Original Research Paper

Volume - 15 | Issue - 03 | March - 2025 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Neurology

AN INTERESTING CASE OF PEDIATRIC OPTIC NEURITIS

Dr Shraddha Kedlaya K*	(MBBS, DNB General Medicine, DM Neurology). Neurologist, Government Medical College And Hospital Krishnagiri *Corresponding Author
Professor Dr. P Chandrasekaran	(MBBS,MD General Medicine, DM Neurology) Medical Superintendent, Government Medical College And Hospital Krishnagiri
Dr. M. Gayathri	(MBBS, MD General Medicine) Assistant Professor, Department Of General Medicine, Government Medical College And Hospital Krishnagiri.
Dr. M Rajkumar	(MBBS, MDRD) Assistant professor, Department Of Radiology, Government Medical College And Hospital Krishnagiri.

ABSTRACT Optic neuritis (ON) is a rare , uncommon condition in the paediatric population and usually underlies a viral cause. In comparison to adult population, paediatric optic neuritis (PON) occurs usually after acute febrile illness and has bilateral optic nerve involvement .Visual loss occurs as prominent symptom .Ocular pain is less common. It has relatively good visual prognosis with near complete visual recovery. Mean age of occurrence is 10 years with female population more affected than male.[1]It has reported incidence of 0.2 per 100,000 children .Children with ON are seropositive for either AQP-4(Aquaporin 4 or MOG Ab (anti-myelin-oligodendrocyte glycoprotein)[2] We hereby report a case of bilateral optic neuritis in 10 year old boy who presented to our institute with sudden onset of headache , retro orbital pain ,bilateral defective vision .

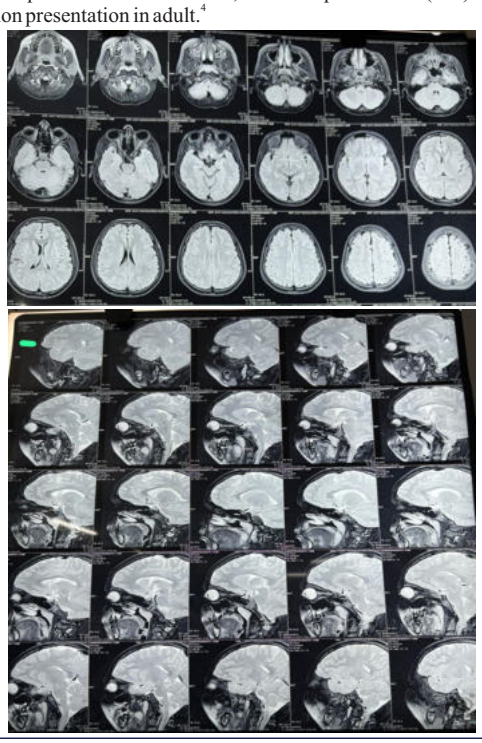
KEYWORDS : Optic neuritis, MOGAD, Demyelinating disease, steroids, IVIG

INTRODUCTION

Optic neuritis is defined as inflammation of optic nerve and has varied etiology including infectious cause, granulomatous ,demyelinating ,autoimmune disease, paraneoplastic disorders.

In comparison to adult population, optic neuritis is rare in pediatric population and it presents as bilateral disease with disc swelling and has good visual outcomes. The mean age of occurrence is 10.6 years with female predominance.[3]

Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) is a demyelinating disorder of the central nervous system (CNS) with IgG autoantibodies targeting MOG with biphasic distribution of the age of onset, with median age of onset overall being 20–30 years. Acute disseminated encephalomyelitis (ADEM) is most common presentation in children ,whereas optic neuritis(ON) is most common presentation in adult.[4]



