



ORIGINAL RESEARCH PAPER

General Medicine

SIGHT STOLEN, STRENGTH TESTED: A YOUNG PATIENT'S BATTLE WITH NMOSD

KEY WORDS: Neuromyelitis Optica Spectrum Disorder, Aquaporin-4 Antibody, Optic Neuritis, Transverse Myelitis, Rituximab.

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ABSTRACT

Neuromyelitis optica spectrum disorder (NMOSD) is a relapsing, antibody-mediated inflammatory disease of the central nervous system that predominantly affects the optic nerves and spinal cord. The disease may result in severe disability, including blindness and paraplegia, if not recognized and treated promptly. We report the case of a 23-year-old female presenting with progressive visual loss and paraparesis, ultimately diagnosed with NMOSD based on positive aquaporin-4 antibody titers and classical MRI findings. Despite corticosteroid and immunosuppressive therapy, the patient continued to relapse, requiring escalation to rituximab and plasmapheresis. This case highlights the aggressive clinical course of NMOSD and emphasizes the need for early diagnosis and initiation of monoclonal antibody therapy to prevent irreversible disability.

INTRODUCTION

- Neuromyelitis optica spectrum disorder (NMOSD) is an inflammatory antibody-mediated disease of the central nervous system with relapsing course.
- Neuromyelitis optica spectrum disorder (NMOSD) term is now used to include optic neuritis with spinal cord manifestations and include other neurologic disorders associated with the serum aquaporin-4 immunoglobulin G antibodies (AQP4-IgG).
- The latest diagnostic guidelines unify antibody-negative and positive forms under the umbrella of NMOSD.
- Early diagnosis and treatment are important to reduce the risk of long-term disability and death.
- It is more frequent in women than men (9:1).

transverse myelitis. Bilateral optic disc atrophy was noted more in right side than left side.

- Serum Anti Aquaporin-4 (AQP4) antibodies : Positive
- Serum Myelin oligodendrocyte glycoprotein (MOG) : Negative
- ANA - 1:320 (+4) and anti-dsDNA: Negative
- ANA profile - Negative

Case Report

► History

A 23 year old female presented with diminution of vision on the left eye subsequently followed by right eye over a period of one year, weakness of bilateral lower limbs and inability to walk, and urinary incontinence and recurrent episodes of vomiting

► Examination

- Vitals - stable
- Visual acuity : Right eye - no perception to light and left eye - finger counting at 1 meter
- Pupillary examination : Both eyes reacting to light with RAPD in right eye
- Fundus examination - Pallor of the both optic disc
- No other cranial nerve deficit
- Tone - Hypertonia in both lower limbs
- Power - 5/5 in bilateral upper limbs and ½ in bilateral lower limbs
- Deep tendon reflexes - all DTR brisk with ankle clonus
- Plantar - Bilateral extensor response
- Sensory examination - Decreased perception of touch, pain and temperature below xiphisternum
- Cerebellar signs - Not present

► Investigations

- CBC, RFT, LFT - WNL
- CEMRI brain and orbit with spine revealed bilateral retrobulbar optic neuritis and longitudinally extensive



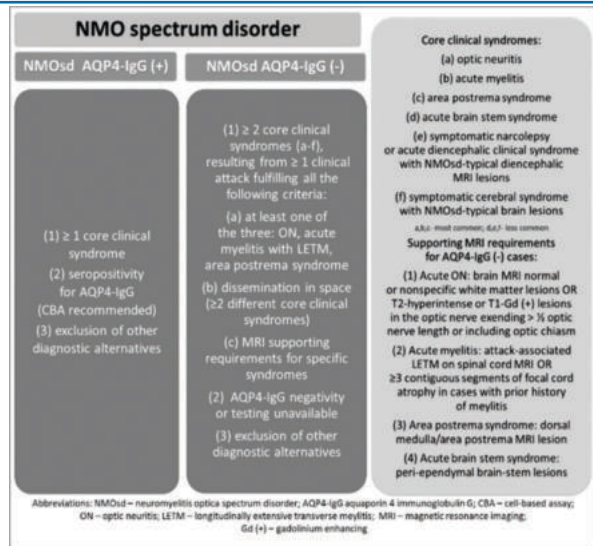


Figure 1: Diagnostic Criteria for NMO Spectrum According to International Panel for NMO Diagnosis (2015).

Management

- The patient was initially treated with pulse dose of IV Methylprednisolone (1 gm for 5 days) and discharged on oral prednisolone but was non compliant to it.
- Patient experienced relapse in the form of myelopathic symptoms for which Azathioprine was started but stopped after 3 days due to leukocytosis and mycophenolate mofetil was started.
- However there was no clinical improvement seen and the patient was started on injection Rituximab for maintenance treatment.
- Patient experienced another relapse, presented with diminished vision. Plasmapheresis (PLEX) planned for relapse in the form of optic neuritis.

DISCUSSION

- Neuromyelitis Optica Spectrum Disorder (NMOSD) is a severe, relapsing autoimmune disorder of the central nervous system characterized by inflammation and demyelination primarily affecting the optic nerves and spinal cord.
- A hallmark of NMOSD is its poorer response to treatment, frequent relapses, high morbidity due to residual neurological deficit compared to conditions like multiple sclerosis (MS) and myelin Oligodendrocytes Glycoprotein Associated Disease (MOGAD)
- Each relapse in NMOSD increases the risk of accumulating permanent neurological disability including blindness, paraplegia and sensory deficits.

CONCLUSION

- NMOSD is a devastating disease that requires aggressive treatment to prevent relapses, mitigate long term disability and improve quality of life.
- Monoclonal antibodies like Eculizumab, Inebilizumab and Satralizumab are recently approved for management for NMOSD and should be considered earlier in the disease course.

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