

# Neuromyelitis Optica Spectrum Disorder (NMOSD) in a Male with Hiccups and Quadriparesis

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## ABSTRACT

**Introduction:** Neuromyelitis-optica (NMO)/ Neuromyelitis-optica spectrum disorders (NMOSD) is an inflammatory disease of central nervous system classically characterized by acute, severe episodes of optic neuritis and longitudinally extensive transverse myelitis (LETM), usually with a relapsing course. The identification of an autoantibody exclusively detected in NMO patients against aquaporin-4 (AQP-4) has allowed identification of cases beyond the classical phenotype. Atypical cases of NMO exist in scientific literature. These cases do not present with all the symptoms.

**Case Report:** We present a case of a 53 year old female who presented to us with quadriparesis, intractable hiccups. MRI revealed longitudinally extensive transverse myelitis and later was diagnosed as a case of NMOSD.

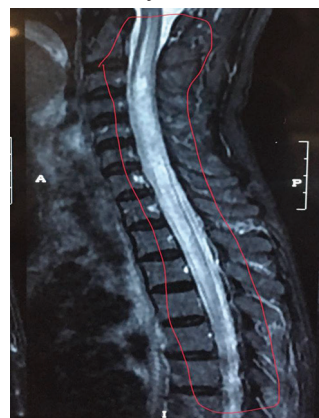
**Conclusion:** Neuromyelitis optica spectrum disorders (NMOSD) are inflammatory disorders of the central nervous system characterized by severe, immune-mediated demyelination and axonal damage predominantly targeting optic nerves and spinal cord. Serum NMO-IgG antibody is specific for the diagnosis.

**Keywords:** Aquaporin-4 (AQP-4), Hiccups, Neuromyelitis-Optica, NMOSD, Quadriparesis.

Respiratory system and Abdominal examination were normal. Fundus examination was done in view of optic neuritis but was found to be normal. Blood investigations like complete blood count, Liver function test, Kidney function test, Thyroid function test were done and found to be within normal limits. Anti-ds DNA, ANA, P-ANCA were done but were negative. However Anti NMO Antibodies were Positive.

CSF Examination was performed and findings were WBC-50cells/hpf (Most of them were Lymphocytes), Sugar was 80, Protein was 120, ADA was Negative. Oligoclonal bands were not present and IgG antibodies levels were normal.

MRI was done and suggestive of Intramedullary altered signal intensity from lower part of medulla to D3 which is predominantly central associated with cord enlargement. It is showing pachy enhancement on intravenous gadolinium contrast administration. Most like longitudinally extensive transverse myelitis.



## Treatment

Patient was started on i/v methylprednisolone 1 g once a day for 5 days and Tab Azathioprine 50mg/day. Nausea was subsided and the weakness improved over next 5 days. Patient

## INTRODUCTION

Neuromyelitisoptica (NMO) or Devic's disease is an inflammatory disease of central nervous system classically characterized by acute, severe episodes of optic neuritis (ON) and longitudinally extensive transverse myelitis.<sup>1</sup> It was first described by Allbutt and was further established as a clinical entity by Devic's in 1894.<sup>2</sup> NMO can also be associated with collagen vascular disease like SLE, Sjogren's Syndrome and infections like pulmonary tuberculosis. The reported incidence of NMO in women is up to 10 times higher than in men.<sup>3-5</sup>

## CASE REPORT

A 53 year old female presented to the emergency department with history of loss of power in all 4 limbs since 14 days and intractable hiccups and nausea since 14 days. There was no history of fever, waxing and waning of symptoms, recent vaccination, tingling and numbness, blurring of vision, diplopia, dysphagia, nasal twang to voice, facial asymmetry, vomiting and seizures.

On examination; Pulse was 80beats/min regular, BP was 130/90 mm Hg in right arm and other general physical examination was normal. CNS Examination revealed upper motor neuron type of sensory motor quadriparesis below C4 level with spastic bladder. Cardiovascular system,

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was put on oral corticosteroid (Tab Prednisolone 40mg/d) and was continued on Azathioprine. Patient was discharged after 1 week and awaiting follow-up after 1 month.

