

SANTRAL SINİR SİSTEMİNİN DİĞER İDİYOPATİK İNFLAMATUAR DEMİYELİZAN HASTALIKLARI VE MULTİPL SKLEROZ VARYANTLARI

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GİRİŞ

Santral sinir sisteminin idiyopatik demiyelinizan hastalıkları nedeni bilinmeyen geniş bir grup hastalığı içerir. En sık görülen multipl sklerozdur(MS). Diğerleri, hastalık başlangıcı, klinik görünümü, hastalık seyri, görüntülemede lezyonların lokalizasyonları ve yapısı, patolojik ve labartuvardan özellikleriyle MS'den farklılıklar gösterirler. Ayrıca MS kullanılan tedavilere yanıt vermemele önemlidir. Bu özelliklerin tanımlanması ve bilinmesi, hastalıkların doğru tanı ve erken tedavisi açısından önemlidir. Bu yazıda bu grup içinde tanımlanmış SSS'in diğer demiyelizan hastalıkları ve MS varyantları kısaca özetlenmiştir.

NÖROMİYELİTİS OPTİKA SPEKTRUM BOZUKLUKLARI

Daha önce Devic sendromu olarak bilinen nöromiyelitis optika (NMO), başlıca optik sinirler ve spinal kordu tutan inflamatuar lezyonlar ile karakterize santral sinir sisteminin (SSS) immun aracılı, ciddi inflamatuar ve demiyelinizan hastalığıdır. Son yıllarda, optik sinirlerin (OS) ve spinal kodun (SK) dışında SSS'in farklı bölgelerinin etkilendiği klinik fenotipleri tanımlanmıştır (1,2). Bu bozuklukların patogenezinin anlaşılması, hastalığa özgü aquaporin-4' e (AQP4) bağlayan antikor ile ilişkileri, nörogörüntüleme özelliklerinin belirlenmesi,

önceki NMO sınıflandırmasının yeniden düzenlenmesi ile daha spesifik tanı kriterleri ve hastalık tedavi kılavuzu 2015 yılında yayınlandı ve bu grup NMO spektrum bozukluğu (NMOSD) olarak adlandırıldı (3). Tanı kriterleri, NMOSD'ları AQP4 antikor varlığına göre: seropozitif ve seronegatif veya AQP4 durumu bilinmeyenler olmak üzere ikiye ayırmıştır (4). Seronegatif grup NMOSD'ların yaklaşık %20-25'ini oluşturur. 2012 yılında seronegatif NMOSD'li hastaların bir kısmında miyelin oligodendrosit antikorlarının (MOG) olduğu bulundu. MOG antikoru bulunan grup AQP4 seronegatif olan NMOSD hastalarının yaklaşık % 30-40'ını oluşturur (5). Bu hastaların klinik fenotip olarak MS ve NMOSD'larda farklı olduğu görüldü ve yakın zamanda MOG antikor hastalığı(MOGAH) ayrı bir nozolojik antite olarak kabul edildi(6).

Epidemiyoloji

NMOSD genellikle sporadiktir, birkaç ailevi olgu tanımlanmıştır (3). Görülme sıklığı ve prevalansı 100.000'de sırasıyla 0.05-0.4 ve 0.52-4.4 arasındadır (7). Tüm ırkları ve etnik kökenleri etkilemesine rağmen beyaz olmayanlarda eğilim vardır. İnsidansı Martinique, Batı Hint Adaları, Asya ve Afro-Amerikan popülasyonlarında, Kafkas popülasyonlarına göre daha yüksektir. Afrikalılar da yüksek ölüm oranları bildirilmiştir (3,5,89).

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jiyle birlikte yürütülen çalışmalar, biyolojik alan-daki ilerlemeler ile SSS'inin yeni idiyopatik demiyelinizan hastalıkları tanımlanmıştır. Gelecekte benzer çalışmalar mevcut hastalıkların daha iyi anlaşılması, tedavilerin belirlenmesi yanında yeni fenotiplerin tanımlanmasında yardımcı olacaktır.

Anahtar Kelimeler: İdiyopatik, demiyelinizan hastalık, multipl skleroz, varyant.

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