

# CASE REPORT

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## EARLY DIAGNOSIS AND MANAGEMENT OF NEUROMYELITIS OPTICA PREVENTS RELAPSES AND LIMITS VISUAL DISABILITY

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**ABSTRACT:** Neuromyelitis optica (NMO) or Devic's disease was considered as a variant of Multiple sclerosis, with recent advances in investigations and with better understanding of etiopathogenesis, NMO is a distinct immune mediated, largely relapsing, inflammatory, demyelinating disease of central nervous system. NMO most commonly involves the optic nerves and spinal cord. Relapsing and monophasic are two variants of NMO, as relapsing NMO is a rapidly disabling disease, prompt diagnosis of the type from the monophasic, helps in the management as well as decreasing the motor and visual morbidity.

**KEYWORDS:** Neuromyelitis optica, Optic neuritis, Multiple sclerosis, Transverse myelitis, Monophasic, Relapse.

**INTRODUCTION:** Since the description of NMO by Eugene Devic by as early as late 18th century, it remained largely regarded as an unusual or severe variant of multiple sclerosis.<sup>[1]</sup> Neuromyelitis optica (NMO) is an inflammatory, demyelinating, necrotising disease of the central nervous system, with a predilection for optic nerves and spinal cord. Clinical, radiological and immunopathological characteristics distinguish it from multiple sclerosis (MS).<sup>[2]</sup> Devic's neuromyelitis optica (NMO) is an autoimmune channelopathy in which the target antigen is Aquaporin - 4 water channel which is found in cell membrane of the foot processes of astrocytes that surround and protect the blood brain barrier in central nervous system.<sup>[1,3]</sup> The disease may be monophasic or relapsing in nature<sup>[3,4]</sup> and the latter is more common. Neuromyelitis optica may present either with the simultaneous occurrence of acute transverse myelitis and optic neuritis or with these index events occurring separately by an indeterminate time interval.<sup>(4)</sup> The purpose of this case report is prompt diagnosis of relapsing NMO and to stress that all cases of transverse myelitis must be subjected thoroughly through complete ophthalmological examination for an early diagnosis of NMO and for vision salvage.

**CASE REPORT:** A 24 yr old female was referred from medical ward with chief complaint of vision loss in both eyes since one day. She was admitted with quadriparesis. Loss of vision is sudden over a period of hours, first in left eye then in right eye with no associated pain. One week before, a prodrome of fever with left upper limb weakness preceded the loss of vision and gradual weakness of all the limbs.

Patient had history of one episode of tonic clonic seizures and recovered without any sequelae.

Patient had history of two episodes of syncopal attacks with duration of one month, of which first attack was uneventful. Second episode was associated with fever, chills and rigors

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with sudden weakness of both lower limbs associated with urinary incontinence and after full investigations transverse myelitis was diagnosed and treated elsewhere and was able to stand with support. Within 6 months duration the patient got admitted again with similar history elsewhere and was referred to govt. general hospital as there was no improvement. On examination of both eyes, pupils were mid dilated not reacting to light with no perception of light. Fundus examination of both eyes showed temporal pallor. Central nervous system examination revealed weakness of both upper limbs and lower limbs. Below chest, sensations were lost. Urinary retention was present. Bilateral babinsky was positive. Haematological investigations were normal, except raised ESR which was 60mm 1<sup>st</sup> hour. Viral markers and other serological tests done were also negative. A N A antibodies were positive, Aquaporin antibody was negative, C S F analysis showed raised protein (78 mg), raised sugar (88mg), WBC 15cells/mm<sup>3</sup> and no oligoclonal bands. VEP shows increased latency of p100. MRI brain showed an ill-defined T2/ FLAIR hyperintense, T1 iso to hypointense signal changes noted in the brain parenchyma. MRI orbits showed signal changes in both optic nerves, optic chiasma and optic tracts suggestive of optic neuritis. MRI cervical spine showed LETM predominantly involving most of the transverse sections of the cord from C1 to C6 vertebral levels.

