Bölüm
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POEMS TANI VE TEDAVİSİ

Aysun GÖNDEREN[1]

GİRİŞ

POEMS sendromu; polinöropati, organomegali, endokrinopati, monoklonal protein, deri değişiklikleri ile karakterize bir sendromdur. Vakaların tümünde gözlenen monoklonal gamopati ve periferik nöropati dışında osteosklerotik kemik lezyonları, Castleman hastalığı (anjiyofoliküler lenf nodu) artmış serum vasküler endotel büyüme faktörü (VEGF) seviyeleri, organomegali, endokrinopati, ödem, tipik cilt değişiklikleri, plevral efüzyon, asitler, eritrositoz veya trombositoz, papilödem gibi özelliklerin bir veya daha fazlasıyla karakterize edilir [1]. POEMS sendromunun daha az sıklıkla kullanılan diğer isimleri osteosklerotik miyelom, Takatsuki sendromu veya Crow-Fukase sendromudur [2,3].

PATOFİZYOLOJİ

VEGF, endotel hücrelerini hedef alması, vasküler geçirgenlikte hızlı ve geri dönüşlü bir artışa neden olması ve anjiyogenezdeki önemli rolü nedeniyle hastalıkla en iyi şekilde ilişkilendirilmiş olan sitokindir [4,5].

POEMS sendromunda mikroanjiyopati, ödem, efüzyon ve artmış vasküler geçirgenlik, neovaskülarizasyon, polinöropati, pulmoner hipertansiyon, lökositoz ve trombositozun da etkisi ile VEGF gibi proinflamatuar sitokinlerin aşırı üretilmesi patofizyolojiden sorumlu tutulmuştur [6,7]. POEMS tanılı hastalar multipl miyelomlu hastalardan daha yüksek düzeyde interlökin-1 beta (IL-1B), tümör nekroz faktörü alfa (TNF-alfa) ve interlökin-6 (IL-6) 'ya sahiptir [6]. Hem IL-1B' nin hem de IL-6'nın VEGF üretimini uyardığı gösterilmiştir [8]. Trombositlerin [9] veya plazma hücrelerinin [10,11], vasküler geçirgenliği arttıran güçlü bir in-

[1] Dr Öğrt Üyesi, Aysun GÖNDEREN, Kütahya Sağlık Bilimleri Üniversitesi Evliya Çelebi EAH, dr.aysunbaz@gmail.com

Tablo 2: POEMS sendromunda kullanılan farklı tedaviler		
Rejim	**Koşullar**	**Sonuç**
Radyoterapi	1-3 kemik lezyonu olan hastalar (kemik iliği tutulumu olmaması)	>%50 yanıt
Melfalan-Dexametazon	ASCT için uygun olmayan hasta	%100 VEGF ve nörolojik yanıt
Otolog kök hücre nakli(ASCT)	ASCT için uygun ve kök hücre toplanabilmiş hasta	%100 yanıt
Talidomid-Dexametazon	Daha önce arteryal olay olmaması	VEGF yanıtı ancak toksik nöropati riski
Lenalidomid-Dexametazon	Daha önce arteryal olay olmaması	Nörolojik ve VEGF yanıtı açısından RT veya ASCT' den önce etkili görünmekte
Bortezomib içeren rejimler	Genellikle ikinci basamakta	VEGF yanıtı % 88, Nörolojik iyileşme % 95

Anti-VEGF antikorları: Bazı vaka raporları, anticytokine /anti-VEGF aktivitesi olan ajanların (örneğin, bevacizumab) kullanılmasının bu hastalığın belirti ve semptomlarının bir kısmının veya tamamının iyileştirilmesinde yardımcı olabileceğini düşündürmekteydi [55,56]. VEGF'nin POEMS sendromunda rolü göz önüne alındığında bevacizumab mantıklı görünmekteydi ancak bu tedavinin bazı vaka serilerinde artmış mortaliteye neden olduğu gösterilmiştir. Artmış mortaliteyi açıklayabilen bir hipotez, yüksek VEGF seviyelerine uzun süre maruz kaldıktan sonra hipertrofik endotelyal hücrelerde VEGF özelliğinin neden olduğu apoptozdur. Bu masif apoptozis, daha sonra, vasküler kaçak sendromunu indükleyen neovaskülarizasyonun yıkımını tetikler [5,51].

POEMS sendromunda kullanılan farklı tedavi yöntemleri tablo 2' de gösterilmiştir [52,57,58,59,60,61].

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