

Neuromyelitis Optica: A Case Report

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Abstract: *Neuromyelitis optica spectrum disorder is an autoimmune disease that causes severe demyelination, especially in the optic nerve and spinal cord with typical clinical manifestations of acute optic neuritis and transverse myelitis. The symptoms can occur simultaneously or separated by a variable period. NMOSD is associated with serum aquaporin antibodies 4 immunoglobulin G (AQP4 - Ig G). Here we report a rare case of neuromyelitisoptica in 27 years old female patient presented with headache and bilateral visual loss. The case emphasizes the existence of neuromyelitisoptica spectrum disorder in clinical settings of the developing world.*

Keywords: neuromyelitisoptica, optic neuritis, transverse myelitis, aquaporin antibodies 4 immunoglobulin G

1. Introduction

Neuromyelitis optica spectrum disorder (NMOSD), formerly known as neuromyelitisoptica (NMO) or Devic's syndrome or Devic's disease, was initially considered as part of multiple sclerosis (MS) because the symptoms were considered overlapping. But now, it is known that the pathophysiology of these two diseases is different [1].

In all populations there is a strong female predominance, with a female - to - male ratio of 3: 1. The mean age of onset is approximately 40 years old, although many cases have been reported in childhood [2].

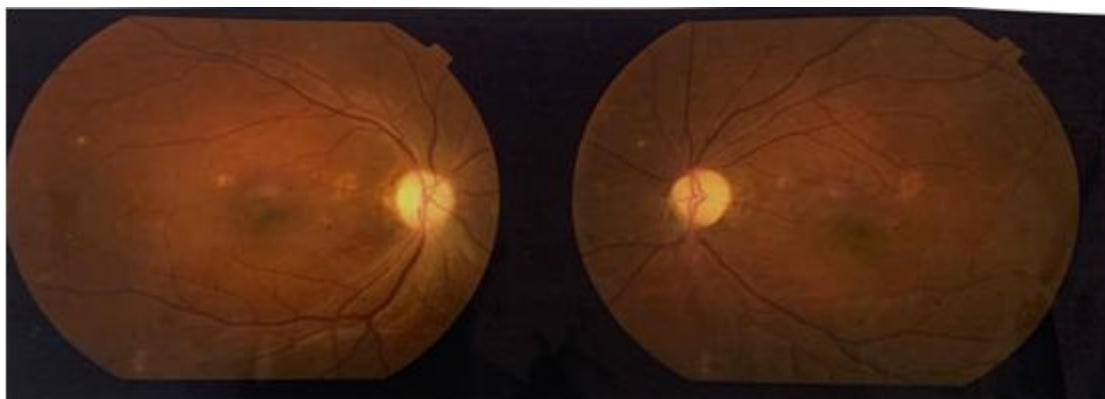
It is important to differentiate NMO from MS early because NMO (especially relapsing NMO) has a more severe morbidity than MS and standard MS modifying therapies may not be effective on NMO [2, 3].

Neuromyelitis optica -immunoglobulin G (NMO - IgG), an IgG auto - antibody binding to the aquaporin - 4 (AQP4) water pump channel localized on the luminal side of blood vessels and astrocytic foot process, has a high sensitivity and specificity to NMO [4, 5]. Thus, NMO - IgG has been incorporated into the updated diagnostic criteria for NMO [3].

2. Case Report

A 27 years old female patient with not known co - morbidities went to the ophthalmology clinic with complain of progressive bilateral visual loss of two months duration associated with sudden in onset, moderate to severe in intensity, throbbing fronto - temporal headache. No history of reddening of eyes, vomiting or neck stiffness. On ophthalmologic examination, by right eye - patient had no perception of light, left eye - patient able to count finger from 2 feet. A fundoscopic examination showed pale disc with disc edema in both eyes, which indicated bilateral optic atrophy. On the basis of this findings they given IV MPS 1gm iv OD for 5 days, transient improvement was noted. The patient was referred to an outpatient medical clinic of SSG hospital for evaluating cause and for further management.