Case Report

13 year old boy was admitted on 20/12/24 in our institution with sudden onset of headache with retro orbital pain since 2 weeks. Retroorbital pain used to increase on ocular movements.10 days later, he also developed sudden onset of blurring of vision which progressed to loss of vision over 3 days. There was no history of fever,giddiness,diplopia,seizure,altered sensorium, limb weakness ,unsteadiness while walking, sensory complaints. On examination ,his vitals were normal .On neurological examination, his visual acuity was only presence of perception of light. Colour vision was defective. Ophthalmoscopic examination showed hyperemic disc. There was no RAPD .There were no other cranial nerve deficit, cognitive , motor, sensory or cerebellar deficit .With clinical suspicion of optic neuritis ,he was started on pulse steroid therapy. MRI(Magnetic Resonance Imaging) brain with orbital cuts with contrast study showed bilateral optic neuritis. Brain parenchyma and optic chiasma were normal. MRI whole spine screening contrast study was normal. Complete blood count, Renal and hepatic function test, electrolytes, peripheral smear, T3 T4 TSH were normal. CRP(C reactive protein) was high(46mg/dl).VDRL ,HIV serology, Hepatitis markers were negative .CSF analysis was normal .Anti MOG (Myelin oligodendrocyte glycoprotein)antibody was strongly positive .Serum aquaporin 4 ,ANA profile were negative. 5 days of pulse intravenous methylprednisolone(30 mg/kg body weight) was given and his visual acquity improved to 6/6.He was discharged on oral steroids 1mg/kg body weight and gradual tapering of steroids was done. He is on low dose of steroids and symptomatically better.

DISCUSSION

Optic neuritis is defined as inflammation of optic nerve and has varied etiology including infectious cause, granulomatous ,demyelinating ,autoimmune disease, paraneoplastic disorders.

In comparison to adult population, optic neuritis is rare in pediatric population and it presents as bilateral disease with disc swelling and has good visual outcomes. The mean age of occurrence is 10.6 years with female predominance. It is usually occurs after febrile illness and periocular pain is less common. Whereas in our case there was no prior febrile illness and onset of disease was with headache and retro orbital pain.[3]

Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) is a demyelinating disorder of the central nervous system (CNS) with IgG autoantibodies targeting MOG. It has biphasic distribution of the age of onset, peaking in children aged 5–10 years as well as in adults aged 20–45 years with median age of onset overall being 20–30 years. Acute disseminated encephalomyelitis (ADEM) is

most common presentation in children ,whereas optic neuritis(ON) is most common presentation in adult. MOGAD is an oligodendr ogliopathy. The trigger for production of antibody in MOGAD is unknown ,possible proposed mechanism is break in immune tolerance causing influx of inflammatory cells from peripheral circulatory system to central nervous system.[4]

Almost one-half of children with MOGAD present with ADEM and one-quarter with ON. In children 11 years and older, ON is more frequent and ADEM is less frequent .It has been observed that most common initial phenotype in paediatric MOGAD is acute disseminated encephalomyelitis, especially among children younger than five years, followed by optic neuritis (ON) and/or transverse myelitis. Approximately one- quarter of children with MOGAD have at least one relapse that commonly occurs within three years of onset of disease and often includes ON, even if ON was not present at onset of disease . Relapsing pattern is less common in children compared with adults with majority of first relapses occur within the first two to three years of diagnosis .Clinical presentations at relapse have varied presentation which is highly relevant in children with MOGAD as this is rarely observed in other paediatric demyelinating disorders such as NMOSD and non-MOG-Ab associated ON and TM. Studies for assessment of predictors of relapse have remained inconclusive .No associations were found between relapsing nature of MOGAD and age ,sex, EDSS at onset ,initial MOG-Ab titre, presence of IgA or IgM MOG-Ab ,cerebral spinal fluid pleocytosis or increased protein concentration ,or location of MRI lesions.

Few studies have reported a strong association between persistent MOG-Ab and relapse.[5]

In majority of studies favourable clinical outcomes were seen in children with MOGAD, however minority of children experienced a more severe course causing neurologic sequelae.

