International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Rare Case of Seronegative Neuromyelitis Optica Spectrum Disorder Presented with Lower Limb Weakness and Blurring of Vision

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Abstract: ***Neuromyelitis optica spectrum disorder (NMOSD) is a rare autoimmune condition primarily affecting the optic nerves and spinal cord. While typically associated with the presence of aquaporin - 4 antibodies (AQP4 - IgG), a subset of patients present as seronegative. Here, we report a case of a 54 - year - old male presenting with bilateral lower limb weakness and blurred vision, ultimately diagnosed with seronegative NMOSD. Clinical examination revealed characteristic findings including optic disc edema and asymmetric limb weakness. Magnetic resonance imaging (MRI monstrated features consistent with acute optic neuritis andand longitudinal extensive transverse myelitis (LETM). Cerebrospinal fluid (CSF) analysis revealed lymphocytic pleocytosis and elevated protein levels, while serological tests for AQP4 - IgG and MOG antibodies were negative. Plasmapheresis was initiated, leading to gradual improvement in symptoms. This case underscores the importance of considering NMOSD in patients presenting with atypical neurological symptoms, even in the absence of seropositivity for aquaporin - 4 antibodies. Continued research is necessary to ter understand and manage seronegative NMOSD cases.***

1. Introduction

Neuromyelitis optica (NMO) is an autoimmune disease of the central nervous system (CNS) that mainly affects the optic nerves and spinal cord. It is sometimes referred to as NMO spectrum disorder or NMOSD.

The prevalence of NMOSD is approximately 0.3 to 4.4 per 100, 000 people. NMOSD characteristically occurs in females (80%) and younger patients between 30 and 40 years of age.

Even using the most sensitive cell - based assays, 12 percent of patients with a clinical diagnosis of NMOSD are seronegative for AQP4 - IgG of which 25% have anti - MOG antibodies positive.

2. Case Report

A 54 yr old male patient presented with Difficulty in walking due to bilateral lower limb weakness since 15 days and blurring of vision since 20 days.

Patient had history of urinary retention 2 months ago and was catheterized for that

Family History –
On Examination
Temp – Normal
Pulse – 110/ min
BP – 140/90 mmhg
RBS – 110mg/dl

RS/CVS – Normal
CNS – Conscious, Oriented to time place and person
Pupil – Bilaterally equally reactive to light
Plantar – Flexor on Left side, Extensor on Right
Power – Normal in upper limb, 3 in right lower limb and 4 in left lower limb
Tone – Normal in all 4 limbs
All Deep Tendon Reflexes present
Fundoscopic examination:
Disc – B/L nasal disc blurring with pale disc i. e. – B/L Disc Edema

Investigation
CBC, Renal function test, Liver function test, Serum electrolytes, TSH – Normal MRI Brain with Orbit with whole spine (p+c)

- Changes of acute optic neuritis involving posterior aspect of intraorbital segment and prechiasmatic segments (more than one half optic nerve length involvement)
- Enhancing focal lesions in left anterior frontal white matter, left cerebral peduncle, both superior cerebellar hemispheres and anterior part of medulla and non enhancing single lesion in left posterior parasagittal frontal white matter. No imaging signs of Dawsons Fingers.
- Patchy confluent T2 hyperintense enhancing lesions involving cervical and dorsal cord extending from C2 to D10 vertebral levels with contiguous plaque like enhancement at D2 to D5 vertebral levels and D6 to D8 vertebral levels (Longitudinal extensive transverse myelitis – LETM). No evident involvement of conus medularis.

CSF examination

- R/M – total count - 21

Neutrophils - 15
Lymphocytes – 85
Protein – 443

- C/S – NAD
- Oligoclonal Bands – Not seen

NMO Antibodies

Aquaporin - 4 antibody – Not seen
MOG antibody - Not seen
Serum ANA – Negative

Management
Patient was started on plasmapheresis.5 cycles of plasmapheresis were done. Gradually patient's limb weakness and visual acuity improved over the course of his admission.

3. Conclusion

In conclusion, this case report highlights a rare presentation of seronegative neuromyelitis optica spectrum disorder (NMOSD) characterized by lower limb weakness and visual disturbances. Despite the absence of typical aquaporin - 4 antibodies, the clinical features, MRI findings, and CSF analysis support the diagnosis of NMOSD. Plasmapheresis was effective in improving the patient's symptoms, emphasizing its role in the management of NMOSD. This case underscores the importance of considering NMOSD as a differential diagnosis in patients presenting with atypical neurological symptoms, even in the absence of seropositivity for aquaporin - 4 antibodies. Continued vigilance and further research are warranted to better understand and manage seronegative NMOSD case.

Acknowledgement
Acknowledgement No words can express my gratitude for my respected PG Teacher and Guide, Dr. Rahul Gambhir Sir (Head of Dept and Professor Medicine Department, PDUMC, Rajkot) who helped me & guided me during whole study with great interest and enthusiasm. Thesis was almost impossible without his invaluable encouragement and much needed constructive criticism. I am also thankful to my parents and family for their restless efforts to always keep my morale high in each & every step of my life.

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