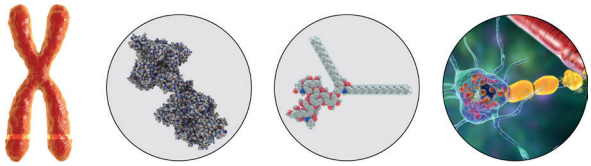

BÖLÜM 7

NÖROMİYELİTİS OPTİKA SPEKTRUM BOZUKLUKLARI

Serdar AYKAÇ[1]

GİRİŞ

Nöromiyelitis optika spekturum bozuklukları (NMOSB) (daha önce Devic hastalığı veya Nöromiyelitis optika olarak bilinen), merkezi sinir sisteminin demiyelinizasyon ve aksonal hasarı ile karakterize immün aracılı inflamatuvar bir hastalığıdır. Ağırlıklı olarak merkezi sinir sisteminde, optik sinirleri ve omuriliği hedef alır (1).

Nöromiyelitis optika terimi ilk olarak 1894 yılında, Devic ve Gault tarafından monofazik bilateral (veya hızlı ardışık) optik nörit ve miyelit seyiri olan bir dizi hasta sunumu ile tanımlanmıştır (2). Aynı araştırmacılar tarafından bazı hastalarda beyin sapı lezyonu bulgularının olabileceği de bildirilmiştir (3,4). Peppo Acchioté'nin 1907 yılındaki makalesinde nöromiyelitis optika'ya "Devic Hastalığı" ismi verilmesi önerilmiştir (5). Yıllar içerisinde hastalığın tanı ve dışlanma kriterleri pek çok kez değiştirilmiştir. Devic ve Gault' un beyin sapı bulgularını tanımlamasına rağmen; ilk dönemlerde relapslar ile giden hastalar, tek taraflı optik nöriti olan hastalar, optik nörit ve transvers miyelit dışında kalan semptomlar NMO tanısında dışlanmıştır (5). 1930 lu yıllarda Russell Brain tarafından NMO'nun bir MS alt tipi olduğu ileri sürülmüştür. Tartışmalar 20.yüzyıl boyunca devam etmiş ve NMO'nun ayrı bir hastalık mı yoksa bir MS alt tipi mi olduğu belirsizliğini korumuştur (5). 1996 yılında bu konuyu açıklığa kavuşturmak için yürütülen bir çalışmada NMO'nun ayrı bir hastalık olduğu sonucuna varılmıştır (6).

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AQP4-IgG için seropozitif olan hastalarda, tek bir atakla başvuranlar da dahil olmak üzere, nüks açısından yüksek risk altında olduklarından, immünsupresyon genellikle en az beş yıl sürdürülür. Bununla birlikte, AQP4 seropozitifliği olsun veya olmasın NMOSB için optimal ilaç rejimi ve tedavi süresi henüz belirlenmemiştir ve birçok otorite ömür boyu tedavi önermektedir (106).

SONUÇ

NMOSB, santral sinir sisteminin nadir görülen antikor aracılı otoimmün bir hastalığıdır. Miyelit, şiddetli optik nörit ve/veya inatçı kusma ve hıçkırık nöbetleri (area postrema sendromu) hastalığın klasik belirtileridir ve klinisyeni tanı konusunda uyarabilir. Relapslar, morbiditeyi önlemek için hızlı ve agresif olarak yüksek doz steroid +/- plazma değişimi ile tedavi edilmelidir. AQP4 antikorları olan tüm hastalar, başka atakları önlemek için süresiz olarak immünsuprese edilmelidir. Nüksün önlenmesi çok önemlidir ve uzun süreli immünsupresyon ile elde edilebilir.

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