BÖLÜM 40

BLASTİK PLAZMASİTOİD DENDRİTİK HÜCRELİ NEOPLAZİ

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GİRİŞ-TARİHÇE

Blastik Plazmasitoid Dendtritik Neoplazi (BPDHN); çok agresif klinik gidişe sahip, cilt tutulumu, kemik iliği tutulumu ve lösemik formda prezente olabilen, nadir görülen bir hematolojik malignitedir (1). Dünya Sağlık Örgütü (WHO) 2016 sınıflamasında (2) ve 2022 yılında yayınlanan WHO Hematopoetik Neoplaziler 5.Güncellemesinde; Myeloid/Dendritik ve Histiyositik Neoplaziler bölümünde ayrı bir başlıkta ele alınmaktadır (3).

İlk olarak 1995 yılında; CD4 pozitif agranüler NK hücreli lösemi olarak tanımlanmıştır (4). 1998 yılında, kutanöz agranülar CD2/CD4+/CD56+ lenfoma terminolojisi kullanılmıştır (5). Morfolojik olarak blastik ve agranüler görünümde olması ve NK hücre markerı olan CD56(+) olması nedeni ile sonrasında blastik NK hücreli lenfoma olarak adlandırılmıştır. 2002 yılında; agranüler CD4+CD56+ hematodermik neoplazm/tümor olarak isimlendirilmiştir (6). 2003 yılında erken plasmasitoid dendritik hücreli lösemi/lenfoma(7) tanımı sonrasında, WHO 2008 güncellemesinde, AML sınıflaması altında blastik plazmasitoid dendritik neoplazi olarak yer almıştır (8).

Epidemiyoloji

Oldukça nadir görülen bir hematolojik neoplazi olması, uzun yıllar terminolojisinin net olmaması nedeni ile tam insidansı bilinmemektedir. Tüm cilt lenfomalarının %0.4'ünü oluşturduğu ifade edilmiş olsa da, cilt tutulumu olmadan da prezente olabilmesi nedeni ile insidansı net bilinmemektedir (9), (10). Tüm yaş gruplarında görülebilmekle birlikte; 6-7. dekattaki erişkinlerde daha sık izlenir ve medyan tanı yaşı 65-67'dir. Erkeklerde kadınlara göre daha fazla görülmektedir (2.5:1) (9). %80-90 vakada izole olarak ortaya çıksa da, hastaların %10-20'sinde bir diğer hematolojik malignitenin (MDS, AML, KMML gibi) seyrinde de görülebilir (10). Dikkat edilmesi gereken bir diğer nokta ise; matür, CD56 negatif

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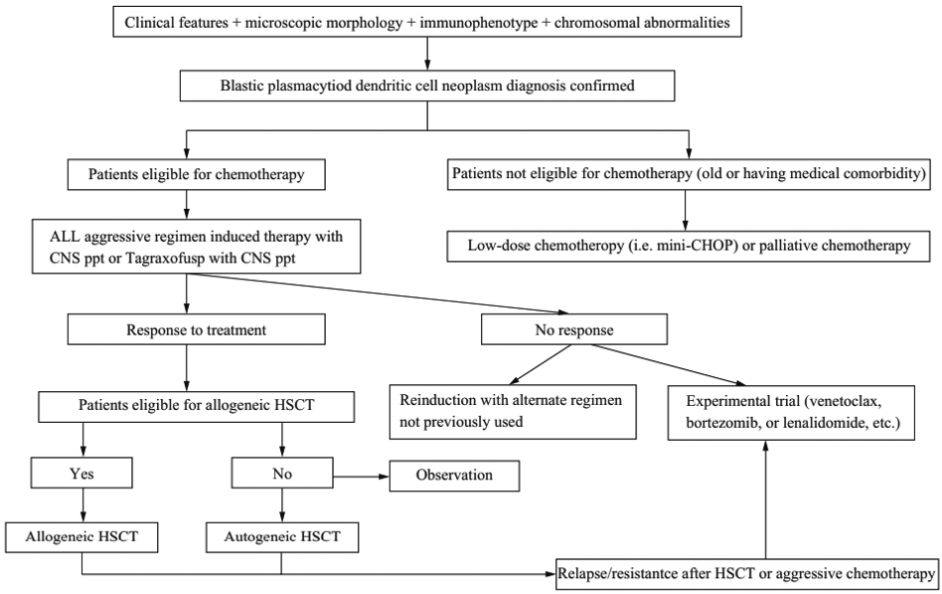
BPDHN'de etkinliğini gösteren bir kaç vaka takdimi bulunmaktadır (46). Halen venetoclax'ın BPDHN'de etkinliği araştıran klinik çalışmalar (NCT03485547), venetoclax + tagraxofusp kombinasyon çalışmaları (NCT04216524) ve yine alltta yatan MDS/KMML tanısı olan hastalarda Tagraxofusp + Venetoclax + Azasitidine çalışması da devam etmektedir (NCT04216524).

CD123 hedefleyen CART hücre tedavisi olan MB-102'nin AML, MDS, BPDHN'de çalışmaları (NCT04109482); CD123 ve CD3'e bağlanan bir BiTE olarak geliştirilen XmAb14045'in çalışmaları yine devam eden çalışmalar arasındadır (NCT02730312). Mevcut onaylı tedaviler doğrultusunda, BPDHN tedavi algoritması Şekil 1 ve Şekil 2'de gösterilmiştir.



Şekil 2. Tedavi Algoritması, Cheng et al (48)

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