Case Report

Anti-myelin oligodendrocyte glycoprotein – antibody presenting as tumefactive demyelination and hemiparesis in a pediatric patient

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ABSTRACT

Anti-myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease is an immune-mediated central nervous system demyelinating disorder with a variety of phenotypic presentations such as acute disseminated encephalomyelitis, optic neuritis, and transverse myelitis but rarely presents as tumefactive lesion, particularly in children. We present a case of an adolescent girl with an anti MOG antibody-related disorder that manifested as a tumefactive lesion.

Key words: Hemiparesis, MOG associated disease, Magnetic resonance imaging, Tumefactive demyelination

Anti-myelin oligodendrocyte glycoprotein (MOG) – antibody-associated disease is an immune-mediated central nervous system demyelinating disorder with a variety of phenotypic presentations including acute disseminated encephalomyelitis (ADEM), optic neuritis, and transverse myelitis, and it rarely presents as a tumefactive lesion. Tumefactive demyelination can cause isolation at the onset or during the progression of other diseases. MOG antibodies are found in one-third of children with ADEM. The presence of MOG antibodies varies with age. Seropositivity rates are highest in young children with ADEM, and lowest in older children with optic neuritis, myelitis, or brain stem symptoms [1].

Tumefactive demyelinating lesion is uncommon in MOG-related disease. Clinical characteristics vary depending on the size and location of the lesion, as well as the degree of mass effect. The most common presenting complaint is hemiparesis or hemiplegia. Aphasia, headache, visual, and cognitive disturbances are among the other symptoms. Tumefactive demyelinating lesion is frequently indistinguishable from brain tumors on magnetic resonance imaging (MRI) and occurs in a variety of autoimmune inflammatory conditions including multiple sclerosis (MS), neuromyelitis optica spectrum disorders, systemic lupus erythematosus, sarcoidosis, and Behcets disease. As a result, early and accurate diagnosis is critical for proper management [2].

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CASE PRESENTATION

A 12 year old developmentally normal female child presented with complaints of one episode of convulsion described as clonic movement of the left upper and lower limb lasting about 5 min, slurring of speech, and weakness of the left upper and lower limb following convulsion, 8 h before admission. The child was in perfect health. The central nervous system examination revealed distal muscle weakness of the left upper and lower limbs (power of 3/5) with no involvement of the cranial nerves, sensory, cerebellar, or meningeal systems. Within 24 h, her left upper limb power improved to 4−/5 and her left lower limb power improved to 4+/5. As shown in Fig. 1, an MRI brain (plain) revealed altered signal intensity involving the cortical and subcortical locations of the right parietal lobe.

Routine blood tests and CSF analysis were within normal ranges. A normal MRI brain contrast was used to rule out tumorous lesions. Indirect immunofluroscence was used to test serum for antibodies against MOG. It is a cell-based immunoassay using transfected cell lines for *in vitro* quantification of human IgG antibodies to anti MOG IgG antibodies, and it was strongly positive, as shown in Fig. 2.

After 5 days of intravenous methyl prednisolone 30mg/kg/day followed by oral steroids prednisolone 1.5 mg/kg/day, her power in the left upper limb improved to 4+/5 and 5/5 in the left lower limb. The child is currently on oral prednisolone maintenance therapy and is being monitored. After 4 weeks, power improved to 5/5 in all four limbs.

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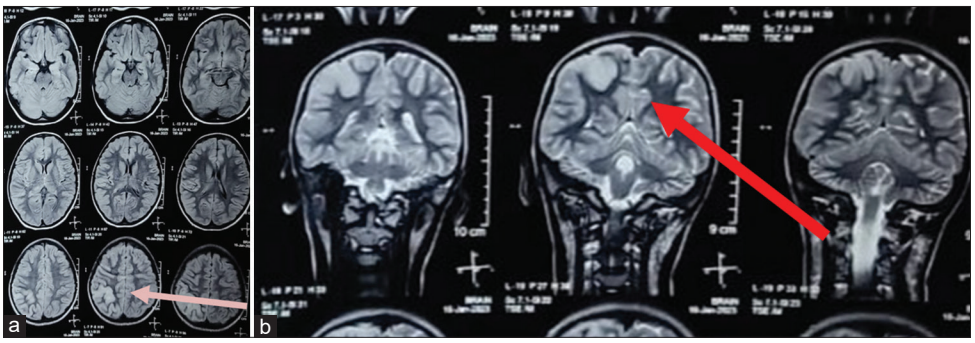

