ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 08 | August - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

ATYPICAL CASE OF MYELIN OLOGODENDROCYTE GLYCOPROTEIN ANTIBODY–POSITIVE BILATERAL OPTIC NEURITIS

Ophthalmology

Dr. Shrinkhal*	MBBS, M.S., DNB, MNAMS, Department of Ophthalmology, AIIMS, Raebareli *Corresponding Author
Dr. Pragati Garg	Department of Ophthalmology, AIIMS, Raebareli
Dr. Swarastra Prakash Singh	Department of Ophthalmology, AIIMS, Raebareli
Dr. Ruchi Shukla	Department of Ophthalmology, AIIMS, Raebareli
Dr. Anirudh Mukherjee	Department of General Medicine, AIIMS, Raebareli
Dr. Ashutosh Kumar Mishra	Department of Neurology, AIIMS, Raebareli

ABSTRACT

A 62 years old male presented with sudden diminution of vision in his both eyes (right eye more than left eye). Anterior segment examination revealed bilateral immature senile cataract and there was relative afferent pupillary defect (RAPD) grade 3 in the right eye and sluggish reaction in the left eye. Posterior segment examination revealed bilateral disc edema (right>left), bilateral posterior vitreous detachment (PVD) and sub-retinal fluid (SRF) at the macula in the right eye. After thorough investigations, patient was diagnosed as anti-myelin-oligodendrocyte glycoprotein (MOG) antibody positive bilateral atypical optic neuritis (right eye>left eye) with presence of SRF in right eye at the macula without transverse myelitis. Patient was given steroids, but did not show significant improvement. This case demonstrates a very unique atypical optic neuritis with several diagnostic dilemmas and focuses on strong clinical acumen to manage these cases.

KEYWORDS

Atypical optic neuritis, Myelin oligodendrocyte glycoprotein-IgG associated disorder (MOG IgG), bilateral disc edema

BACKGROUND:

Optic neuritis (ON) is an inflammatory condition of the optic nerve, which presents as sudden diminution of vision in the affected eye. A typical case of optic neuritis is usually easy to diagnose and manage. Optic neuritis with associated demyelinating disease; Multiple sclerosis (MS) is classified as "typical ON" which usually presents at an early age. Atypical ON can of be infectious origin, neuromyelitis optica (NMO) or NMO spectrum disorder (NMOSD) or Myelin oligodendrocyte glycoprotein-IgG associated disorder (MOG IgG). Such types of ON are generally seen in Asia and vary from typical ON in several aspects as the management approach, and final neurological prognosis. It is important to diagnose early these atypical cases for aggressive approach towards management. Myelin oligodendrocyte glycoprotein (MOG) is a transmembrane protein, located on the surface of oligodendrocytes. MOG antibody is a biomarker of MOG-associated disorder. Patients with MOG positive optic neuritis have optic disc oedema, which is usually bilateral, and often recurrent. Optic neuritis associated with MOG-IgG seropositivity can occur with or without other neurologic symptoms and is recurrent in most cases. [1,2] Vision loss is usually profound, but recovery is usually better than seen with AQP4-IgG optic neuritis. JJ Chen *et al*. in their study found that only 6–10 percent of patients with MOG-IgG positive optic neuritis had a final visual result worse than 20/60 compared to 50 percent of patients with AQP4-IgG optic neuritis [3]. Our patient is an example of anti-myelin-oligodendrocyte glycoprotein (MOG) antibody positive bilateral atypical optic neuritis with poor visual outcome.

Case Summary:

A 62 years old male, vegetarian by diet, presented with chief complaint of sudden, painless diminution of vision in his right eye for last 10 days and in his left eye for last 3 days. It was not associated with any fever, headache, trauma, weakness, facial or scalp pain and tenderness. There was no associated lower back pain, weakness in the arms or legs, and the bowel habits were normal. He was non-diabetic, non-hypertensive with no history of alcohol and tobacco intake or any long-term drug intake. On examination, his visual acuity was hand movement close to face (HMCF) with accurate projection of rays (PR) in all four quadrants in the right eye and finger count at one meter with accurate PR in all four quadrants in his left eye. His intra-ocular pressure was 16 mm of Hg in both the eyes on applanation tonometry. His extra ocular movements were also non-painful, full and free in all the direction in both the eyes. Anterior segment examination revealed bilateral early stage immature senile cataract and there was relative afferent pupillary defect (RAPD) grade 3 in the right eye and sluggish reaction in the left eye. Color vision was grossly abnormal as the patient was only able to read introductory plate on Ishihara chart. Posterior segment examination revealed bilateral disc edema (right>left) with dull foveal reflex in the right eye [Figure 1A blue arrow].

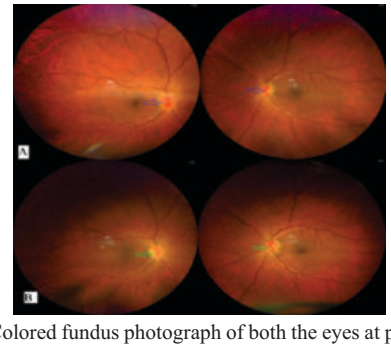

Figure 1: Colored fundus photograph of both the eyes at presentation showing bilateral disc edema (1A blue arrow) and after treatment showing resolving disc edema with some pallor (1B green arrow).