The patient was presenting in our hospital with complain of headache and bilateral visual loss since three months, so for further evaluation admitted in female medical ward, and sent all routine investigation on first day. On the next day evaluated by Neuro - physician, bilateral optic atrophy was detected on fundoscopic examination (figure 1 - 1). There was no history of memory deficit, other cranial nerve palsies, sensory or motor deficit, or bladder or bowel dysfunction. There was normal CSF chemistry and cellularity.



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On examination a conscious and well oriented, with regular heart rate of 80 beats/min, blood pressure of 120/80 mmHg and respiratory rate 16/min. the patient could count fingers at one meters. There was no other cranial nerve palsy. Power in all 4 limbs at all joints and in all range of movements were 5/5. Plantar response were flexor on both sides. Findings on other systems were unremarkable. On investigation, complete blood count, urinalysis and serum liver biochemical and renal function test were within normal limits. Serological tests for HIV, hepatitis B and hepatitis C virus were negative. CSF examination suggestive of normal results. Serum aquaporin - 4 (AQP4) antibody and ANA

were sent, which came positive, but ANA profile was negative.

MRI of brain (P+C) with whole spine screening + Orbit done which revealed –subtle abnormal signal in periventricular white matter of bilateral temporal lobes and lower medulla and cervical medullary junction circumferential on both sides along optic radiation, possibility of Neuromyelitis optica spectrum disorder likely. Focal poorly defined long segmental abnormal signal in right optic nerve extending into right optic chiasm s/o optic neuritis.

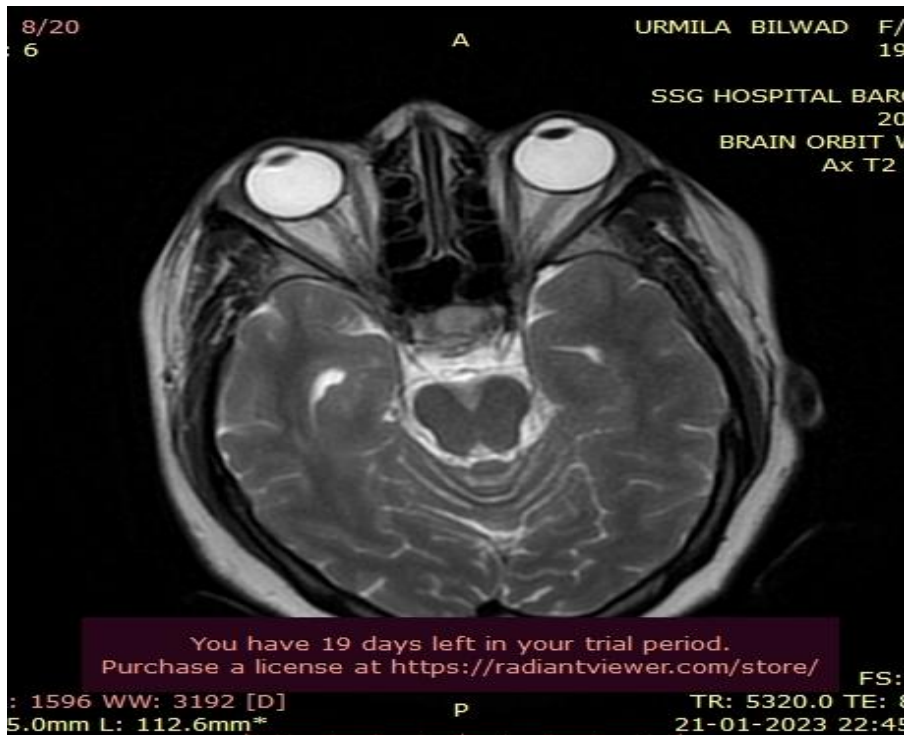


Figure 2: MRI brain with orbit

VEP Done by ophthalmology department s/o - both eye optic neuropathy.

