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Case Report

Myelin oligodendrocyte glycoprotein antibody associated disease – two distinct clinical presentations

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ABSTRACT

Myelin oligodendrocyte glycoprotein antibody associated disease (MOGAD) is an inflammatory demyelinating disorder caused by the development of immunoglobulin G autoantibody targeting MOG, which is expressed exclusively on the outer surface of the myelin sheath and plasma membrane of oligodendrocyte. MOGAD can present in different clinical spectrum including acute disseminated encephalomyelitis (ADEM), optic neuritis (ON), transverse myelitis and relapsing courses of these presentations. Depending on the severity, acute attacks can be treated with I V methyl prednisolone, plasma exchange or IVIG as in any demyelinating disease. This report is about 2 children with MOG antibody positive disease who presented with different symptomatology, severity and outcome.

Keywords: Myelin oligodendrocyte glycoprotein, Demyelinating disorder, Acute disseminated encephalomyelitis, Optic neuritis

INTRODUCTION

Myelin oligodendrocyte glycoprotein antibody associated disease (MOGAD) is an inflammatory demyelinating disorder affecting the central nervous system. It is caused by the development of immunoglobulin G autoantibody targeting MOG, which is expressed exclusively on the outer surface of the myelin sheath and plasma membrane of oligodendrocyte.[1] In the background of increasing awareness about this condition in pediatric population, here by describe 2 cases of MOGAD with 2 distinct way of presentation admitted during same period of time.

CASE REPORT

Case 1

7 years old girl with no significant illness in the past, normal development, fully immunised having normal growth parameters presented with features of acute encephalitis. Child was active in all her daily activities. Within a period of 1 month, mother noticed general inattention in activities and worsening of scholastic performance. She had a 3 weeks history of intermittent fever, abdominal pain, headache and behavioral changes like making loud inappropriate sounds, decreased sleep, clenching of teeth and restlessness. Later on child developed generalised tonic clonic seizure (status epilepticus) and went in to coma having a Glasgow coma scale (GCS) score of E1V1M3. Her hemodynamic parameters were stable, child was intubated and mechanically ventilated. Treatment was started empirically with iv antibiotics, antiviral, anti-tubercular drugs, anti-epileptics and other supportive measures. Magnetic resonance imaging (MRI) showed bilateral asymmetric multifocal gyral edema, gangliothalamic, callosal and brain stem T2 flair hyper intensities with nodular, parenchymal, leptomeningeal and cervical cord enhancement and mild bilateral optic neuritis. Cerebrospinal fluid (CSF): elevated protein and

pleocytosis. Intravenous methyl prednisolone 30 mg/kg/day and intravenous immunoglobulin (IVIG) 400 mg/kg/day were given for 5 days, then switched over to oral presnisolone. CSF MOG antibodies came to be positive and neuromyelitis optica (NMO) antibodies negative. Due to the devastating presentation of the disease, weekly I V rituximab was given for 4 weeks. Patient showed gradual clinical improvement, but ophthalmology check-up revealed bilateral optic atrophy. Now she is conscious, able to walk, speaks with poor coherence and totally blind.

Case 2

A 11 years old boy who is developmentally normal with adequate growth, normal intelligence and behaviour having past history of neurological issues admitted with 3 episodes of seizures. 1st episode was a left sided focal seizure turned out to be a generalised seizure, 2nd and 3rd episodes being left focal seizure with no loss of consciousness. Physical and neurological examination were normal with no involvement of cranial nerves.

His problems started at the age of 5 years with history of high grade fever of 2 weeks duration along with vomiting and gradual development of difficulty in speech and irritability. MRI brain showed bilateral asymmetric multifocal ill-defined cortical, subcortical T2 flair hyperintensities. The episode was treated with IV methyl prednisolone for 5 days followed by oral steroid. While on oral steroids, after 1 month he developed acute onset visual loss and pain of left eye. MRI taken was suggestive of optic neuritis and visual evoke potential was absent in left eye. Serum for NMO antibodies were negative. Another 5 days course with methyl prednisolone results in significant improvement in vision. 5 months later, he again presented with acute onset visual loss and pain of same eye. Managed with the same treatment protocol and showed good improvement in vision. Continued on oral steroids for few more months, but they discontinued the treatment and lost follow up. The next 5 years were uneventful. This time presented with seizure, but no visual loss. MRI brain was repeated again, showed bilateral asymmetric cortical subcortical T2 flair hyperintensities consistent with demyelinating etiology (Figure 1).

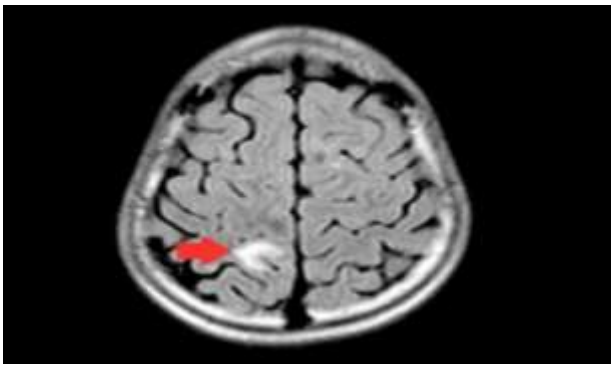

Figure 1: Area of demyelination (arrow).

Treatment started with methyl prednisolone 30 mg/kg/day for 5 days followed by oral steroids. MOG Antibody became positive and NMO antibody negative. CSF study was normal. CSF oligoclonal bands were also negative. Due to the relapsing course of disease, immunotherapy was started with oral azathioprine. After initiation of treatment he was asymptomatic and now is under follow up.

DISCUSSION

MOGAD is an acquired demyelinating disease associated with MOG-antibodies having a wide clinical spectrum with monophasic diseases such as acute disseminated encephalomyelitis (ADEM), optic neuritis (ON), and transverse myelitis and also relapsing courses of these presentations.[2] In almost all patients present with relapsing ADEM, MOG antibody is positive. It is more common in children and presentation is heterogenous.[3] In our study 1st child had acute demyelination presented as encephalitis with optic neuritis where as the 2nd child had a relapsing course. There are studies which correlating MOG antibody titre with clinical time course and prognosis.[4,5] MOG positivity is possible even in the absence of suggestive MRI findings.[6] Perivenous and confluent white matter demyelination, MOG-dominant myelin loss, intracortical demyelination, predominant CD4+ T-cell and granulocytic inflammation, complement deposition within active white matter lesions, partial axonal preservation, and reactive gliosis are the neuropathological features of MOGAD described which is similar to that seen in ADEM.[7-10] Depending on the severity ,acute attacks can be treated with IV methyl prednisolone, plasma exchange or IVIG as in any demyelinating disease. Though the efficacy is not clear, azathioprine and mycofenolate mofetil are considered for long term use and rituximab as a second line agent.[3] MOG antibody disease is having relatively favorable prognosis on long term.[11]

CONCLUSION

MOGAD is a clinical condition which can present with varying symptomatology, severity and outcome. Investigating a child who is presenting with features of acute encephalitis with serum or CSF MOG antibodies will help to identify this condition. Early identification and treatment with IV methyl prednisolone or IVIG will influence the prognosis.

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