MRI of optic nerves with fat suppression technique and contrast is the investigation of choice. It shows Longitudinally extensive optic neuritis where more than 50% of the pre- chiasmal optic nerve length is involved .Optic chiasmal involvement, when it occurs in MOG-ON, is more likely to be part of longitudinally extensive disease extending up to the chiasm, in comparison to AQP4-ON where isolated chiasmal involvement can occur. Optic peri neuritis is seen in MRI where in there is circumferential, 'tram-track' enhancement of the optic nerve sheath, which may extend into the surrounding orbital fat.

CSF Analysis is contributory to diagnosis ,but is not sensitive or specific. CSF pleocytosis with mononuclear predominance, elevated CSF protein are seen in MOGAD, more commonly in myelitis as presentation when compared to optic neuritis. Detection of serum MOG IgG using a live cell-based assay is sensitive and specific test for diagnosis of MOGAD.[4]

The majority of patients with MOGAD are treated with intravenous steroids (IV methylprednisolone IVMP) during acute attack/relapse . Dose of IVMP is 20-30 mg/kg/day for 3-5 days. Treatment should be escalated to IVIG or PLEX, in case of insufficient response to IVMP, since treatment failure can lead to severe disability.

Intravenous immunoglobulin (IVIG) in the acute phase after or in addition to IVMP has been used and is associated with good recovery in MOGAD. IVIG is usually administered with total dosage of 1–2 g/kg over a course of 1–5 days .PLEX as an established treatment escalation, has been done during the acute phase in adult and paediatric MOGAD patients.

Relapse rates are reported as 30–60% in MOGAD .There is a proportion of patients who have a monophasic illness . Role of corticosteroid-sparing maintenance immunosuppression at ON onset to prevent relapse is still unclear.[6]

In majority of studies, it was found that relapsed patients had a longer interval from onset to diagnosis, higher MOG antibody titre at onset, longer duration of persisting MOG positivity. The most relapsed attacks occurred in patients with persistent MOG antibody positivity. It was also observed that compared with patients without sequelae, patients with sequelae also had higher MOG antibody titer at onset, longer duration of persisting MOG antibody positive, and higher relapse rate.

Hence it is recommended to slowly taper and maintain low dose of oral prednisolone for six months after the initial attack, especially in MOG antibody-positive patients .Mycophenolate mofetil, monthly IVIG, and maintenance of a low dose of oral prednisone can decrease the relapse frequency in most patients with MOGAD.[7]

REFERENCES

1). Pérez-Cambrodí RJ, Gómez-Hurtado Cubillana A, Merino-Suárez ML, Piñero-Llorens DP, Laria-Ochaita C. Optic neuritis in paediatric population: a review in current tendencies of diagnosis and management. J Optom . 2014 Jul-Sep;7(3):125-30.

2) Nair A, Sankhyan N, Sukhija J, Saini AG, Vyas S, Suthar R, Sahu JK, Rawat A. Clinical outcomes and Anti-MOG antibodies in paediatric optic neuritis: A prospective observational study. Eur J Paediatr Neurol. 2024 Mar;49:1-5

3) Kowsalya A, Ramalingam U, Chaudhary S, Kumar M. Clinical features and visual outcomes of paediatric optic neuritis in the Indian population: A prospective study. Indian J Ophthalmol 2023;71:637-42.

4) Niroshan Jeyakumar, Magdalena Lerch, Russell C. Dale and Sudarshini Ramanathan. Eye (2024) 38:2289–2301

5) J. D. Santoro et al. Attack phenotypes and disease course in paediatric MOGAD. Annals of Clinical and Translational Neurology 2023; 10(5): 672–685

6. Bruijstens et al.E.U. paediatric MOG consortium consensus: Part 5 – Treatment of paediatric myelin oligodendrocyte glycoprotein antibody-associated disorders.European Journal of Paediatric Neurology. Volume 29, November 2020

7) Li X et al. Paediatric myelin oligodendrocyte glycoprotein antibody-associated disease in southern China: analysis of 93 cases. Front Immunol. 2023 Jun 2