Anti-MOG encephalitis mimicking small vessel CNS vasculitis

Objective To report 2 patients with anti–myelin oligodendrocyte glycoprotein (MOG)-associated encephalitis who were initially misdiagnosed with small vessel primary CNS vasculitis.

Methods Review of symptoms, MRI and neuropathologic features, and response to treatment. MOG antibodies were determined in serum and CSF using a cell-based assay.

Results Symptoms included fever, headache, and progressive mental status changes and focal neurologic deficits. CSF studies revealed lymphocytic pleocytosis, and both patients had abnormal brain MRIs. Brain biopsy samples showed prominent lymphocytic infiltration of the wall of small vessels; these findings initially suggested small vessel CNS vasculitis, and both patients were treated accordingly. Although 1 patient had a relapsing-remitting course not responsive to cyclophosphamide, the other one (also treated with cyclophosphamide) did not relapse. Retrospective assessment of serum and CSF demonstrated MOG antibodies in both cases, and review of biopsy specimens showed absence of fibrinoid necrosis (a pathologic requirement for small vessel CNS vasculitis).

Conclusions Anti–MOG-associated encephalitis can be mistaken for small vessel CNS vasculitis. This is important because the diagnosis of anti–MOG-associated encephalitis does not require brain biopsy and can be established with a serologic test.

Real-world validation of the 2017 McDonald criteria for pediatric MS

Objective To compare the diagnostic accuracy of the McDonald 2017 vs the McDonald 2010 criteria to predict a second attack of multiple sclerosis (MS) (clinically definite MS [CDMS]) at the first attack of acquired demyelinating syndromes (ADS).

Methods One hundred sixty-four children (aged <18 years) with an incident attack of ADS were included in a prospective multicenter study between June 2006 and December 2016. Brain (and spinal if available) MRI was performed ≤3 months after symptom onset. Sensitivity, specificity, positive predictive value, negative predictive value, and accuracy were compared at baseline between the 2010 and 2017 criteria.

Results Among the 164 patients, 110 patients (67%) presented without encephalopathy (ADS−, female 63%; median age, 14.8 years; interquartile range [IQR], 11.3–16.1 years) and 54 (33%) with encephalopathy (acute disseminated encephalomyelitis [ADEM], female 52%; median age, 4.0 years; IQR, 2.6–6.1 years). Of the 110 ADS− patients, 52 (47%) were diagnosed with CDMS within a median follow-up of 4.5 years (IQR, 2.6–6.7 years). The sensitivity was higher for the 2017 criteria than for the 2010 criteria (83%; 95% confidence interval [CI], 67–92, vs 49%; 95% CI, 33–65; p < 0.001), but the specificity was lower (73%; 95% CI, 59–84 vs 87%; 95% CI, 74–94, p = 0.02). At baseline, 48 patients fulfilled the 2017 criteria compared with 27 patients when using the 2010 criteria. The results for children aged <12 years without encephalopathy were similar. In patients with ADEM, 8% fulfilled the 2010 criteria and 10% fulfilled the 2017 criteria at baseline, but no patient fulfilled the criteria for CDMS.

Conclusions The McDonald 2017 criteria are more sensitive than the McDonald 2010 criteria for predicting CDMS at baseline. These criteria can also be applied in children aged <12 years without encephalopathy but not in children with ADEM.