Figure 1: (a) Fluid attenuated inversion recovery MRI brain showing hyperintensity in the right parietal subcortical region adjacent to the gyri (marked with the arrow). (b) Magnetic resonance imaging brain showing circumscribed lesion in the right parietal region (marked with the arrow)

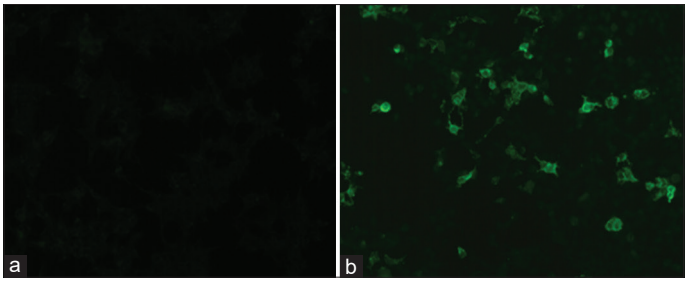

Figure 2: Comparison of MOG-IgG negative and positive samples by live cell-based assay immunofluorescence (CBA-IF). Representative fluorescence image of (a) negative and (b) positive samples

DISCUSSION

Anti-MOG antibody-associated disease (MOGAD) manifesting as tumefactive demyelination is uncommon. The onset age ranges from 6 to 66 years [1,2] MOGAD is a newly identified autoimmune disease whose phenotype is expanding with each new patient case. While positive serum MOG antibodies were once thought to be a biological marker for MS, it is now widely accepted that the presence of MOG antibodies typically indicates a non-MS course. The prevalence of tumefactive demyelination is estimated to be 1–2/1000 MS cases. There is no clear gender preference [3].

The initial symptoms differed depending on the location of the lesion. The majority of them responded favorably to steroids. MOG does not have a distinct radiographic pattern, but leptomeningeal enhancement, thalamic lesions, pontine lesions, deep white matter lesions, tumefactive, poorly defined lesions, and cortical lesions were more common in MOGAD than in NMOSD or non-MOG antibody cases. The lesions usually improve or disappear completely over time [3,4]. Tumefactive demyelination is defined as demyelinating lesions (2 cm or greater) with or without cortical involvement or mass effect, and is distinguished from brain tumor by incomplete ring enhancement, mixed T2 weighted iso- and hyperintensity of enhanced regions with or without cortical involvement or mass effect. In the reported cases, brain biopsies revealed mostly MS pathological pattern II or III [2].

The severity of the initial presentation, recovery from the initial attack, and even antibody titers cannot predict which patients will relapse. Relapsing disease rates vary by series, but have been reported to be as high as 72–88% [5] MOG has a better prognosis than aquaporin four positive disease, with faster recovery and less disability [6]. Because half or more of MOGAD patients will have a monophasic disease course, deciding whether or not to start chronic immunosuppression at the time of initial presentation is difficult. Tocilizumab has been used off-label in adult patients with refractory MOGAD and in both adult and pediatric NMOSD patients [7,8].

This case report describes the clinical and radiologic manifestations of anti-MOG antibody-associated disease in a child who responded well to steroids. This should always be taken into account when making a diagnosis of tumefactive lesions.

In our case, the child was diagnosed in 2 days and treatment was started right away, resulting in complete improvement in distal muscle power. The lack of CSF anti-MOG antibodies and histopathological correlation of the lesion in our case was the study's limitation.

CONCLUSION

Tumefactive demyelination is a symptom of a number of immunologic-mediated neurologic diseases that can be difficult to diagnose; distinguishing this demyelinating lesion from malignancy or infection is critical for management. More research is needed to fully understand the pathogenesis of the disease.

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