ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 01 | January - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A CASE REPORT OF INTERESTING NEUROMYELITIS OPTICA

Ophthalmology

Dr.N.V.N . Prasanna Bharathi* M.S Ophthalmology, former Resident of OMC, Hyderabad. *Correspondig Author

Dr.Tanveer Fatima M.S Ophthalmology, former Resident of OMC, Hyderabad.

Dr.Santhosh Kumar Kraleti Public health specialist ,member, NPCB.

ABSTRACT

Back ground: NMO is an autoimmune disorder of the Central nervous system, dominated by inflammation of the Optic nerve and the Spinal cord. Though recent development of different modalities for the treatment of autoimmune disorders, Neuromyelitis Optica recurrent attacks well respond to intravenous methyl prednisolone.

Case presentation: Here we report a case of neuromyelitis optica ,in which a young male patient had a first presentation as Bilateral sudden loss of vision due to optic neuritis at the age of 16 years and second remission at the age of 32 years without any spinal cord effected symptoms and recovered well with only Intravenous methyl prednisolone who is Anti-MOG (Myelin oligodendrocyte glycoprotein) Antibody positive and Anti-Aquaporin-4 Antibody negative , a Monophasic NMO.

Conclusion: Treating Neurologists and Ophthalmologists using Intra venous methyl prednisolone in acute attack with all precautions can save vision and reduce incidence of Optic atrophy after acute attacks.

KEYWORDS

Methyl prednisolone, Neuromyelitis optica, Devic's disease,Loss of vision,Recovery.

INTRODUCTION:

Devic's disease is also called neuromyelitis optica or NMO; it is a condition often confused with multiple sclerosis. Devic's disease is often characterized by immune attacks on the optic nerve and the spinal cord. Patients may experience these attacks at the same time or at different times. They may also have just problems with the optic nerves alone or just problems with the spinal cord alone and still have Devic's disease. Recent research has suggested that Devic's disease is probably a different disease in which there is a specific immune attack on a molecule known as aquaporin 4. Our understanding of Devic's disease is changing quickly at the present time due to new research in this important disorder like NMO with aquaporin-4 antibodies, NMO without aquaporin-4 antibodies (or not tested).

Early in the course of the disease, it may be difficult to distinguish between NMO and multiple sclerosis because both may cause optic neuritis and myelitis as symptoms. However, the optic neuritis and myelitis tend to be more severe in NMO; the brain MRI is more commonly normal, and the spinal fluid analysis does not usually show oligo clonal bands in NMO, which are features that help to distinguish it from MS.

In most cases of NMO, the initial symptoms of vision loss or paralysis improve with standard treatment with high dose corticosteroids, and partial recovery of vision, motor, sensory, or bladder function occurs. However, in recurring cases, NMO frequently causes significant permanent disturbances of vision and/or spinal cord function leading to blindness or impaired mobility.

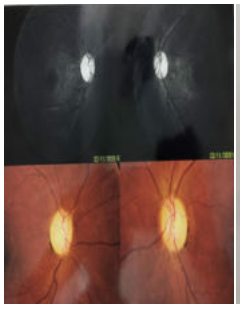
Some patients with NMO have only recurrent myelitis or only recurrent optic neuritis. NMO occurs in individuals of all races. The prevalence of NMO is approximately 1-10 per 100,000 individuals. For acute attacks, the standard treatment is high-dose intravenous corticosteroids, typically methylprednisolone. Plasma exchange may be effective in patients who experience acute severe attacks that do not respond to intravenous corticosteroids.

In 2020, Uplinza (inebilizumab-cdon) and Enspryng (satralizumab-mwge) were approved for the treatment of NMOSD in adult patients with the same AQP4 for long-term suppression of the disease, a variety of immunosuppressive drugs like Corticosteroids, azathioprine, mycophenolate mofetil and rituximab .

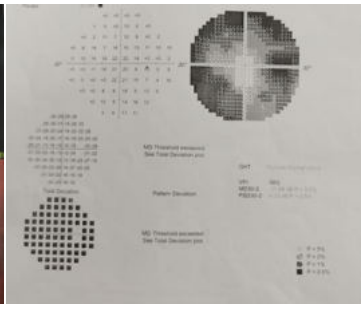
Case presentation:

We report a case of 32yrs old male presented with the complaint of mild pain in the left eye followed by sudden loss of vision in both the eyes, left eye more than right eye without any comorbidities. Patient had similar complaints at the age of 16 years and was treated with 3 doses of Intravenous methyl prednisolone and his vision recovered completely. He was not thoroughly investigated in the past.

