Pediatric Onset Myelin Oligodendrocyte Glycoprotein-Associated Disease: A Series of Four Cases

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Abstract

Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease (MOG-AD) is a rare inflammatory demyelinating disease of the central nervous system with monophasic and relapsing manifestations. The most common presenting phenotypes of MOG-AD in pediatric population include acute disseminated encephalomyelitis (ADEM), optic neuritis (ON), transverse myelitis (TM) and brainstem syndromes. Here, we report four cases of MOG-AD in pediatric patients. Patients presented with headache, gait disorder, dysarthria, seizure. The first case had weakness of both lower limbs with irritability, 2nd case had altered conscious level with seizure along with walking and speech difficulty. The 3rd case had relapsing features, visual and speech problem with optic neuritis after 2 years of treatment. The 4th case had headache and unsteady gait, visual impairment and speech difficulty. Most of the patients had demyelinating features in magnetic resonance imaging (MRI) of brain. All the four cases were positive for anti-MOG antibody. Treatment was done according to the protocol of the institute with steroid, intravenous immunoglobulin and rituximab.

Key words: children, myelin-oligodendrocyte glycoprotein, myelin oligodendrocyte glycoprotein antibody-associated disease.

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Introduction

Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease (MOG-AD) is an inflammatory demyelinating disease of the central nervous system mediated by MOG antibodies, characterized by loss of myelin in the white matter, optic nerves and spinal cord. MOG is a myelin protein expressed on the outermost layer of the myelin sheath and oligodendrocyte membranes in the central nervous system. MOG-AD represents various clinical phenotypes, with a higher occurrence in children than in adults. It is commonly associated with an acute disseminated encephalomyelitis (ADEM), optic neuritis and/or transverse myelitis in the pediatric population.

The clinical manifestations of MOG-AD are related to age. Previous studies suggested that the clinical presentation in children with MOG-AD exhibits a bimodal distribution. Among children aged 4 to 8 years, encephalopathy is more frequently dealt, while in children aged 13 to 18, optic neuritis (ON) is more commonly seen.[1] Phenotypes are: ADEM which is common in pediatric age, ON (41-63%), transverse myelitis (30%), brainstem syndrome (30%). The disease course can be monophasic or multiphasic (relapsing-remitting), with approximately 30–40 % of children experiencing a multiphasic course.[2]

The incidence of MOG-AD ranges between 1.6 and 3.4 per 1,000,000/years. Male and female ratio is 1:1. Children

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account for about 50% cases. Median age of onset is 20-30 years. Geographic distribution is variable. Pediatric population have more heterogeneous presentation than adults.[3] In 2023, the international MOG-AD panel established formal consensus criteria for diagnosis of MOG-AD as a distinct entity. The criteria requires detection of MOG antibodies and a presence of a core clinical demyelinating event, including ADEM, ON, transverse myelitis, cerebral cortical encephalitis often with seizures, brainstem or cerebellar deficits and cerebral monofocal or polyfocal deficits. MRI is essential for diagnosis as well as for follow-up of MOG-AD patients.[4] In this paper, we present four pediatric cases of MOG-AD with diverse presentation, with one having refractory MOG-AD (Table-1).

Case Series

Case 1

A 2-year-old immunized, male child, 2nd issue of non-consanguineous parents, got admitted with the complaints of weakness of both lower limbs for 1 month manifested by difficulty in walking, frequent fall, inability to sit without support along with irritability and restlessness. He had history of low-grade fever for 3 days, 20 days prior to this illness. On examination, he was ill looking, irritable, restless, upper motor neuron lesion was present in both lower limbs. He was treated with IV methylprednisolone (IVMP) for 5 days followed by oral prednisolone. Then again after 40 days of onset of illness, he came with dimness of vision. Fundoscopy was normal. Gradually he developed ataxia and tremor of all 4 limbs after an episode of febrile illness. This time he was treated with Inj. Immunoglobulin (IVIG). His first MRI of brain findings were suggestive of ADEM (Fig-1A). His 2nd MRI of brain showed, T2 -Flair hyperintense in both para ventricular and supra ventricular deep white matter (Fig-1A). MRI of orbit and spine was normal (Fig-1B). Serum MOG antibody titre was positive. After first relapse, he was given monthly IV MP for total 7 cycles and IVIG for 2 cycles and started mycophenolate mofetil (MMF) which was planned to be continued for at least 2 years. Currently he is in remission.

