

## A rare case of Quadripareisis

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### Abstract

Neuromyelitisoptica (NMO) is an inflammatory CNS syndrome distinct from Multiple Sclerosis (MS) that is associated with serum aquaporin 4 immunoglobulin G antibodies (AQP4-IgG).

**Keywords:** NMO (Neuromyelitis optica), MS (Multiple sclerosis), AQP4-IgG(aquaporin 4 immunoglobulin G antibodies)

### Introduction

Neuromyelitisoptica (NMO) is an inflammatory CNS disorder distinct from multiple sclerosis (MS) [1, 2 Traditionally], NMO was considered a monophasic disorder consisting of simultaneous bilateral optic neuritis and transverse myelitis but relapsing cases were described in the 20th century [3].

**Case study:** 15 year old girl presented with complaints of numbness and weakness of both upper limbs in the form of difficulty in mixing food, holding objects, combing hair followed by weakness of both lower limbs in the form of difficulty in wearing slippers and walking and difficulty in getting off the bed, since 1 month. H/O constipation, urine retention, absent sweating, loss of hot and cold perception below neck. No H/O fever, vaccination, no symptoms suggestive of cranial nerve, cerebellar or meningeal involvement.

O/E: Patient conscious, coherent, well oriented in place, person, and time. General examination: Unremarkable. Vitals stable.

CVS: NAD, RS: NAD, Abdomen: NAD, Single breath count: 17

### CNS

1. Higher functions: Normal
2. Cranial nerves: Normal
3. Motor system: Bulk-both UL have wasted thenar and hypothenar eminences(as shown in Fig.2), both LL normal Tone-Hypotonia in both UL, normal tone in both LL Power-3/5 both UL, 1/5 both LL Reflexes-Superficial reflexes: Conjunctival and corneal normal, Abdominal absent, Plantars bilateral extensors Deep tendon reflexes: Bilateral biceps, triceps, supinator absent, knee and ankle 3+
4. Sensory system: Graded sensory loss to pain, temperature and touch (LL>UL), Vibration and JPS: Lost
5. Cerebellum: Normal
6. ANS: Normal
7. Skull, spine, meninges: normal

### Investigations

1. CBP: Normal
2. ESR: 20 mm first hour
3. CUE: Normal
4. Renal and Liver parameters: Normal
5. Electrolytes: Normal
6. HIV, HCV, HBsAg: Non-reactive
7. TFT (Thyroid function test): Normal
8. ANA (Antinuclear Antibodies): Negative
9. CSF analysis: Unremarkable
10. MRI Brain: Normal
11. MRI Spine: Bulky cervical spinal cord with altered signal with patchy hyperintense areas and minimally prominent central canal at C2 to D1 levels-Features are highly suggestive of acute myelitis (as shown in Fig 3)
12. AQP4IgG: Positive
13. VEP (Visual evoked potential): Normal
14. Fundus examination: Normal

### Diagnosis: Non Compressive Myelopathy Longitudinal Extensive Transverse Myelitis Letm (C2 to D1) - Devices Disease

Patient was started on steroid pulse therapy followed by immunosuppressive therapy with Azathioprine 50 mg BID, with close monitoring of blood counts, supportive therapy with physiotherapy and bowel and bladder care. Patient responded dramatically and after 2 months patient is able to stand (Fig 4.) and walk without support and is able to carry out all her routine activities independently.

### Discussion

Neuromyelitisoptica (NMO) is an inflammatory CNS disorder distinct from multiple sclerosis (MS). [1, 2] It became known as Devic disease following a seminal 1894 report. [3] Traditionally, NMO was considered a monophasic disorder consisting of simultaneous bilateral optic neuritis and transverse myelitis but relapsing cases were described in the 20th century. [3] The nosology of NMO, especially whether it represented a topographically restricted form of MS, remained controversial.

