MOG-IgG serological status matters in paediatric ADEM

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Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating disease of the central nervous system (CNS) characterised by encephalopathy and other neurological manifestations. ADEM is a heterogeneous syndrome predominantly found in the paediatric population. ADEM usually takes a monophasic course, but some patients have a relapsing disease. Patients diagnosed with ADEM have multifocal CNS lesions including of the spinal cord and/or optic nerves. Therefore, ADEM should be distinguished from other diseases such as multiple sclerosis (MS) and neuromyelitis optica spectrum disorders (NMOSD). Over the past years, several disease-specific autoantibodies such as aquaporin-4 (AQP4)-IgG in NMOSD have been discovered, revolutionising the understanding of underlying pathogenic mechanisms and identifying a broader clinical spectrum related to the disease, as seen in NMOSD.1 However, antibodies against myelin oligodendrocyte glycoprotein (MOG) have been reported in some inflammatory CNS diseases and early assays using denatured protein (western blot) or linear peptides (ELISA) showed low specificity. The interest on MOG-IgG has risen again recently after the development of assays that can detect conformational sensitive antibodies in unique subgroups of patients with paediatric ADEM and NMOSD.2,3 Baumann et al4 report clinical and MRI features of paediatric ADEM with (n=19) and without (n=14) serum MOG-IgG. None were AQP4-IgG-positive. They found that ADEM patients with MOG-IgG had large, widespread brain lesions with ill-defined borders and more commonly longitudinally extensive spinal cord lesions on the MRI. On the other hand, MRI lesions atypical for ADEM, most of which are typical for MS, were rarely seen in MOG-IgG-positive children. MOG-IgG seropositivity was associated with significantly better clinical outcome. The majority of patients showed drastic reduction of MOG-IgG in the follow-up and had a single attack, but three patients remained with high-titres of MOG-IgG and relapsed, suggesting that longitudinal testing of MOG-IgG may have prognostic implications. Taken together, MOG-IgG-positive ADEM appears to have uniform MRI features and better clinical recovery. However, further research is required to reveal the pathogenic role of the MOG-IgG and to uncover the whole scope of neurological diseases associated with MOG-IgG.

REFERENCES
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