BÖLÜM 104

İMMÜN-ARACILI SEREBELLAR ATAKSİ

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GİRİŞ

Serebellar ataksi (CA) gelişiminden sorumlu patogenetik etki hücre ölümüne yol açan immün aracılı disfonksiyon mekanizmasıdır.[1] İmmüm mekanizmaya bağlı gelişen serebellar ataksiler (ICMA) karekteristik olarak; gluten ataksi (GA), paraneoplastik serebellar dejenerason (PCD), anti-glutamat dekarboksilaz 65 antikor-ilişkili serebellar ataksi (anti-GAD65-ilişkili CA), post-infeksiyöz serebellit ve opsoklonus miyoklonus sendromu (OMS) gibi çeşitli oto-immün temelli etiyolojileri kapsar. Teröpatik yanıtın IMCA etiyolojisine göre değiştiği düşünülmektedir.[1-9] CA yönetiminde farklılaşan etyolojilere, tedavi yöntemlerine ve sağlam doku kapasitesinin korunması olarak tanımlanan serebellar rezervin halen yeterli olduğu zaman aralığında erken immünoterapinin önemini vurgulayan klavuzlar yayınlanmıştır.[10-12] Yakın dönemli çalışmalarda IMCA patogenezine ilişkin birçok hücre ve antikor aracılı immün mekanizma ile ilgili yeni bulgular tanımlanmıştır. Bu çalışmalar erken tedavinin önemine ışık tutmakta olup ICMA klinik alt tiplerinin prevelansını ortaya koymaktadır.[13,14] Bununla birlikte ataksilerin büyük kısmına halen tanı konamamaktadır. Çünkü ICMA'nın progresif formları dejeneratif CA profillerini taklit eder. Klinik girişimlerdeki gecikmeler maalesef tedavi fırsatlarının kaybı ile ilişkilidir.[13,14]

SINIFLAMA

Öykü

İmmün aracılı CA'leri ilk olarak 1868 yılında JM Charcot tanımlamıştır.[1] Bir sonraki tarihi mihenk taşı ise 1919 yılında B. Brouwer tarafından CA'ler ile malignansiler arasındaki ilişkinin tanımlanmasıdır.[15] Son otuz yılda çeşitli malignansi tipleri ile CA arasında ki ilişki araştırılmıştır. Sonuç olarak çeşitli neoplazi kategorilerinde anti-Yo, anti-Hu, anti-Tr, anti-CV2, anti-Ri, anti-Ma2 ve anti-VGCC gibi spesifik oto-antikorlar belirlenmiştir.[3-8] Yüzyılın başında GA ve anti-GAD65 antikor ilişkili CA üzerine çeşitli çalışmalar yapılması dönüm noktası oluşturmuştur. Her iki klinik durumda da hafif serebellar atrofi varlığı veya atrofi olmaması ile oto-antikorların birlikteliği karekterizedir. MS ve PCD gibi klasik hastalıklar ile birlikte, klinik IMCA kategorileri artık ataksioloji olarak yerleşmiştir.[1,10,16]

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