A major advance was the discovery that most patients with NMO have detectable serum antibodies that target the water channel aquaporin-4 (AQP4-immunoglobulin G [IgG]) [4, 5] are highly specific for clinically diagnosed NMO, and have pathogenic potential [6] In 2006, AQP4-IgG serology was incorporated into revised NMO diagnostic criteria that relaxed clinical requirements by permitting unilateral optic neuritis or asymptomatic brain MRI lesions but retained the requirement for both myelitis and optic neuritis [2] The specificity of AQP4-IgG facilitated observations that further broadened the clinical and neuroimaging spectrum of NMO. In 2007, the term NMO spectrum disorders (NMOSD) was introduced to include AQP4-IgG-seropositive patients with limited or inaugural forms of NMO (e.g., first-attack LETM or recurrent or bilateral optic neuritis) who were at high risk for future attacks. [1] The NMOSD term also encompassed the cerebral, diencephalic, and brainstem lesions that occur in a minority of patients with otherwise typical NMO. It also included AQP4-IgG-seropositive patients with coexisting autoimmune disorders (e.g., systemic lupus erythematosus [SLE] or Sjögren syndrome [SS]). Finally, NMOSD potentially included patients diagnosed with opticospinal MS, an MS phenotype prominent in Asia and distinguished from Western MS. [7]

**NMOSD diagnostic criteria for adult patients**

Diagnostic criteria for NMOSD with AQP4-IgG

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

Diagnostic criteria for NMOSD without AQP4-IgG or NMOSD with unknown AQP4-IgG status

1. At least 2 core clinical characteristics occurring as a result of one or more clinical attacks and meeting all of the following requirements: **a.** At least 1 core clinical characteristic must be optic neuritis, acute myelitis with LETM, or area postrema syndrome **b.** Dissemination in space (2 or more different core clinical characteristics) **c.** Fulfilment of additional MRI requirements, as applicable
2. Negative tests for AQP4-IgG using best available detection method, or testing unavailable
3. Exclusion of alternative diagnoses

Core clinical characteristics

1. 1.Optic neuritis
2. 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

Additional MRI requirements for NMOSD without AQP4-IgG and NMOSD with unknown AQP4-IgG status

1. Acute optic neuritis: requires brain MRI showing (a) normal findings or only nonspecific white matter lesions, OR (b) optic nerve MRI with T2-hyperintense lesion or T1-weighted gadolinium enhancing lesion extending over 1/2 optic nerve length or involving optic chiasm
2. Acute myelitis: requires associated intramedullary MRI lesion extending over 3 contiguous segments (LETM) OR 3 contiguous segments of focal spinal cord atrophy in patients with history compatible with acute myelitis
3. Area postrema syndrome: requires associated dorsal medulla/area postrema lesions
4. Acute brainstem syndrome: requires associated periependymal brainstem lesions

Abbreviations: AQP4 5 aquaporin-4; IgG 5 immunoglobulin G; LETM 5 longitudinally extensive transverse myelitis lesions; NMOSD 5 neuromyelitisoptica spectrum disorders.

**Treatment**

There is no cure for neuro myelitisoptica (NMO) at this time, and no medications have been specifically approved to treat it. The standard of care for an initial attack of NMO includes the following:

- Intravenous (into the vein) high-dose corticosteroids (methylprednisolone)
- Plasma Exchange (PLEX) if no improvement occurs with corticosteroids. The goal of PLEX is to lower the level of NMO-IgG in the blood. PLEX involves removing blood from the body through a needle and tubing. Through a series of steps, the plasma (the liquid part of the blood) is separated from blood cells and replaced with an artificial plasma substitute; the plasma substitute and blood cells are combined and returned to the body through an intravenous line. The procedure lasts several hours and may be repeated multiple times over a number of days.

Because the likelihood of recurrence is greater than 90 percent and attacks are generally severe, ongoing treatment to suppress the immune system is considered necessary. The following medications are used for maintenance therapy:

- mycophenolatemofetil
- rituximab
- azathioprine



Fig 1

Fig 2

Fig 3

Fig 4

## References

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