sitarabin) hiçbir hematolojik tam remisyon bildirilmemiştir, yalnızca akselere fazda olan bir hastada ikinci bir kronik faz elde ederek başırılı olunmuştur, lösemik olan hastalar indüksiyon tedavisi sonrası ya ölmüş veya hipoplastik kemik iliği saptanmıştır(Willard & arkadaşları 2001, Kobayashi & arkadaşları 2002)

3-Hematopoetik kök hücre nakli

Akselere veya blastik dönüşüm potansiyeli göz önüne alındığında allojenik kök hücre nakli tek tedavi yöntemidir, buna rağmen literatürde nispeten az sayıda ve tek vaka şeklinde raporlar mevcuttur, yaşları 15, 23, 26, 40 (n=29, 49(n=2) ve 60(n=2) ' olan dokuz vaka raporlanmış , nakil 6 hastada başarılı olmuş ve sırasyıla 78,73,54,36,7,1 aydan fazla remisyon saptanmıştır(Elliot & arkadaşları 2005,Bohm & arkadaşları 2002,Lee & arkadaşları 2015,Hasle & arkadaşları 1996,Goto & arkadaşları2009). Şuan için KNL için kordon kanı veya haploidentikal nakil hakkında bilgi yoktur.

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