Optical coherence tomography (OCT) revealed presence of bilateral disc edema (right eye >left eye), bilateral posterior vitreous detachment (PVD) and sub-retinal fluid (SRF) of around 130 micron at the macula in the right eye [Figure 2A].

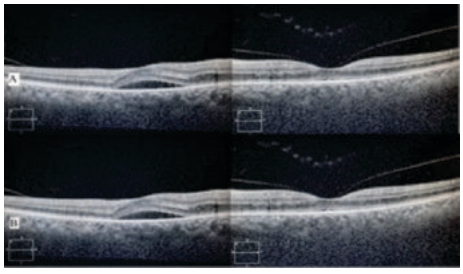

Figure 2: Optical coherence tomography (OCT) of macula of the eyes

at presentation (2A) and after treatment (2B), showing presence of bilateral posterior vitreous detachment (PVD) and sub-retinal fluid (SRF) at the macula in the right eye

Magnetic resonance imaging (MRI) of the brain, orbit and spine revealed bilateral optic neuritits [Figure 3], with no cerebral and spinal involvement [Figure 4].

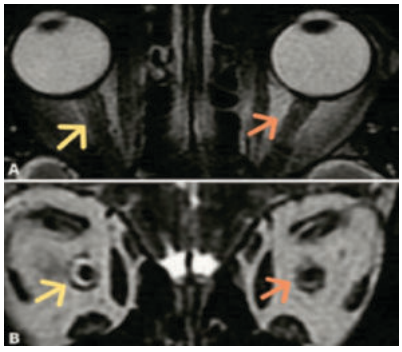

Figure 3: MRI Orbit images: T2 axial(a) and T2 coronal images show mild hyperintensity in bilateral optic nerves and sheath in the retrobulbar segment. (Yellow arrow: Right Optic nerve. Orange arrow: Left Optic nerve.)

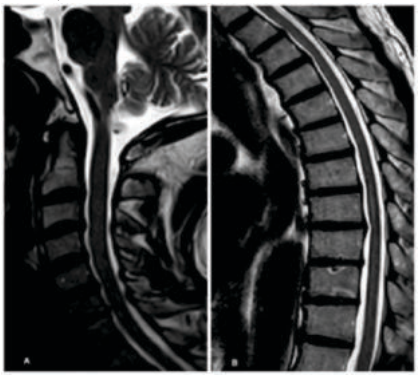

Figure 4: MRI Spine images: T2 sagittal cervical spine(a) and dorsal spine(b) images show normal signal intensity of spinal cord.

On the basis of all clinical, diagnostic and radiological findings, provisional diagnosis of bilateral immature senile cataract with bilateral atypical optic neuritis (right eye >left eye) with presence of SRF in right eye at the macula and without transverse myelitis was made.

Patient was immediately started on high dose intravenous (i.v.) methylprednisolone (1 gm) once daily with strict monitoring of blood sugar and blood pressure. Also he was started on topical nepafenac 0.1% I drop 3 times a day in the right eye, oral pantoprazole 40 mg twice daily, oral acetazolamide 250 mg twice daily, oral multi-vitamin with Vitamin B12 once daily, oral calcium 500 mg and Vitamin D3 500 IU once daily.

Patient was investigated in due course for the causes of optic neuritis. His complete blood count (CBC) revealed leukocytosis (15.4*10^3/microliter) with predominant neutrophilia (96%). Peripheral blood picture (PBP) showed normocytic normochromic picture with neutrophilia. Blood culture report was normal. Procalcitonin was within normal limits (0.105 ng/ml, N= 0.077-0.500 ng/ml). C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were <0.2 mg/dl and 03 mm/hr respectively, i.e. were also within normal limits. His chest X-ray, thyroid profile, viral markers, anti-nuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies (ANCA), rheumatoid factor, urine analysis, interferon gamma assay for tuberculosis, test for syphilis, lyme disease were all normal. His serum vitamin B12 levels were marginally low (169 pg/ml, N=239-931 pg/ml). His anti neuromyelitis optica (NMO) antibody was negative, but anti-myelin-oligodendrocyte glycoprotein (MOG) antibody came positive.

Patient's final diagnosis was **anti myelin-oligodendrocyte glycoprotein (MOG) antibody positive bilateral atypical optic neuritis (right eye>left eye) with presence of SRF in right eye at the macula without transverse myelitis with bilateral immature senile cataract.**

Patient was continued on i.v. methylprednisolone once daily for 3 days along with all other medications but did not show any significant improvement and discharged on oral prednisolone (1mg/kg) for 11 days with sudden taper in 3 days, topical nepafenac 0.1% I drop 3 times a day in the right eye, oral pantoprazole 40 mg twice daily, oral acetazolamide 250 mg twice daily for 14 days, oral multi-vitamin with Vitamin B12 once daily for 30 days, oral calcium 500 mg and Vitamin D3 500 IU once daily for 30 days. After completion of the oral steroids treatment, there was very slight improvement symptomatically in the left eye, however in the right eye, mild disc pallor [Figure 1B and 2B] developed with slight resolution of SRF to 78 microns. His best corrected visual acuity after 3 days of treatment was hand movement close to face (HMCF) with accurate projection of rays (PR) in all four quadrants in the right eye and finger count at three meter with accurate PR in all four quadrants in the left eye.

