Bölüm 12

WALDENSTRÖM MAKROGLOBULİNEMİSİ GÜNCEL TANI VE TEDAVİ YAKLAŞIMI

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GİRİŞ

Waldenström makroglobunemisi (WM) serum monokonal IgM artışı ve kemik iliğinde lenfoplazmasitik hücre infiltrasyonu ile karekterize lenfoproliferatif bir hastalıktır.[1]

WM' nin ana fiziksel bulguları hepatomegali (20%), splenomegali (15%) ve lenfadenopatidir (15%).[2] En sık görülen semptom normositik anemiye bağlı yorgunluktur. Tanı esnasındaki ortalama hemoglobin 10 g/dl' dir.[3] WM tanı esnasında genellikle asemptomatiktir.[4]

Tanı anında ortanca yaş zencilerde 63, beyazlarda 73'tür. Siyahlar beyazlara göre daha kısa yaşam süresine sahiptir.[5] Erkeklerde görülme sıklığı (0.92/100.000) iken kadınlarda bu oran (0.30/100.000)' dur.[6] 70 yaş altı hastalarda ortanca sağ kalım 10 yıl, 70-79 arası 7 yıl, 80 yaş ve üzerinde ise 4 yıldır.[7] En önemli ölüm nedeni hastalığın ilerlemesi, tedavi komplikasyonları ve yüksek dereceli lenfomaya dönüşümdür.[8]

TANI

WM monoklonal IgM artışı ile seyreden bir lenfoplazmasitik lenfomadır.[1] Kemik iliği ve lenf nodları maturasyonunun farklı evrelerinde olan pleomorik B hücreleri ile infiltredir.[9] Kemik iliği incelemesinde intertrabeküler alanda tutulum vardır.[1] Klonal WM hücrelerinin immünfenotipik özellikleri; pan B yüzey belirteçleri (CD19, CD20, CD22), yüzey/ sitoplazmik IgM, FMC7 ve CD79a pozitif iken CD5, CD10, CD11c, CD23 ve CD 103 negatiftir. [10] Kötü prognozla ilişkili del 6 q anomalisi olguların %42' sinde mevcuttur.[11] 11q delesyonu ve trizomi 4'de kötü prognazla ilişkilidir.[12] MYD88 L265P geni olguların % 90 'nında saptanmaktadır. MYD88 geninin tipi prognozla ilişkilidir. MYD88MUT da 10 yıllık yaşam beklentisi %90 iken, MYD88WT de %73'tür.[13] MYD88 çevresel kandan PCR ile saptanabilir.[14]Hastaların % 30'unda kötü prognozla ilişkili CXCR4 mu-

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Yeni tanı Waldenström Makroglobunemisi yönetimi		
IgM MGUS Asemptomatik makroglobunemi Hb ≥11 g/dL Plt ≥ 120.000 /L ▼▼ İZLEM	Hgb <11 g/dL Plt <120.000 IgM ilişkili nötropeni WM ilişliki hemolitik anemi Semptomatik kriyoglobunemi ▼▼ TEK AJAN RITUXIMAB	Bulky hastalık Sitopeni -hb<11 -plt<120.000 Konstisyonel semptomlar Hiperviskozite semptomları ▼▼ Hipervizkosite sendrom var yok ▼▼ ▼▼ Plazmaferez R-B x 4/6 . kür R-B x 4/6 kür

Şekil-1 Mayo klinik çalışma grubunun yeni tanı WM yönetimi. Kısaltmalar; hb hemoglobin; plt trombosit, IgM immünglobulin M; MGUS anlamı belirlenemeyen monoklonal gamopati; RB rituximab, bendasmustine.

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