With the above clinical features and investigations and based on Wingerchuck et al's Revised diagnostic criteria, diagnosis of relapsing N M O was made.<sup>[3,4]</sup>

The patient was treated with iv methyl prednisolone 1gm/ day for 5 days, followed by oral prednisolone 1mg/ kg body weight for 3 weeks. As there was no significant improvement, later the patient was treated with Azathioprine 2 mg/ kg body weight and was on maintenance dose of azathioprine and prednisolone. With this treatment there is significant improvement in her motor function but no improvement in her vision was noted. The patient is in follow up till date with no significant change in vision.

<b>Absolute Criteria</b>	<b>SUPPORTIVE CRITERIA</b>
Optic neuritis	Brain MRI not meeting criteria for M S at disease onset
Acute myelitis	Spinal cord MRI with contiguous T2 – weighted signal abnormality extended over 3 or more vertebral segments indicating relatively large lesion in the spinal cord
	N M O ig G seropositive status

Table 1: Wingerchuck et al Revised Diagnostic Criteria for NMO

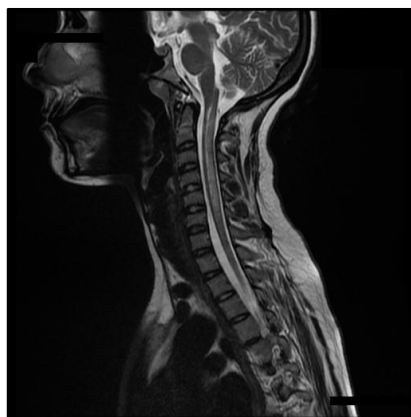
The new guidelines require two absolute criteria with at least two of three supportive criteria.

So our case fulfills 2 absolute criteria and 2 out of 3 supportive criteria (first two supportive criteria).

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**Fig. 1: T2 W coronal view MRI brain image showing**



**Fig. 2: T2W MRI sagittal scan Cervical spinal cord showing hyperintense**

Hyperintense optic nerves C1 to C6 segments.

**DISCUSSION:** In NMO, there is considerable time overlap between occurrence of ON and TM. In our case there is a time overlap of about 6 months or the patient might have gone through subclinical optic neuritis attacks which might be cumulative for her vision loss.<sup>[5,6]</sup> Involvement of both Optic nerves is rare with TM and the incidence is about 10% in NMO, which happened in our case. Severity of the Index events at the first episode may give clues regarding monophasic or relapsing nature of the disease.<sup>[6]</sup> There can be associated co-existing autoimmune disorders in devics disease.<sup>[6]</sup> In this patient even though there is no clinical evidence of connective tissue disorder, serum ANA antibodies were positive.

Features	Monophasic	Relapse
Frequency	Less common	More common
Age of onset	29 years	39 years
Sex ratio	About 50% female	80 to 90% Female
History of autoimmune disease	Uncommon	About 50%
Index events: ON or myelitis only	48%	90%
Bilateral ON	17%	8%
Simultaneous ON + myelitis	31%	0%
Recovery	Good	Fair

Table 2: Characteristics of Monophasic and Relapsing NMO<sup>(9)</sup>

Relapsing NMO: distinctive features.<sup>[3]</sup>

1. Marked female preponderance.
2. Higher age at onset.
3. Higher frequency of associated autoimmune disorders and serum autoantibodies.

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4. Less severe index events.
5. Worst outcome.

**TREATMENT:** There is no cure for NMO at this time,<sup>[7]</sup> and no medication have been specifically approved to treat it. The standard of care for an initial attack of NMO include.

- High dose iv corticosteroids (iv methylprednisolone 1gm/day for 5 days followed by oral prednisolone 1mg/kg body wt for 3 days).
- PLEX – plasma exchange – if no improvement with corticosteroids.

Because of likely hood of recurrences is more than 80%<sup>[4,6,7]</sup> and attacks are generally severe, ongoing treatment to suppress the immune system is considered necessary. The following medications are used for maintenance therapy: mycophenolate mofetil, azathioprine, rituximab.

Therapy under study aimed at anti-aquaporin 4 Abs-Aquaporumab, Sivelestat, Culizumab.

**CONCLUSION:** As there is considerable time lapse between Optic neuritis and Transverse myelitis, every case of Transverse myelitis need to be referred to an ophthalmologist for thorough ophthalmological examination, for early diagnosis and management of NMO to prevent the relapses and to limit visual disability. It is also necessary for careful assessment of severity of initial index events so as to assess the monophasic versus relapsing type of NMO and to plan management.

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