BÖLÜM 12

SEREBRAL VASKÜLİT-VASKÜLOPATİLER

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GİRİŞ

Santral sinir sistemi (SSS) vaskülitleri; nedenine, yerleşim yerine ve etkilenen damarın boyutuna veya nöropatolojik bulgulara göre sınıflandırılabilir. Uluslararası Chapel Hill Konsensüs Konferansı'nda belirlenen ve 2012 yılında revize edilen sınıflandırma, sistemik vaskülitlerin adlandırılmasında en yaygın kullanılan sınıflandırmadır (1). Damar boyutuna göre sınıflandırıldığında; büyük damar vaskülitleri aortu ve ana dallarını, vertebral arteri, baziler arteri, internal karotid arteri (İCA), external karotid arteri (ECA), anterior serebral arterin (ACA) A1 segmentini, orta serebral arterin (MCA) M1 segmentini ve posterior serebral arterin (PCA) P1 segmentini etkiler. Orta damar vasküliti, MCA'nın bifurkasyon düzeyi distalindeki dallarının yanı sıra anterior ve posterior kominikan arterleri etkiler. Küçük damar vasküliti ise arteriyolleri, venülleri ve kılcal damarları etkiler (2).

SSS vasküliti beyin, spinal kord ve menenkslerdeki vasküler yapıların inflamasyonu ve destrüksiyonu ile karakterize geniş bir hastalık grubudur. SSS vaskülitleri primer ve sekonder olarak sınıflandırılabilir. Vaskülit tutulumu SSS ile sınırlı ise primer, diğer sistemlerde de tutuluma neden olan sistemik bir inflamatuar veya enfeksiyöz sürece sekonder ortaya çıkarsa sekonder olarak adlandırılır. Sekonder vaskülite neden olabilecek hastalıklar arasında konnektif bağ doku hastlalıkları, sistemik vaskülitler, kronik inflamatuar hastalıklar ve varisella gibi enfeksiyon hastalıkları yer alır (3).

SANTRAL SİNİR SİSTEMİNİN PRİMER ANJİİTİ

SSS primer anjiti ilk olarak 1959 yılında beyin otopsi incelemeleri sonucunda rapor edildi (4). 1980'lere dek ölümcül bir hastalık olarak kabul edilmekteydi (5). Tanı yöntemlerindeki ilerlemeler, hastalık hakkında artan farkındalık ve artan başarılı tedavi oranlarıyla vaka sayısında artış izlendi (6). SSS primer anjiti nadir görülen bir hastalık olup: yıllık insidansı ortalama milyonda 2.4 vakadır (7). Her yaştan hastayı etkileyebilir, ancak ortalama 50 yaş civarında pik

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(77). Takayasu arteriti, damar duvarlarında inflamasyon ve fibrozise neden olup lümende darlık, oklüzyon, dilatasyon ve anevrizma oluşumuna neden olur. Etkilenen hastalarda kollarda kladikasyo ve nörolojik semptomlarla görülür. Takayasu arteritinin serebrovasküler bulguları geçici iskemik atak, inme ve hipertansif ensefalopatiyi içerir (77,78).

Tanıda erken dönemde yüzeyel USG ile intima-media kalınlık artışı görülür (78). Kontrastsız BT, kalsifikasyonlarla birlikte aort ve dallarındaki kalınlaşmayı ve dansite artışını gösterebilir. Kontrastlı BT ile de damar duvarında kontrastlanma gösterilebilir (79). MRG'de T2 ağırlıklı sekanslarda, inflame vasküler yapının içinde ve çevresinde ince duvar kalınlaşması ve T2 hiperintens sinyal gözlenir. Akut faz sırasında, damar duvarında ve periadventisyal yumuşak dokularda kontrast tutulumu gözlenebilir. Geç fazda ise stenoz bölgeleri ile buna sekonder segmental dilatasyon gözlenebilir (80,81). DSA'da aortun veya en az iki orta büyüklükteki vasküler dalın tutulumu tanı için gereklidir (77) (Resim 8).

Dev Hücreli (Temporal) Arterit

Dev hücreli arterit, büyük arterleri tutan kronik, granülomatöz bir vaskülittir. Çoğunlukla süperfisyal temporal arteri tutar, ancak oksipital arteri de tutabilir ve genellikle 55 yaşın üzerindeki hastalarda görülür. Dev hücreli arteritte iki yaygın klinik bulgu grubu vardır: temporal arterit ve polimiyalji romatika. Dev hücreli arteritin semptomları arasında tek taraflı baş ağrısı, yüz ağrısı, çene kladikasyosu ve görme kaybı bulunur. Dev hücreli arterit tanısı temporal arter biyopsisi ile konur (30,52,78).

USG'de kalınlaşmış bir hipoekojen arter duvarı (halo işareti), temporal arterin dev hücreli arterit tutulumunun karakteristik gri skala özelliğidir. Renkli Doppler USG ile etkilenen damarda türbülan akım ve darlık gösterilebilir (52). Kontrastlı yüksek çözünürlüklü MRG'de duvar kalınlaşması ve duvarda ve komşuluğunda kontrast tutulumu görülür (82). T2 ağırlıklı sekasnlarda damar duvarında ödem saptanır (83).

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