Case Report

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Steroid-refractory Bilateral Optic Neuritis as a Presentation of Paediatric MOG Antibody-associated Disease: A Case Report

Ophthalmology Section

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ABSTRACT

Myelin Oligodendrocyte Glycoprotein Antibody-associated Disease (MOGAD) commonly presents in children as bilateral optic neuritis with disc oedema and favourable steroid responsiveness. The MOGAD is an immune-mediated demyelinating disorder of the central nervous system, increasingly recognised as distinct from multiple sclerosis and neuromyelitis optica spectrum disorder. In children, optic neuritis is the most common manifestation and is frequently bilateral, associated with optic disc oedema and good visual recovery following corticosteroid therapy. However, emerging evidence suggests that MOGAD encompasses a broader clinical spectrum, including atypical neurological features and variable treatment response. Hereby, the authors present a paediatric case of a 13-year-old girl with severe bilateral visual loss, early neurological symptoms, papilledema-like fundus appearance, and poor response to intravenous corticosteroids, requiring Intravenous Immunoglobulin (IVIg) for visual recovery. The present case highlights important deviations from typical MOGAD presentations and underscores the need for early recognition of steroid-refractory disease. The deviations from typical MOGAD presentations include poor initial response to steroids, whereas most cases show rapid steroid responsiveness. In addition, the paediatric onset of disease with aggressive course and need for second-line immunotherapy is less commonly reported in classic MOGAD. The paediatric case described herein diverges from the classical phenotype, emphasising diagnostic and therapeutic challenges.

Keywords: Autoimmune disease of nervous system, Corticosteroids, Demyelinating disease, Immunoglobulin G, Immunotherapy, Myelin oligodendrocyte glycoprotein

CASE REPORT

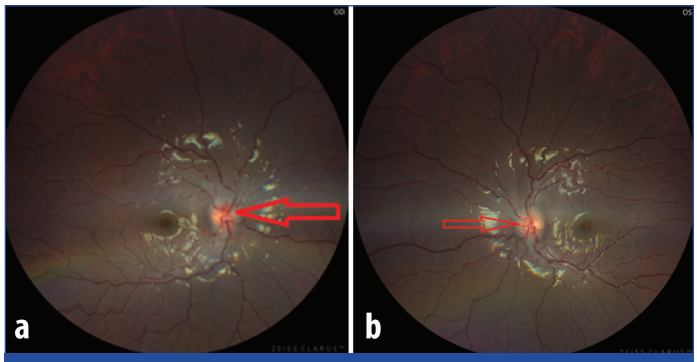
A 13-year-old girl presented with acute, progressive bilateral vision loss of 20 days' duration. This was preceded by generalised tonic clonic seizures one week prior to vision loss, followed by episodes of headache over temporal and occipital region. There was no history of fever, trauma, or similar episodes in the past. She was born of a 3rd degree consanguineous marriage with uneventful perinatal and postnatal period.

On examination, best-corrected visual acuity was Counting Fingers (CF) at one meter in both eyes. Colour vision was markedly reduced (5/17 in the right eye and 6/17 in the left eye on Ishihara testing). Afferent pupillary defect was present in both eyes. Anterior segment examination was unremarkable. Fundus examination revealed bilateral hyperemic optic disc swelling with blurred margins, grade 1 papilloedema with Frisen scale [Table/Fig-1a,b] [1].

