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Case Study

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NEUROMYELITIS OPTICA: A RARE CASE WITH ATYPICAL PRESENTATION

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ABSTRACT

Neuromyelitis optica is a rare disease presenting with bilateral optic neuritis with severe visual loss and longitudinal extensive transverse myelitis involving three or more contiguous vertebral segments. We are reporting a case of a 35 year old lady who presented with tingling and numbness in all 4 limbs with a history of 10 days. The diagnosis was made after a clinical history, examination, MRI of the whole spine and Lab investigations. Patient was managed conservatively and recovery was seen on subsequent follow up. Though patient was not presented typically as NMO presents, where transverse myelitis or MS are probable diagnosis but we should keep NMO to avoid unnecessary cost of treatment and patient distress.

KEYWORDS: Neuromyelitis optica, Optic neuritis, Transverse myelitis, Endocrinopathy, Pleocytosis, Astrocytopathy.

CASE PRESENTATION

A 35 year old lady presented with tingling and numbness in all four limbs with weakness since 10 days. She had a history of band like sensation at T4-5 level. There was no history of visual disturbances, seizures, fever, neck pain, intractable hiccoughs or vomiting, or bladder and bowel involvements.

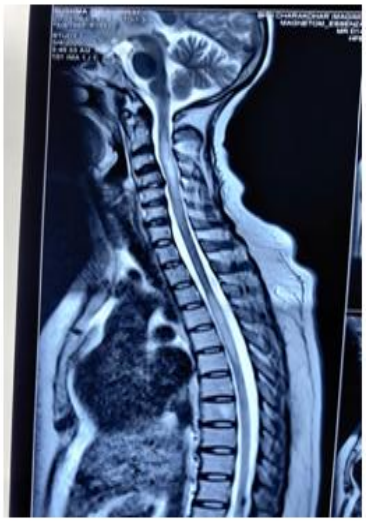
She also had no previous such episodes or any chronic illnesses in the past.

On examination she had 3+ reflexes with equivocal plantar response. Her motor and sensory examination were WNL. Blood investigations revealed.

Parameter	Patient value	Normal range
HB	12.3 gm/dl	11-15
TLC	9770/ul	4000-10000
DLC(N/L/E/M)	58/34/0.6/7.4	
PLATELET	192000/ul	150000-450000
MCV	92fl	80-100
LDH	245 U/L	105-248
RENAL Profile		
CREATININE	0.8 mg/dl	0.5-1.4
UREA	37.3 mg/dl	15-45
SODIUM	138	
POTASSIUM	4.2	
CALCIUM	9.4 mg/ dl	8.5-11.0
LIVER Profile		
SGOT/SGPT	29/40	9-37/10-40
TB/DB	0.3/0.2	0.1-1.2/0.01-0.3
TOTAL PROTEIN/ ALB	7.7/4.5	6.4-8.5
ALP	268 U/L	110-310
THYROID PROFILE		
T3/T4/TSH	99/7.1/4.5	60-200/4.5-12/0.3-5.5
HbA1c	5.0%	
ANA	0.4	<1.2

CSF findings s/o**. Pleocytosis with N/L 10/90**
The FUNDUS examination was **Within normal limits**.
Viral markers (HIV/HbsAg/anti-HCV) : Non Reactive
Serum NMO; Anti – Aquaporin 4, IgG **: POSITIVE**
MOG ANTIBODIES : Negative.
CSF OCB bands : NOT seen

MRI CERVICO-DORSAL spine



MRI cervical dorsal spine: patchy areas of intramedullary T2W hyperintensity seen at cervicodorsal cord at C2-5, D4-6 and D11-12 levels, likely areas of demyelination.

MRI BRAIN: no significant abnormality noted.

The patient was managed with high dose methylprednisolone 1 gm/day x 5 days followed by prednisone tapering dose plus azathioprine 50 mg BD for prophylaxis. On follow up, the patient was partially improved and tingling and weakness subsided.

DISCUSSION

The incidence and prevalence of NMO shows considerable variation between populations and geographic regions, with prevalence estimates that range from <1 to > 4 per 100,000.[1,2,3,4] Neuromyelitis optica (NMO; Devic's disease) is an aggressive inflammatory disorder characterized by recurrent attacks of Optic neuritis and myelitis[2]; NMO is more frequent in women than men (9:1)[5], and typically begins in adulthood but can arise at any age (The median age of onset is 32 to 41 years).[6,7,8,9] In patients with NMO, attacks of ON can be bilateral and produce severe visual loss (uncommon in MS); myelitis can be severe and transverse (rare in MS) and is typically longitudinally extensive involving three or more contiguous vertebral segments.[3] The brain MRI lesion in hypothalamus causing an endocrinopathy; the area postrema in the lower medulla presenting as intractable hiccoughs or vomiting; or the cerebral hemispheres producing focal symptoms, encephalopathy, or seizures.[4,11,12] Spinal cord MRI lesions typically consist of focal enhancing areas of swelling and tissue destruction, extending over three or more spinal cord segments.[8,14] Cerebrospinal fluid (CSF) findings include pleocytosis greater than that observed in MS,OCBs are uncommon, occurring in <20% of NMO patients.[16,17] The pathology of NMO is a distinctive astrocytopathy with inflammation, loss of astrocytes, and an absence of staining of the water channel protein AQP4 by immunohistochemistry, plus thickened blood vessel walls, demyelination, and deposition of antibody and complement.[2]

The key diagnostic test in NMO is the NMO antibody. Depending on the assay employed, sensitivity ranges from 50% to 75% with specificity close to 100%.[10] Patients with NMO often have positive antibodies for other conditions such as Sjogren's syndrome or lupus. NMO is typically a recurrent disease; the course is monophasic in fewer than 10% of patients. Untreated NMO is usually quite disabling over time; in one series, respiratory failure from cervical myelitis was present in one-third of patients, and 8 years after onset, 60% of patients were blind and more than half had permanent paralysis of one or more limbs.[4,13,15] There is limited data indicating that the long-term course of NMO has been substantially improved with the development of therapies to treat acute attacks and prevent relapses.[5]

CONCLUSION

Neuromyelitis optica is a rare disease but should be kept in mind as a differential diagnosis of Long extended transverse myelitis [LETM]. Early disease recognition is important because it is an acutely progressive disease which results in permanent debilitation such as paralysis, visual loss. The diagnosis is confirmed by MRI imaging and NMO antibody test. The patient should be kept on long term follow up so as to ascertain resolution, detect recurrences or development of debilitation.

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