## DISCUSSION

Neuromyelitis optica (NMO, previously known as Devic's disease) and Neuromyelitis optica spectrum disorders (NMOSD) are inflammatory disorders of the central nervous system characterized by severe, immune-mediated demyelination and axonal damage predominantly targeting optic nerves and spinal cord. The discovery of a disease-specific serum NMO-IgG antibody that selectively binds aquaporin-4 (AQP4) has led to increased understanding of a diverse spectrum of disorders. Hallmark features of NMO include acute attacks of bilateral or rapidly sequential optic neuritis (leading to severe visual loss) or transverse myelitis (often causing limb weakness, sensory loss, and bladder dysfunction) with a typically relapsing course. Attacks most often occur over days, with variable degrees of recovery over weeks to months.<sup>6</sup> Central nervous system involvement outside of the optic nerves and spinal cord is recognized in patients with NMO and NMOSD. Other suggestive symptoms include episodes of intractable nausea, vomiting, hiccups, excessive daytime somnolence or narcolepsy, reversible posterior leukoencephalopathy syndrome, neuroendocrine disorders, and (in children) seizures. While no clinical features are disease-specific, some are highly characteristic. Neuromyelitis optica (NMO) usually tends to affect children or young adults. The average age at onset of symptoms is around 40 years, but all ages can be affected; it has a higher prevalence in females (F: M = 9:1). There is a higher prevalence of this condition in Western countries but Japan is the country with the highest proportion of NMO patients.<sup>7,1</sup> NMO is an idiopathic inflammatory disease of the central nervous system, episodically affecting the optic nerves and spinal cord. It is characterized by destruction of the myelin sheath, in association with areas of necrosis, cavity formation, small areas of perivascular lymphocytosis and in moderately advanced cases the formation of glial tissue.<sup>8</sup> Apart from this typical NMO atypical cases are also known which differ in clinical presentation and prognosis. The lesions of NMO can involve atypical sites like brainstem and hypothalamus. When the medulla is involved patient manifests with hiccups, nausea and vomiting due to involvement of the medullary vomiting centre (area postrema). The definition of typical NMO phenotype could be illustrated according to Wingerchuk's revised diagnostic criteria (2006)<sup>9</sup> as patients with ON, acute myelitis, and at least two of the following three supportive criteria: [1] longitudinally extensive spinal cord lesions contiguous over three or more vertebral segments; [2] lack of brain lesions in the magnetic resonance imaging (MRI) fulfilling MS criteria at disease onset and [3] serum positivity for AQP-4 antibody. Some patients with NMOSD present with brainstem symptoms due to medullary involvement. In particular, the area postrema clinical syndrome of nausea and vomiting or hiccups, sometimes intractable, with associated medullary lesions on MRI occurs

with an incidence of 16 to 43 percent in NMOSD. Brainstem involvement may lead to acute neurogenic respiratory failure and death.<sup>10</sup>

Diagnostic criteria — Revised consensus criteria published in 2015 unify the concepts of NMO and NMOSD and base the diagnosis on the presence of core clinical characteristics, AQP4 antibody status, and MRI neuroimaging features.<sup>11</sup>

1. Optic neuritis
2. Acute myelitis
3. Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting
4. Acute brainstem syndrome
5. Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
6. Symptomatic cerebral syndrome with NMOSD-typical brain lesions

The criteria recognize six core clinical characteristics, which are:

1. At least one core clinical characteristic
2. A positive test for AQP4-IgG using the best available detection method (cell-based assay strongly recommended)
3. Exclusion of alternative diagnoses

The diagnosis of NMOSD with AQP4 IgG antibodies requires the following<sup>11</sup>:

Our patient had acute longitudinally extensive transverse myelitis, area postrema syndrome and positive antibody status.

## CONCLUSION

Neuromyelitis optica (NMO, previously known as Devic's disease) and Neuromyelitis optica spectrum disorders (NMOSD) are inflammatory disorders cause immune-mediated demyelination and axonal damage predominantly targeting optic nerves and spinal cord. The discovery of a disease-specific serum NMO-IgG antibody that selectively binds aquaporin-4 (AQP4) is hallmark. Clinically rapidly progressive optic neuritis with transverse myelitis occurs with a relapsing and remitting pattern. Wingerchuk's revised diagnostic criteria is used for the diagnosis. Treatment is usually done with corticosteroids and other immunosuppressants like azathioprine.

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