An Adult Case of Anti-Myelin Oligodendrocyte Glycoprotein (MOG) Antibody-associated Multiphasic Acute Disseminated Encephalomyelitis at 33-year Intervals

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Abstract

Acute disseminated encephalomyelitis (ADEM) followed by optic neuritis (ON) has been reported as a distinct phenotype associated with anti-myelin oligodendrocyte protein (MOG) antibody. We herein report the case of a 37-year-old woman who was diagnosed with ADEM at 4 years of age and who subsequently developed ON followed by recurrent ADEM 33 years after the initial onset. A serum analysis showed anti-MOG antibody positivity. This phenotype has only previously been reported in pediatric cases. Neurologists thus need to be aware that the phenotype may occur in adult patients, in whom it may be assumed to be atypical multiple sclerosis.

Key words: MOG, ADEM, optic neuritis, adult


Introduction

Acute disseminated encephalomyelitis (ADEM) is defined as a monophasic inflammatory demyelinating disorder (1). However, due to its heterogeneity, some recent series have blurred the distinction between ADEM and other demyelinating disorders of the central nervous system (CNS) by proposing an expanded spectrum of ADEM, which includes multiphasic forms (2). Recently, ADEM followed by optic neuritis (ON) has been reported as a recognizable and distinct phenotype of multiphasic ADEM associated with anti-myelin oligodendrocyte protein (MOG) antibodies (3-5). This phenotype has only previously been reported in pediatric cases. However, the long-term prognosis of the phenotype remains unclear. We herein report an adult case of ADEM in which ON with anti-MOG antibodies and recurrent ADEM occurred more than 30 years after the initial event.

Case Report

The patient was a 37-year-old woman with no history of collagen disease. She had been diagnosed with ADEM at 4 years of age. She developed an altered mental state, after experiencing headache and nausea, without any apparent visual impairment. Her symptoms were not associated with an antecedent infection or vaccination. Steroid therapy was administered and she recovered without any residual deficits. Since then, she had experienced no neurological problems until one day in 2014 when she developed a headache, fever, and orbital pain. One week after the onset of symptoms, she was admitted due to a vision disturbance in her right eye. She had given birth to her second child four months before the onset symptoms. Neither infection nor recent vaccination was noted prior to the onset of these symptoms.
Her right visual acuity was reduced to counting fingers. There were no apparent signs of meningeal irritation. Her deep tendon reflexes and plantar reflexes were normal. The laboratory results showed a white blood cell count of 7,500/μL, and a C-reactive protein level of 0.01 mg/dL. Negative results were obtained for anti-neutrophil cytoplasmic antibodies and anti-nuclear antibodies, including SS-A and SS-B antibodies. In the cerebrospinal fluid (CSF), the cell count was 17/mm³ (monocytes, 16/mm³), the protein concentration was 31 mg/dL, and the myelin basic protein (MBP) level was abnormally elevated [191 pg/mL (normal, <102 pg/mL)]; however, no oligoclonal bands (OCBs) were evident. The immunoglobulin (Ig)G index was within the normal range. The patient showed strongly positive results for serum anti-MOG antibodies in a cell-based assay, which was described by Sato et al. (6), but negative findings for serum anti-aquaporin 4 (AQP4) antibodies. Contrast-enhanced MRI showed right optic nerve swelling with enhancement (Figure a, b). Meningeal enhancement was also observed along the interhemispheric fissure (Figure c). MRI revealed no spinal cord abnormalities. Her visual acuity improved immediately after the initiation of steroid pulse therapy [methylprednisolone (1 g/day for 3 days), intravenously] and recovered completely to 20/20 vision. She was discharged 21 days after the onset of symptoms. However, she was readmitted 4 days later due to headache and numbness. At her readmission, an abnormal sensation in the right extremities, signs of meningeal irritation, and right Horner’s syndrome were observed. She still had normal deep tendon reflexes, which were slightly brisker than at her previous admission. Her planter response was normal. A CSF analysis showed a cell count of 24/mm³ (monocytes, 6/mm³), and a protein level of 29 mg/dL. Her MBP level remained high, at 144 pg/mL. Neither OCBs nor the elevation of her IgG index level were detected. Hyperintense lesions were observed in the right pallidum, right posterior limb of the internal cap-
The anti-MOG antibodies in the phenotype were detected from the time of first onset of ADEM and remained present through the recurrence of ON or ADEM (3, 4). Therefore, we suggest that the anti-MOG antibodies in the present case may have been present from the first onset of ADEM and remained positive for more than 30 years. Few reports have examined the length of time for which anti-MOG antibodies remain positive. According to a study which followed pediatric patients with anti-MOG antibodies associated with demyelinating disorders for 5 years, these antibodies occasionally persist in patients with multiphasic events (14). However, the long-term changes to the levels of this antibody that take place over decades remain unclear (14). Further reports on prospectively followed patients with this phenotype are needed.

The present case suggests that multiphasic ADEM with ON can be associated with anti-MOG antibodies in both children and adults. Neurologists need to be aware of this phenotype, which may be assumed to be atypical MS.

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References