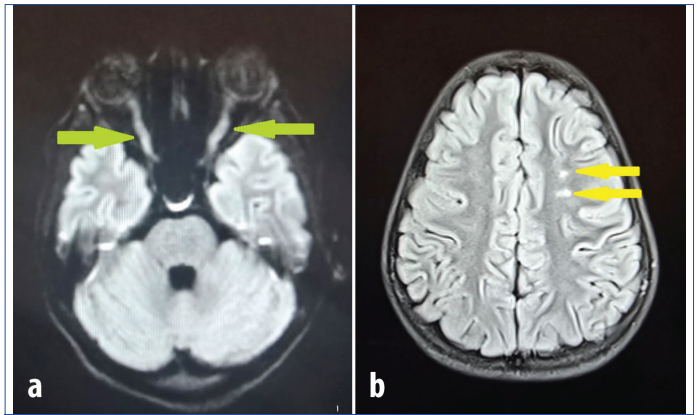
Given the combination of acute bilateral visual loss and neurological symptoms, an inflammatory optic neuropathy with central nervous system involvement was suspected. Cerebrospinal fluid analysis showed normal opening pressure (7 to 10 cm H_2O) mildly elevated protein, 55 mg/dL (Normal: < 50 mg/dL), lymphocytic pleocytosis (20 cells/mm^3), and sterile cultures. Magnetic Resonance Imaging (MRI) of the brain and orbits with contrast demonstrated bilateral swollen and tortuous optic nerves with enhancement, longitudinally [Table/Fig-2a], along with lesions at the grey-white matter junction [Table/Fig-2b]. Magnetic Resonance (MR) venography was normal.

Differentials for present case included MOGAD, Neuromyelitis Optica Spectrum Disorder (NMOSD), Multiple sclerosis, Idiopathic intracranial hypertension and infectious or auto-immune encephalitis. Serological testing revealed positive serum MOG-IgG antibodies, while aquaporin-4 antibodies were negative, confirming the diagnosis of MOGAD.

The patient was treated with intravenous methylprednisolone at 30 mg/kg/day for five days. Despite completion of therapy, visual improvement was minimal (CF 1 mt in both eyes). In view of persistent severe visual impairment, IVIg was administered at a



[Table/Fig-1]: a) Right eye colour fundus picture showing blurring of disc margin as pointed by red arrow; b) Left eye colour fundus picture showing blurring of disc margin as pointed by red arrow.



[Table/Fig-2]: a) MRI brain in axial section at the level of middle cerebellar peduncle showing swollen and tortuous optic nerves as shown by the solid green arrows; b) MRI brain in axial section at the level of high convexity showing lesions at the gray white junction interface as pointed out by the solid yellow arrows.

dose of 2 g/kg over five days, followed by oral prednisolone (1 mg/kg/day) with gradual tapering.

Within five days of IVIg therapy, visual acuity improved dramatically to 6/9 in the right eye and 6/6 in the left eye. Optic disc oedema showed significant resolution on follow-up.

DISCUSSION

The present case illustrates several clinically important features that are not typically emphasised in classic descriptions of Paediatric MOGAD. Although bilateral optic neuritis with disc oedema is well recognised, severe steroid-refractory visual loss remains under-reported despite its implications for irreversible optic nerve damage if treatment escalation is delayed [2-4]. Maran JJ et al., in their case report have highlighted the presentation of MOGAD with isolated intracranial hypertension and bilateral optic disc oedema resolving with intravenous methylprednisolone [5]. In present patient, meaningful recovery occurred only after IVIg administration.

The presence of early neurological symptoms, including epileptic episodes and cortical MRI lesions, highlights that MOGAD may present with combined optic neuritis and encephalitic features rather than isolated visual involvement [2,3,6]. Additionally, the papilledema-like fundus appearance can be misleading for headache associated with other entities and may delay consideration of inflammatory optic neuropathy.

The use of IVIg has emerged as an effective second-line therapy in steroid-resistant MOGAD by neutralising pathogenic antibodies and modulating immune responses [2,7]. In a case report by Patel NA et al., improvement in cognitive function in a patient with MOGAD has been reported after the use of IVIg [8]. Early recognition of inadequate steroid response is therefore critical, particularly in children. In a study by da Klein da Costa B et al., it was reported that almost 66% of paediatric patients with MOGAD, showed improvement with the use of IVIg [7].

Patient experienced loss of vision in both eyes, which was frightening and did not improve despite receiving high dose steroid treatment. Although the delayed diagnosis of MOG antibody-associated disease was challenging for patient and her family, understanding the cause of her illness helped guide further care and gave us hope for recovery.

CONCLUSION(S)

Paediatric MOGAD may present with atypical features including early neurological symptoms, papilledema-like optic disc oedema, and poor steroid responsiveness. The present case emphasises the importance of prompt antibody testing and early escalation to IVIg in steroid-refractory optic neuritis to achieve optimal visual outcomes.

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