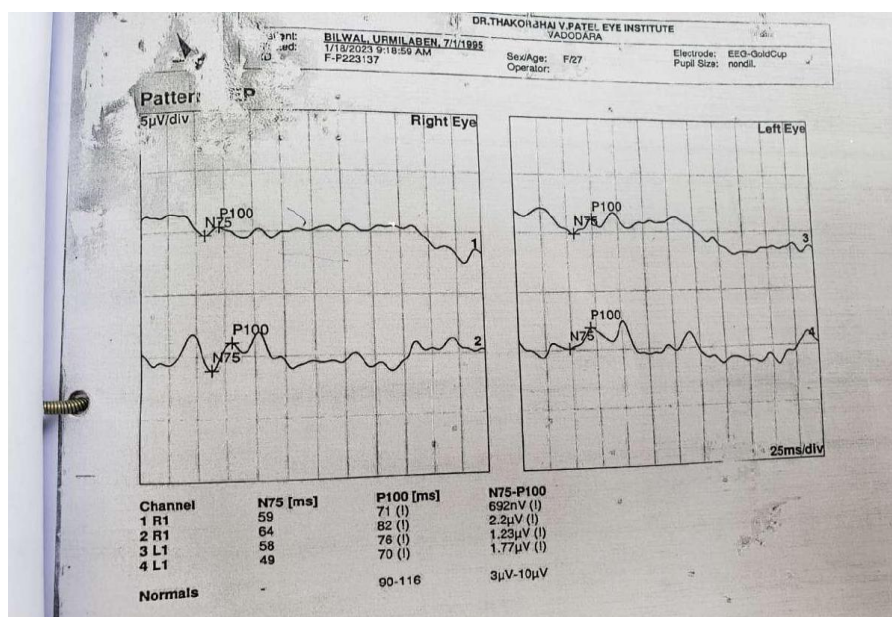


Figure 3: Visual evoked potential report

VEP report s/o - optic neuropathy changes present in both eyes.

A diagnosis of NMO spectrum disorder was made based on the typical imaging findings, serological investigation and clinical feature such as optic neuropathy. The patient was started on injection Methylprednisolone 1 gm iv OD for 5 days then changed to prednisolone 1mg/kg (40mg) PO daily. Five cycles of plasmapheresis was done. The patient was discharged on tab. Azathioprine 50 mg PO twice daily with oral prednisolone.

3. Discussion

NMOSD is an autoimmune disease that causes severe demyelination, especially in the optic nerve and spinal cord with typical clinical manifestations of acute optic neuritis and transverse myelitis. It has been associated with serum AQP4 - IgG [6]. NMOSD is a rare syndrome with less than 1% demyelinating disease and the incidence varies in various countries. In general, the incidence of NMOSD ranges from 0.05 - 4.4 per 100, 000 [1], [6]. It generally occurs in Asian, African and Hispanic descendants. The clinical features of NMOSD are severe recurrent attacks of myelitis and bilateral and unilateral optical neuritis that can occur simultaneously. It is more common to be found in the form of polyphasic (90%) than monophasic (10%) [1]. Serum AQP4 - Ig is detected in 60 - 90% of patients who met NMOSD clinical and radiological criteria.

Cerebrospinal fluid examination in positive NMOSD patients with AQP4 - Ab can be found moderate with normal pleocytosis in about 40% of cases during acute recurrence. Oligoclonal bands (OCB) are usually not found, the intrathecal polyspecific antiviral immune response against - Measles, Rubella and Varicella - Zoster viruses (MRZ reaction) are negative, and increased glial fibrillary acidic protein and neurofilament heavy chain (nfH) are commonly found [7]. In this patient, the cerebrospinal fluid examination gave a normal impression and OCB was negative.

Corticosteroids are the main choice in the acute phase. Intravenous methylprednisolone is administered with a dose of 1 - 1.5 grams in 3 - 5 days [1], [2], [6], [7]. Intravenous dexamethasone at a dose of 5 mg can be also a choice of corticosteroids. Therapeutic plasma exchange (TPE) can be considered if the patient's condition does not improve or neurological symptoms worsen. Therapeutic plasma exchange dosage is carried out by giving 5 - 7 cycles in a period of 2 weeks with a dose of 1 - 1.5 plasma per time TPE. This patient has been treated with a dose of 50 mg azathioprine twice a day to prevent relapse.

The probability of recurrence of disease activity is greater than 90% [7]. Attacks on NMOSD can be very severe, NMOSD can be life - threatening if the lesion extends to the cervical spinal cord and brain stem because it has the potential to cause respiratory failure.

4. Conclusion

The case emphasizes the existence of neuromyelitisoptica spectrum disorder in clinical settings of the developing

world. High index of suspicion of this rare disease is required to avoid delayed diagnosis and treatment.

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