On examination of the patient vision reduced to perception of light in the left eye and right eye has 20/30.There is reduced color vision and contrast sensitivity in left eye. Relative afferent pupillary defect is present in left eye. Left eye was diagnosed as optic neuritis on fundus examination.

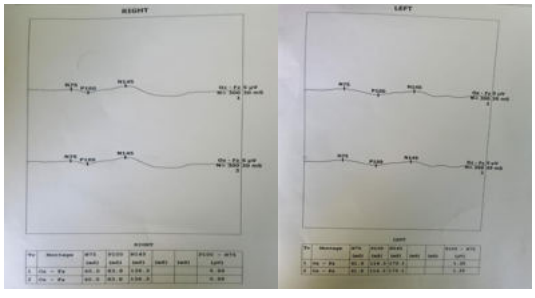


Picture: 1 Fundus of RE and LE



Picture: 2 Visual field of LE

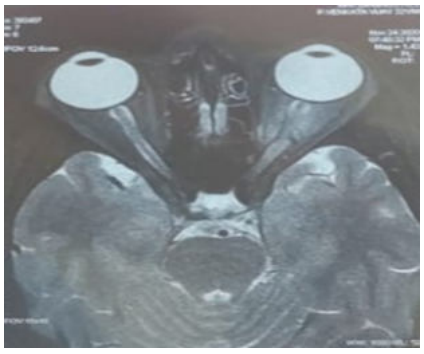
Humphrey Visual field testing showed left eye diffuse visual field loss with complete scotoma. Visual evoked potential showed prolonged latency of p100 and N145 in left eye.



Picture 3: VEP of RE and LE, Prolongation of p100 in LE

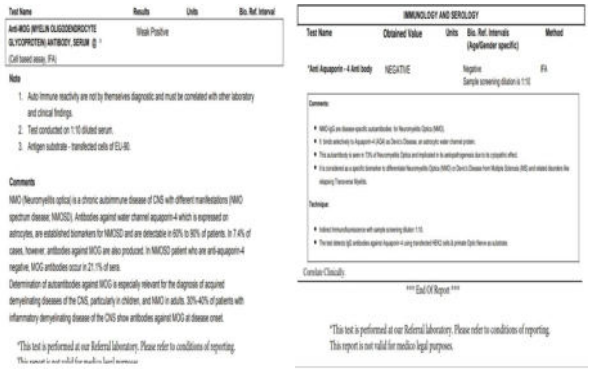
He consulted neurologist and he advised MRI of the brain with and without contrast. MRI of the brain is normal. MRI with contrast showed left optic nerve is mildly hyper intense with effacement of

perioptic space and diffusion restriction. No significant post contrast enhancement of optic nerve was seen.



Picture4: MRI of Optic nerve with contrast

Patient was advised serum Anti-MOG antibody and Anti aquaporin 4 antibodies to differentiate Multiple sclerosis and Neuromyelitis Optica



Test Name	Results	Units	Bio. Ref. Interval
Anti-MOG (MYELIN OLIGODENDROCYTE GLYCOPROTEIN) ANTIBODY, SERUM @ *	Weak Positive		
Cell based assay, IFA			

Note

1. Auto immune reactivity are not by themselves diagnostic and must be correlated with other laboratory and clinical findings.
2. Test conducted on 1:10 diluted serum.
3. Antigen substrate - transfected cells of EU-90.

Comments

MAO (Neuromyelitis optica) is a chronic autoimmune disease of CNS with different manifestations (NMO spectrum disease; NMOSD). Antibodies against water channel aquaporin-4 which is expressed on astrocytes, are established biomarkers for NMOSD and are detectable in 60% to 80% of patients. In 7.4% of cases, however, antibodies against MOG are also produced. In NMOSD patient who are anti-aquaporin-4 negative, MOG antibodies occur in 21.1% of sera.
Determination of autoantibodies against MOG is especially relevant for the diagnosis of acquired demyelinating diseases of the CNS, particularly in children, and NMO in adults. 30%-40% of patients with inflammatory demyelinating disease of the CNS show antibodies against MOG at disease onset.

*This test is performed at our Referral laboratory. Please refer to conditions of reporting.

IMMUNOLOGY AND SEROLOGY

Test Name	Obtained Value	Units	Bio. Ref. Intervals (Age/Gender specific)	Method
*Anti Aquaporin - 4 Anti body	NEGATIVE		Negative Sample screening dilution is 1:10	IFA

Comments

- NMO IgG are disease-specific autoantibodies for Neuromyelitis Optica (NMO).
- It binds selectively to Aquaporin-4 (AQ4) as Devic's Disease, an astrocytic water channel protein.
- This autoantibody is seen in 73% of Neuromyelitis Optica and implicated in its astrocytopathogenesis due to its cytopathic effect.
- It is considered as a specific biomarker to differentiate Neuromyelitis Optica (NMO) or Devic's Disease from Multiple Sclerosis (MS) and related disorders like relapsing Transverse Myelitis.

