Bölüm 3

GÜNCEL EDİNSEL APLASTİK ANEMİ PATOGENEZ, KLİNİK BULGULAR, TEŞHİS VE TEDAVİ YAKLAŞIMI-2020

Hikmettullah BATGİ[1]

GİRİŞ

Edinsel ağır aplastik anemi (AA), önemli morbidite ve mortalite ile ilişkili nadir bir hematolojik hastalıktır. Tanısı, pansitopeniye neden olabilecek diğer bozuklukların dışlanması ve iyi bilinen Camitta kriterlerine dayanır.[1] Ağır AA, immün aracılı kemik iliği hipoplazisi ve pansitopeni ile karakterize olup immünsüpresif tedaviyle (İST) etkili bir şekilde tedavi edilebilir.[2] Esas olarak antitimosit globulin (ATG) ve siklosporin veya allojenik kök hücre transplantasyonu ile tedavi edilir.[3] Hastaların üçte birinde kalıcı şiddetli sitopeni ve transfüzyon gereksinimi vardır ve bunlar İST'ye dirençlidir.[2]

TANIMLAR

AA, kemik iliği hipoplazisi ve aplazisiyle ilişkili pansitopeni anlamına gelir ve altta yatan çeşitli nedenleri vardır (Tablo 1).[4] AA teşhisi için kriterler aşağıda açıklanmaktadır.

Ağır AA teşhisi aşağıdaki kriterlerin her ikisini de gerektirir[5];

- Kemik iliği hücreselliği <% 25 veya % 25-50 olduğunda rezidüel hematopoetik hücrelerin oranı <%30 olmalı ve
- şağıdaki 3 durumdan en az ikisi bulunmalıdır.
  - Periferik kan mutlak nötrofil sayısı (ANC) <500 / microL (<0,5 X 109 / L)
  - Periferik kan trombosit sayısı <20.000 / microL
  - Periferik kan retikülosit sayısı <20.000 / microL

Çok ağır AA[6] tanısı için kriterler;

- Ağır AA ve ANC <200 / microL'dir.
- Ağır olmayan AA için kriterler şunlardır;
- Hiposellüler kemik iliği (Ağır AA için açıklandığı gibi)

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Tablo 1: Edinsel aplastik aneminin başlıca nedenleri	
İdiyopatik	**Viral enfeksiyonlar**
Sitotoksik ilçalar ve radyasyon	Epstein Barr Virüsü
Kanser tedavisi	Seronegatif (A ila -G olmayan) hepatit
İlaç reaksiyonu	İnsan immün yetmezlik virüsü (HIV)
Antiepileptikler:Karbamazepin, fenitoin,diğerleri	Diğer herpes virüsleri
Antibiyotikler: sülfonamidler, kloramfenikol	**İmmün bozukluklar**
Nonsteroidal antiinflamatuarlar: fenilbutazon, indometazin	Eozinofilik fasiit
Antitroidal ialçlar: metimazol,propiltyouracil	Sistemik lupus eritematoz
Altın	Greft-versus-host hastalığı
Arsenik	Paroksismal noktürnal hemoglobinüri
Toksik kimyasallar	**Diğer nedenler**
Benzen	Timoma
Çözücüler	Gebelik
Tutkal buharlar	Anoreksiya nervoza

Tablo 2: Kemik iliği yetmezliğinin başlıca nedenleri	
Kalıtsal kemik iliği yetmezliği sendromları	**Edinsel kemik iliği yetmezliğinin nedenleri**
Fanconi anemisi	İlaçlar, kimyasallar veya radyasyonun neden olduğu aplastik anemi
Diskeratoz konjenita	Viral enfeksiyon veya immün bozukluk ile ilişkili edinsel aplastik anemi
Shwachman-Diamond sendromu	Miyelodisplastik sendromlar
Amegakaryositik trombositopeni	Paroksismal noktürnal hemoglobinüri
Retiküler disgenez	Büyük granüler lenfositik lösemi
Diğer	

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