Patient was explained poor visual prognosis of the disease and informed about next line of management as plasma exchange therapy, immunosuppressive drugs etc.

DISCUSSION:

Optic neuritis is an inflammatory condition of the optic nerve. A typical case of optic neuritis occurs in young age group, usually 20-40 years and presents with sudden diminution of vision in one eye with associated pain on extra-ocular movements. A complete ophthalmic, neurologic and systemic examinations should be done for diagnosis of ON [4, 5].

Our patient was 62 years of age and presented with sequential loss of vision (right eye followed by left eye) within 7 days. The presentation with sudden profound vision loss without any associated nausea, vomiting or any transient visual obscuration, excluded the diagnosis of papilloedema. There was no associated pain on extra-ocular movements. At first, very quick sequential involvement of both the eyes along with absence of pain on extraocular movements gave the impression of anterior ischemic optic neuropathy (AION). Most important was to exclude the probability of giant cell arteritis, which is the most common and dreaded cause of arteritic AION. However, presence of severely affected color vision and absence of peri-papillary retinal haemorrhage excluded the diagnosis of anterior ischemic optic neuropathy (AION) clinically. It was further supported by the findings of normal ESR and CRP values and thrombocytopenia in CBC. Usually, ESR is raised up to 70-100 mm/hour or greater in cases of giant cell arteritis, but can be normal in around 16% of the cases. Also, there is associated thrombocytosis in CBC in giant cell arteritis, however can be normal also. There was also no associated other clinical features suggestive of giant cell arteritis, like pain in any distribution of external carotid artery, except sequential bilateral severe vision loss.

Leukocytosis with predominant neutrophilia gave the initial impression of sepsis. However, absence of fever, normal ESR, CRP and serum procalcitonin and sterile blood culture excluded any infective cause. Other thorough investigations done for general causes of infection also were negative. So, the final clinical and pathological impression for leukocytosis with neutrophilia was steroid induced, and thus was not started on any antibiotics.

Now, the diagnosis was tapered to only atypical optic neuritis. Features initially suggestive of atypical optic neuritis were Asian patient, age greater than 50 years, bilateral involvement and painless full and free extra-ocular movements. Usually, it is associated with long segment optic nerve involvement, extending up to chiasm and tract and associated area posterma syndrome with clinical features like nausea, vomiting, intractable hiccups, and excessive day-time sleepiness (symptomatic narcolepsy). There is usually associated transverse myelitis with MRI findings of contiguous signal abnormality extending up to 3 or more vertebral segments.

However, our patient in spite of positive anti-myelin-oligodendrocyte glycoprotein (MOG) antibody, did not demonstrate any clinical or radiological feature of brainstem or spinal involvement. The bilateral optic nerve enhancement due to bilateral optic neuritis showed only short segment involvement, not even extending up to chiasm.

Our patient did not show significant clinical improvement even after

extensive medical management. So, this case with an aggressive presentation and without any specific common clinical features of atypical optic neuritis, created diagnostic dilemma at several points.

CONCLUSION:

Clinicians should be well aware of the clinical features of atypical optic neuritis and also that it can have a unique and varied presentation, requiring an aggressive approach towards the diagnosis and management.

Decleration Of The Patient Consent:

The authors certify that they have obtained all appropriate patient consent. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial Support And Sponsonship: Nil

Conflict Of Interest: There are no conflicts of interest.

REFERENCES:

1. Jurynczyk M, Messina S, Woodhall MR, et al. Clinical presentation and prognosis in MOG-antibody disease: a UK study. Brain 2017;140(12):3128–3138.
2. Pache F, Zimmermann H, Mikolajczak J, et al. MOG-IgG in NMO and related disorders: a multicenter study of 50 patients. Part 4: Afferent visual system damage after optic neuritis in MOG-IgG-seropositive versus AQP4-IgG-sero-positive patients. J Neuroinflammation 2016;13(1):282.
3. Chen JJ, Pittock SJ, Flanagan EP, Lennon VA, Bhatti MT. Optic neuritis in the era of biomarkers Surv Ophthalmol. 2020;65:12–7.
4. Menon V, Saxena R, Misra R, Phuljhele S. Management of optic neuritis. Indian J Ophthalmol 2011; 59(2): 117-22.
5. Mehrotra A, Jaffery N, Saxena R, Menon V. Comparative evaluation of megadose methylprednisolone with dexamethasone for treatment of primary typical optic neuritis. Indian J Ophthalmol 2007; 55(5): 355-9.