Technique

- Indirect Immunofluorescence with sample screening dilution 1:10.
- The test detects IgG antibodies against Aquaporin-4 using transfected HEK cells & primate Optic Nerve as substrate.

Correlate Clinically

*** End Of Report ***

*This test is performed at our Referral laboratory. Please refer to conditions of reporting.
This report is not valid for medico legal purposes.

Picture5: Lab reports of Anti-MOG antibodies and Anti aquaporin - 4 antibodies

Anti-MOG antibodies are positive in Monophasic NMOSD and Anti aquaporin 4 antibodies are absent in Multiple sclerosis. In this patient Serum sample confirmed Anti-MOG (MYELIN OLIGODENDROCYTE GLYCOPROTEIN) Antibody, suggestive of Neuromyelitis Optica (NMO). After 16 years of similar complaints, remission of the disease occurred at the age of 32 years. He was treated with 5 doses of Intravenous methyl prednisolone one gram in 100ml of Normal saline by neurologist. After that patient was shifted to oral steroids 1mg /kg and tapered in 2 months as no other complaints were present.He responded well to the treatment and vision recovered completely in right eye and left eye near normal with improved color vision after 2 weeks of the treatment. Patient was advised rituximab weekly 1 injection for 4 weeks to prevent further remission of NMO.

CONCLUSION:

Use of Intravenous methyl prednisolone in acute attacks of optic neuritis before differentiating Multiple sclerosis and Devic's disease can save vision and prevent complications. As Neuromyelitis Optica is seen in young individuals right time of administration of intravenous steroids prevents permanent damage to the optic nerve in acute attacks and recurrences may occur in future.

Declarations: Informed consent of the patient has taken.
No financial interest

REFERENCES:

1. Kim W, Kim SH, Kim HJ. New insights into neuromyelitis optica. J Clin Neurol. 2011;7(3):115–127. doi: 10.3988/jcn.2011.7.3.115. [PMC free article] [PubMed] [CrossRef] [Google Scholar]
2. Jacob A, Weinshenker BG, Violich I, et al. Treatment of neuromyelitis optica with rituximab: retrospective analysis of 25 patients. Archives of Neurology. 2008; 65 (11):1443-8.
3. Shimizu Y, Yokoyama K, Misu T, et al. Development of extensive brain lesions following interferon beta therapy in relapsing neuromyelitis optica and longitudinally extensive myelitis. Journal of Neurology. 2008;255(2):305-7.
4. Pittock SJ, Lennon VA, de Seze J, et al. Neuromyelitis Optica and non-organ-specific Autoimmunity. Archives of Neurology. 2008;65(1):78-83.
5. Keegan BM, Pittock SJ, Lennon VA. Autoimmune myelopathy associated with collapsin response-mediator protein-5 immunoglobulin G. Ann Neurol. 2008; 63 (4):531-4.
6. Weinshenker BG and Wingerchuk DM. Neuromyelitis Spectrum Disorders. Mayo Clin Proc. 2017;92(4):663-679. https://www.mayoclinicproceedings.org/article/S0025-6196(16)30849-7/pdf?code=jmcp-site
7. Jarius et al. MOG-IgG in NMO and related disorders: a multicenter study of 50 patients. Part 1: Frequency, syndrome specificity, influence of disease activity, long-term course, association with AQP4-IgG, and origin Journal of Neuroinflammation 2016; 13:279.
8. Jarius et al. MOG-IgG in NMO and related disorders: a multicenter study of 50 patients. Part 2: Epidemiology, clinical presentation, radiological and laboratory features, treatment responses, and long-term outcome Journal of Neuroinflammation 2016; 13:280.
9. Jarius et al. MOG-IgG in NMO and related disorders: a multicenter study of 50 patients. Part 3: Brainstem involvement – frequency, presentation and outcome Journal of Neuroinflammation 2016; 3:281.
10. Davide Mangioni, Alessandro Soria, Laura Brighina, Alessandra Bandera, Carlo Ferrarese, Andrea Gori :A case of classic neuromyelitis optica (Devic's syndrome) triggered by pegylated-interferon α BMC Pharmacol Toxicol. 2014; 15: 56. Published online 2014 Sep 30. doi: 10.1186/2050-6511-15-56.