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**Case Report**

# Neuromyelitis optica spectrum: a case report

**Harshini Kancherla*, Maheswari S. Kadimella, Ramya Bala Prabha Gelly, Ramo R. Tadikonda**

Department of PharmD, CMR College of Pharmacy, Kandlakoya, Hyderabad, India

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***Correspondence:**
Harshini Kancherla,
E-mail: kancherlaharshini@gmail.com



## ABSTRACT

Neuromyelitis optica (NMO) or Devic's disease is a rare inflammatory, demyelinating autoimmune disease of the central nervous system that primarily affects women, most commonly in their 30s and 40s. NMOSD has been linked to several different illnesses including autoimmune disorders. Recurrent bouts of optic neuritis, myelitis and brainstem disorders are caused by serum antibodies (Abs) against the aquaporin-4 (AQP4) water channel. No AQP4-Abs are detected in some patients with NMOSD symptoms, but Abs against myelin-oligodendrocyte-glycoprotein (MOG). The term MOG-encephalomyelitis is now commonly used to describe this clinical disorder. Long segments of inflammation in the spinal cord (myelitis), severe optic neuritis and bouts of persistent vomiting and hiccoughs are all characteristics symptoms. This was the case of 30 year old female who presented with a chief complaint of weakness of both lower limbs, unable to walk, tingling and numbness. Based on her MRI findings revealed that the patient was diagnosed with NMO spectrum. She was treated with immunosuppressants and corticosteroids.

**Keywords:** Aquaporin-4, Demyelinating, Devic's disease, Neuromyelitis optic spectrum

## INTRODUCTION

NMO, also known as Devic's disease, is a devastating, idiopathic, inflammatory demyelinating sickness of the central nervous system (CNS).[1] It is typically characterized by longitudinally extensive transverse myelitis is a type of myelitis that affects three or more vertebral segments and cause debilitation at the time of presentation. It can also cause unilateral or bilateral optic neuritis. Individuals can also show little lesions. It used to be often misdiagnosed as multiple sclerosis (MS) because it's regarded to be a variety of MS.[2] Abs against the astrocytes AQP4 water channel is found in the majority of patients with NMOSD, patients often suffer from recurrent attack if severe optic neuritis or/and myelitis.[3-7] Brainstem and brain involvement such as region postrema syndrome or diencephalon syndrome can occur in rare circumstances.[3]

NMOSD has a consistent prevalence of 1/100,000 population among Whites, with an annual incidence of 1 per million population. The incidence is higher among East Asians, at 3.5 per 100,000 people, whereas it may be as high as 10 per 100,000 people among Black. So far hospital-based research on MOG-antibody illness has found no apparent racial majority.[4] Eugene Devic and his doctorate student Fernand Gault originally described NMO (derived from neuromyelitis optique) in 1894. As a result, the sickness was previously known as Devic's disease. It wasn't until 2004 that the potential antigenic target, the AQP4 water channel was found and the two disorders could be successfully differentiated by AQP4 Abs detection. Antibody-negative and positive variants of NMOSDs are now grouped under the umbrella of NMOSDs in the most recent generation of diagnostic recommendations.[5] As a result, neurologic impairments commonly accrue during recurrent NMOSD, which accounts for around 80-85 per cent of cases. Patients who

do not receive long-term immunosuppressive medication have a worse prognosis and are more likely to die.[6] NMOSD is a severe inflammatory demyelinating illness of the central nervous system, says the beginning of most NMOSD papers. In reality, the international classification of diseases (2019 ICD-10-CM diagnosis code G36.0) classifies NMO (Devic) as a demyelinating disease of the CNS. Pathological examinations of AQP4 antibody-positive NMOSD cases, on the other hand, clearly shows that AQP4-expressing astrocyte is the primary target of immune attack and that astrocytic destruction is more severe and extensive than demyelination in the disease.

There are currently five lines of evidence: massive astrocytic damage is visible (severe loss of immunostaining for AQP4 and glial fibrillary acidic protein (GFAP)) in the NMO lesions, which is more extensive than myelin destruction; GFAP levels in the CSF of AQP4-antibody-seropositive NMOSD patients during acute exacerbations are extremely high, but CSF-GFAP levels in normal multiple sclerosis are not raised. Although CSF-myelin basic protein levels are higher in NMOSD than in multiple sclerosis; in the cervical cord of AQP4-antibody-seropositive NMOSD, Myo-inositol was measured by 1H-magnetic resonance spectroscopy; in experimental research, AQP4-antibody is harmful to astrocytes, AQP4-antibody is primarily IgG1 and may readily activate complements; complement-mediated cytotoxicity is a key cause of AQP4-antibody harm; foveal thickness is markedly reduced in AQP4-antibody-seropositive NMOSD compared to healthy controls on optical coherence tomography, and the foveal alteration near Muller cell-rich fovea suggests a retinal astro cytopathy.[7]

AQP4 was a transmembrane water channel that was extensively expressed in the end feet of CNS astrocytes and was required for optimal BBB function and maintenance of CNS water homeostasis.[8]

## CASE REPORT

A 30 years old female had chief complaints of weakness of both lower limbs for one week, unable to walk, tingling and numbness in the last 3 days and decreased urine output. In her history, she had similar complaints 2 years back.