Case 2

A 12-year-old immunized girl, 2nd issue of non-consanguineous parents presented with history of recurrent episodes of encephalopathy evident by altered level of consciousness, seizure, headache and walking and speech difficulty for 4 years. First attack was followed by a febrile illness. On examination, she was ill looking, vital signs within normal limit, anthropometrically well thrived. Nervous system examination showed slow slurred speech, impaired coordination and ataxic gait. MRI of brain revealed bilateral, diffuse, extensive ill-defined hyperintense lesions in periventricular and subcortical white matter region (Fig-1C). Microscopic examination of CSF showed lymphocytosis (100 cell/cmm). Serum for antibody titre for MOG: 1: 32 (positive). EEG showed generalized burst of spike and polyspike wave discharges. She was treated with pulse IV MP, rituximab and MMF. In follow up, she has behavioural disorder.

Case 3

A 6-year-6-month-old boy, 2nd issue of non-consanguineous parents, immunized, presented with the complaints of speech and feeding difficulty for 5 days. Initially at 4 years and 6 months of age he had history of walking and speech difficulty, abnormal behavior, drooping of eyelid followed by a febrile illness. He was treated with IVMP and was improved. He again developed walking and speech difficulty, blurring of vision following a febrile illness after 9 months of initial illness. MRI of brain revealed, diffuse patchy hyperintense lesions involving cortex, juxtracortical, deep and periventricular white matter of bilateral cerebral-hemispheres (Fig-1D). There was diffuse leptomeningeal enchancement along the surface of brain stem also. CSF study showed-MOG Ab-positive. He was treated with Inj. MP, IVIG, rituximab subsequently. He developed severe drug reaction with rituximab. After that, he was treated with tablet MMF. The patient is now on MMF and clinically stable.

Case 4

A 4-year-6-month-old boy, only issue of non-consanguineous parents presented with high grade and continued fever, lethargy, headache which was daily and progressive in nature. He also had vomiting, generalized tonic clonic seizure, visual disturbances. Gradually he developed hemiparesis and difficulty in speech. Examination revealed, hypertension and right

sided facial palsy, left hemiparesis. Eye assessment showed optic disc edema in both eyes, right eye had optic neuritis. Investigation showed neutrophillic leukocytosis (TC of WBC 33000, N-90%, Hb% 11.2, TPC-220.000), CSF study was normal. His blood for MOG antibody was positive. MRI of brain showed, Flair and T2-hyperintense lesions in supratentorial and infratentorial compartment involving gray, white matter and patchy enhancement in cerebellum (Fig-1E). He was treated with IVMP. Patient is clinically stable now.