On physical examination, vital signs were blood pressure=110/70 mmhg, pulse rate=112 bpm and $SpO_2$=94%.

The blood examination of complete blood count, liver function test and electrolytes were normal. Her blood urea level was 29.7 mg% and PT=13.8, INR=1.02. Her MRI spine revealed hypodensity noted in the spinal cord from D4-D7 level. Her cerebrospinal fluid showed glucose 54 mg/dl, proteins=25, chloride=116 mEq/l, her aquaporin antibodies were positive, her RF factor, anti ds DNA, lupus profile ANA was negative. Her ESR was 100 mm/hr.

The patient was started on injection methylprednisolone 1 g IV OD for 5 days, tablet liofer-XL 10 mg OD, tablet optineuron 10 mg OD, tablet prothiode 75 mg OD, tablet pantoprazole 40 mg OD. In her history she had received 6 cycles of cyclophosphamide therapy, the last cycle was received 6 weeks ago, she had also taken tablet beclofen 10 mg OD, tablet dosulepin 10 mg OD, tablet optineuron OD. The patient was discharged and didn't show up for follow up.

## DISCUSSION

In humans, two monomers were present, M1AQP4 and M23AQP4. AQP4ab played a crucial role in the pathogenesis of NMOSD. NMOSD patients were categorised by IgG, IgM and compliment deposit with a rosette pattern as well as cellular infiltrates of granulocytes. The loss of AQP4 on astrocytes was a crucial aspect. Other characteristic astrocytic markers such as GFAP were present in some lesions. AQP4 depletion appeared to precede astrocyte death. The severity of the disease as well as the related demyelination, was determined by the severity of the condition. Both grey and white matter were affected by demyelination. Necrosis and cavitation were sometimes present as well as thickening vessels that had been hyalinized, these results pointed to the autoimmune response in NMOSD primarily affecting astrocytes and were initiated by autoantibody-mediated loss of AQP4.[9]

Based on clinical and laboratory findings revealed that the patient had been diagnosed with NMO spectrum disorder. Initially, the main aim of therapy was to reduce hypodensity. Her MRI spine revealed hypodensity noted in the spinal cord from D4-D7 levels.

It can be more difficult to distinguish between MS and seronegative. To make an accurate diagnosis a careful review on history exam imaging and CSF results were essential.[10] So, the diagnosis and treatment were challenging in NMOSD patients.

However, Nurgul et al reported that in NMO patients the number of cells was more than 50 and polymorphs were seen and proteins may be increased our findings were corelating to their study. In our study also proteins were normal and polymorphs were seen and IgG index was negative.[11]

Therapy in NMOSD in acute exacerbation phase was to reduce the risk of relapse and long-term care. Corticosteroids were the main choice in the acute phase, intravenous methylprednisolone with a dose of 1 gm for five days which was supporting to our study. In our case also they had prescribed methylprednisolone 1 gm for 5 days.[12]

***Medications to avoid in NMOSD***

Interferon beta, natalizumab and fingolimod were used to treated MS which act as disease modifying agents but several studies suggested inefficacy or worsening of NMOSD due to use of those drugs. Hence, we avoided these drugs for NMOSD.[13]

In this case, she was started with methylprednisolone 1 g IV for 5 days, tablet prothiadm 75 mg OD, tablet optineuron 10 mg OD. On day 3 her vitals were normal but she had a complaint of joint position slightly impaired, same medications were continued on day 3, tablet carbamazepine 300 mg OD was added. On day 5 to day 9 same treatment was continued, on day 10 tablet wysolone 60 mg OD was added, on day 12 plan to start rituximab on discharge same medications were followed but the patient didn't show up for follow up.

## CONCLUSION

NMO spectrum is an inflammatory, demyelinating disease of central nervous system. Early diagnosis and initiation of therapy in patients to avoid persistent attacks. Long term use of immunosuppressants and corticosteroids will improve the quality life of an individual and reduces the disease activity. Potential cure for autoimmune disease may be achieved by the induction of immune tolerance to the autoantigen by vaccination.

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