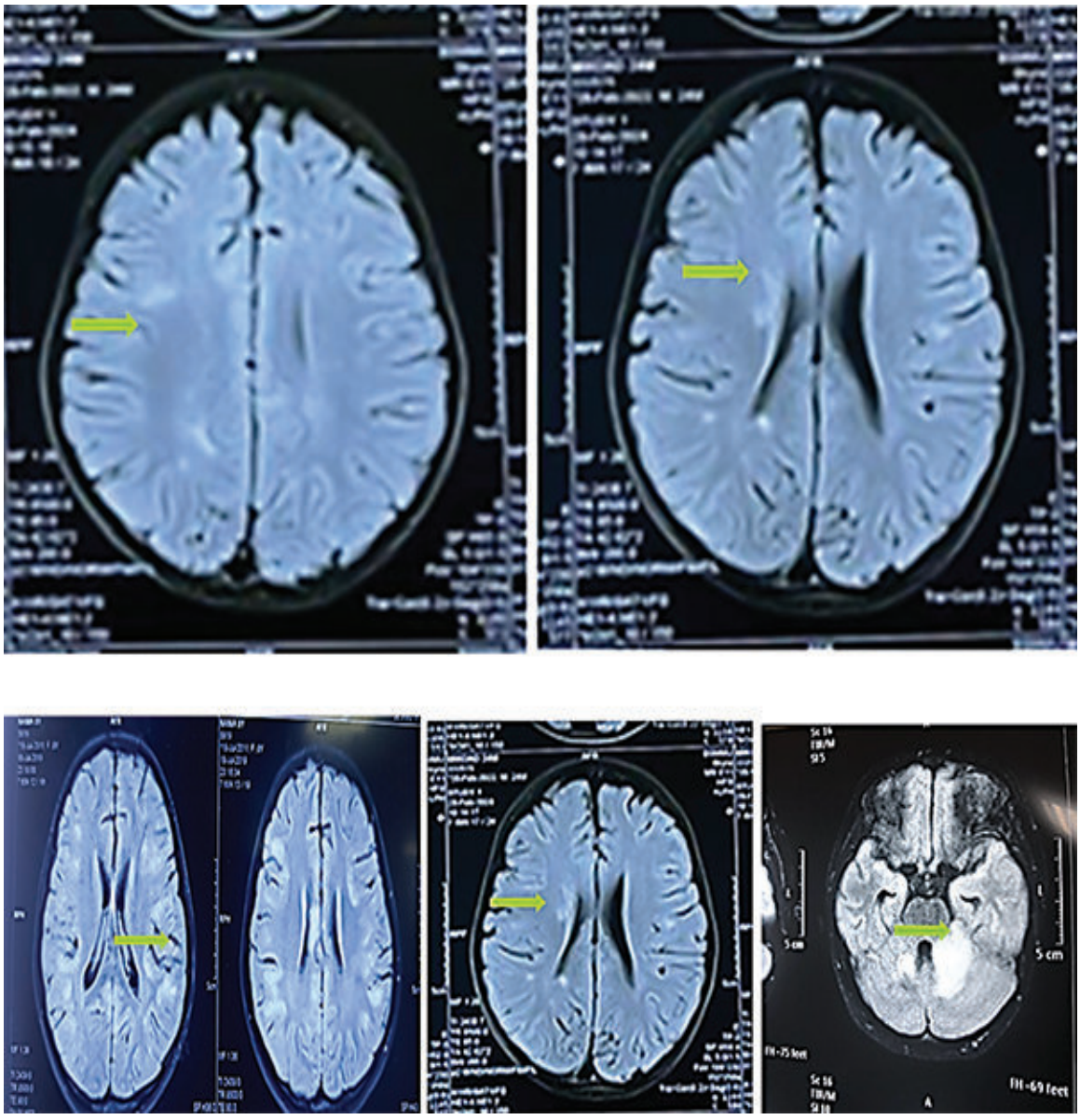

Figure 1: *A, B showing T2-flair hyperintense areas are present in both para ventricular and supra ventricular deep white matter (Case 1), C: Flair image (axial view) showing bilateral asymmetrical multiple ill-defined hyperintense lesions in periventricular and subcortical white matter region. (Case 2), D: Showing T2-flair hyperintense areas are present in both para ventricular white matter,(Case 3),* E: Showing hyperintense lesions in cerebellum.(Case 4)

Table-I

Profiles of the patients

Name of the patients	Age of onset of the patient (in year)	Clinical presentation	MRI of Brain with contrast	MRI of Spine	MRI of orbit
Case 1	2 years	Irritability, restlessness, walking difficulty, H/O fever prior to limb weakness.	T2 -Flair hyperintense areas are present in both para ventricular and supra ventricular deep white matter.	Unremarkable	Normal
Case 2	12 years	Altered level of consciousness, seizure, headache, walking and speech difficulty	Flair image in axial view shows bilateral, diffuse, extensive hyperintense lesions in periventricular, subcortical white matter region.	Normal	Normal
Case 3	6 years	Behavioral abnormality and walking difficulty, visual impairment and speech problem.	Diffuse patchy flair hyperintense lesions involving cortex, juxtracortical , deep and periventricular white matter of bilateral cerebral-hemispheres.Diffuse leptomeningeal enchancement along the surface of brain stem.	Longitudinal spinal cord involvement (>3 vertebral segments).	Diffuse enhancement of bilateral optic nerves
Case 4	4 years	Headache, unsteady gait, visual disturbances and speech difficulty	Flair and T2-Hyperintense lesions in supratentorial and infratentorial compartment involving gray, white matter and patchy enhancement in cerebellum. 2nd MRI: new lesion in cerebellum	Normal	Normal

Discussion:

This paper highlights clinical features, investigations and management of four paediatric cases of MOG-AD. Of the four cases, two cases were under 6 years; two patients showed manifestations of ADEM; one child presented with cerebellar ataxia; one exhibited ON. De Mol CL et al. (2020) revealed in their study, the most common phenotypes in children were monophasic ADEM, ON and neuromyelitis optica.[4] In our case series, 3 were male out of four patients. However, there is no racial or gender predominance in MOG-AD.[5]

The most characteristic neuro-radiological findings in children positive for MOG antibodies tend to have larger intracranial lesions that are scattered in distribution,

predominantly in the midbrain, pons, cerebellum and the gray matter and juxtacortical white matter regions of the frontal, temporal and parietal lobes.[1] More recently, distinct non-ADEM cerebral phenotypes characterized by cortical gray matter inflammation have also been reported.[6] MRI features can help in differentiating MOG-AD from multiple sclerosis and neuromyelitis optica.[7] Seropositivity status for MOG-IgG autoantibodies is important for diagnosing MOG-AD, in the context of high clinical and neuroimaging suspicion.[8]

Disease course can be either monophasic or relapsing, with subsequent relapses most commonly involving the optic nerve. Some MOG antibody–positive patients with central nervous system demyelinating events present with isolated seizures.[9] Cranial neuropathy is an emerging phenotype in MOG-AD. Residual disability develops in 50–80% of patients.[10]

Among the four children we mentioned, clinical characteristics at the onset included, fever, headache, hemiparesis, irritability, behaviour abnormality, vision and speech problem, seizure and ptosis. Brain MRI abnormality were observed in all 4 children and spinal MRI abnormality was present in 1 child. Serum neuromyelitis optica and CSF oligoclonal bands were negative in all cases. Here, the majority presented with ADEM-like lesions characterized by multiple or isolated white matter demyelinating lesions. In addition to typical ADEM-like lesions, widespread cortical lesions, brainstem or cerebellar abnormalities, and deep gray matter lesions were also observed, consistent with previous reports. One patient with spinal involvement showed longitudinal spinal cord involvement (>3 vertebral segments).

Regarding treatment, in the acute attack, IVMP leads to a favourable outcome in the majority of patients and can be followed by tapering of oral steroids up to a maximum of three months to maintain the benefit of acute treatment by suppressing disease activity. IVIG and plasmapheresis constitute second-line therapies in case of insufficient response to IVMP (Figure-2).[11] After a first relapse, maintenance treatment should be started in order to prevent further relapses and the possibility of permanent sequelae.[1] The main indication for long-term treatment is relapsing disease course.[12] Most children have a good response to acute phase treatment. For those who relapse, immunotherapies can be added as maintenance therapy and the clinical prognosis is generally good.[13] Disease course is more often monophasic but relapses are possible. Excellent motor recovery occurs after treatment. Follow-up MRI every 6 months is recommended for patients with relapsing disease on maintenance treatment.[14]

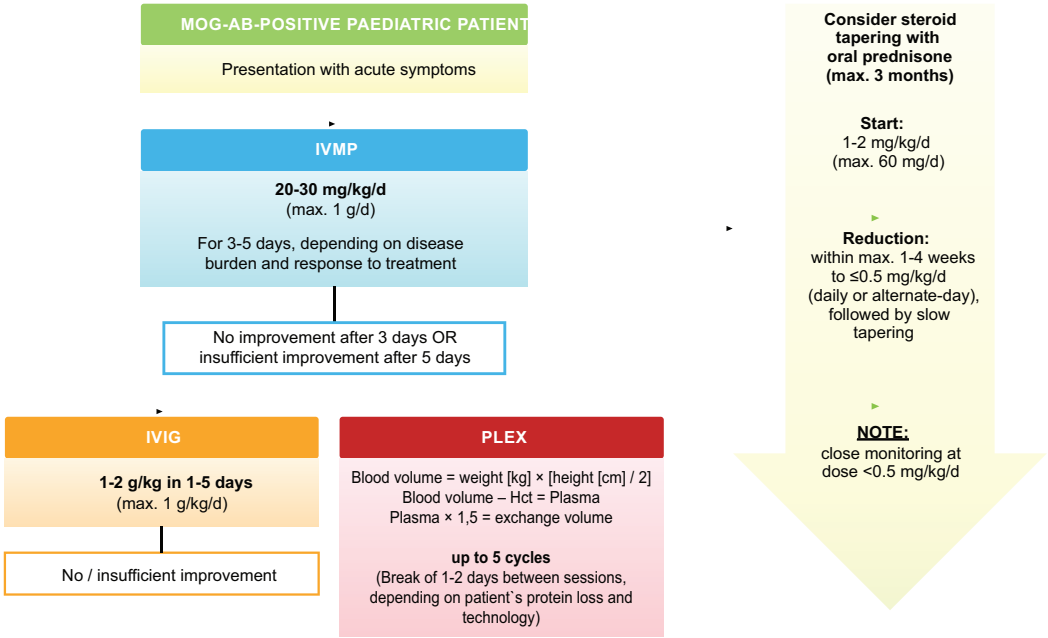

Figure 2: *Treatment Algorithm of MOGAD*

All the four children mentioned in this case series, were treated with IVMP. Among them, three children achieved disease control after getting IV MP, IV IG and MMF.

Conclusions

Here we present four cases of pediatric MOG-AD with diverse presentations. The key clinical features were fever, headache, behaviour abnormality, hemiparesis, vision and speech problem. Immunotherapy was the main modality of treatment for these children.

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Conflict of interest:

Nothing to disclose.

Ethical approval

Informed consent was taken from the parents of the patient.

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