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A Case Series of Neuromyelitis Optica Spectrum Disorders

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Introduction

Neuromyelitis optica (also known as Devic's disease) is an idiopathic, severe, demyelinating disease of the central nervous system that preferentially attacks the optic nerves and spinal cord producing optic neuritis and myelitis.

It was thought to be a variant of multiple sclerosis, however, clinical, laboratory, immunological and pathological features that distinguish it from multiple sclerosis are now recognized.

It is up to 9 times more common in women than men (F: M=9:1) .The median age of onset is 39.

In 10% of the cases, NMO affects the hypothalamus and brainstem, especially the areas around the fourth ventricle.

Brainstem symptoms include vomiting, vertigo, hearing loss, facial weakness, trigeminal neuralgia, diplopia, ptosis and nystagmus.

Some patients have nausea and intractable hiccups.

Aim of the Study

To Evaluate and Analyze Cases of Neuromyelitis Optica Spectrum Disorders Presenting to our Hospital, Great Eastern Medical School and Hospital, Srikakulam, Andhra Pradesh.

Materials and Methods

The study was conducted in Great Eastern Medical School and Hospital , Srikakulam, Andhra Pradesh, in the period of 1 year from September 2019 to August 2020.

All the cases diagnosed to have NMO / NMO SPECTRUM DISORDER were included in the study.

Evaluated clinically and underwent following investigations:

1. Bio-chemical: complete blood counts, blood sugar, liver and renal function tests
2. Chest x-ray
3. ECG

- Radio imaging - MRI BRAIN/SPINE
- Serologic work up(NMO- IgG, HIV, HSV, HBsAg)
- Lumbar puncture and CSF analysis,
- Evoked potentials (selected patients)
- Rheumatologic work up(ANA, RA, Anti-Ro/SSA)

Diagnostic criteria for NMO (Wingerchuk's criteria)

- Required criteria
    1) Acute myelitis
    2) Optic neuritis

- Supportive criteria (at least 2 of the following)
  1) MRI brain non diagnostic for multiple sclerosis at disease onset.
  2) MRI spinal cord lesion extending over > 3 vertebral segments.
  3) NMO IgG seropositivity.

Neuromyelitis optica spectrum disorders

- Limited forms of Neuromyelitis optica.

Idiopathic single or recurrent events of longitudinally extensive Myelitis (≥ 3 vertebral-segment spinal cord lesion seen on MRI)

Optic neuritis: recurrent or simultaneous bilateral.

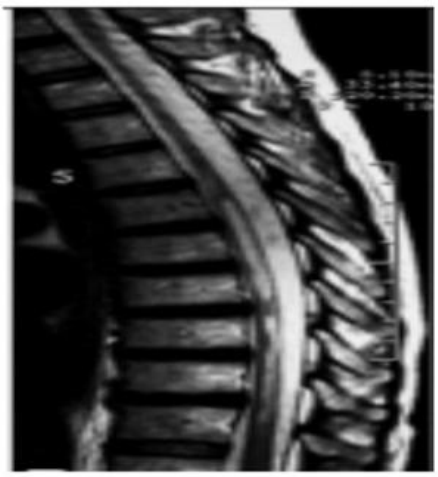
- Asian optic spinal Multiple Sclerosis.
- Optic neuritis/longitudinally extensive myelitis associated with systemic autoimmune disease.
- Optic neuritis/myelitis associated with NMO typical brain lesions (hypothalamic, corpus callosal, periventricular, brainstem)

	CASE 1 Female, age 50 years	CASE 2 Female ,age 18 years
History	Acute onset paraplegia of 2 days duration	Acute onset Hiccups, vomiting, diminished vision and ataxia of 8 days duration.
Clinical features	Spastic paraplegia with sensory level at D4 with bowel and bladder involvement	B/l complete ophthalmoplegia with truncal and gait ataxia
Past history	2 prior episodes of acute myelitis, 2 yrs and 1 yr back. Responded well to steroids	--
MRI BRAIN/SPINE	Long segment demyelination (D4 to D9)	Brain stem demyelinating lesions. (peri 4th ventricle)
NMO IgG	POSITIVE	POSITIVE
TREATMENT	Responded to I/V methyl prednisolone and presently on rituximab .	Responded to I/V methyl prednisolone.

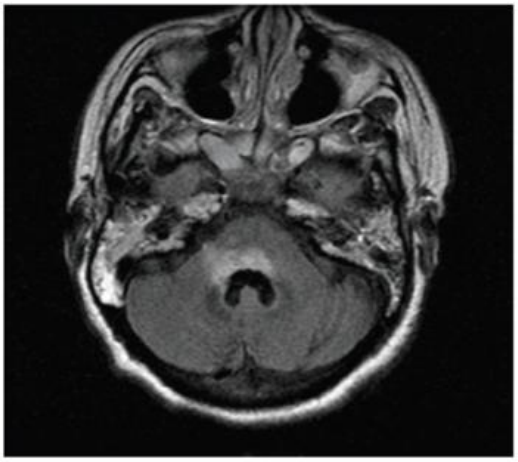
	CASE 3 Male ,age 58 years	CASE 4 Female , age 28 years
History	Acute onset paraplegia with urinary retention and constipation since 1 day	Acute onset diminished vision in left eye since 3 days
Clinical features	B/L spastic paraplegia with sensory level at D4 .	B/L optic atrophic changes.
Past history	Prior h/o B/L paraperesis 1 year back . Responded well to steroids.	Acute onset Diminished vision in both eyes 5 years back which improved partially with retrobulbar steroids.
MRI BRAIN/SPINE	Long segment demyelination -D5 to D11.	Normal
NMO IgG	POSITIVE	POSITIVE
TREATMENT	I/V methyl prednisolone was given and receiving rituximab presently.	I/V methyl prednisolone given and presently on azathioprine.

	Case 5 ,female,age 72 yrs	
History	Acute onset weakness of b/l lower limbs since 1 day.	
Clinical features	B/L spastic paraplegia with sensory level at D4 .	
Past history	--	
MRI BRAIN/SPINE	Long segment demyelination -D5 to D11.	
NMO IgG	positive	
TREATMENT	I/V methyl prednisolone given and presently on azathioprine.	

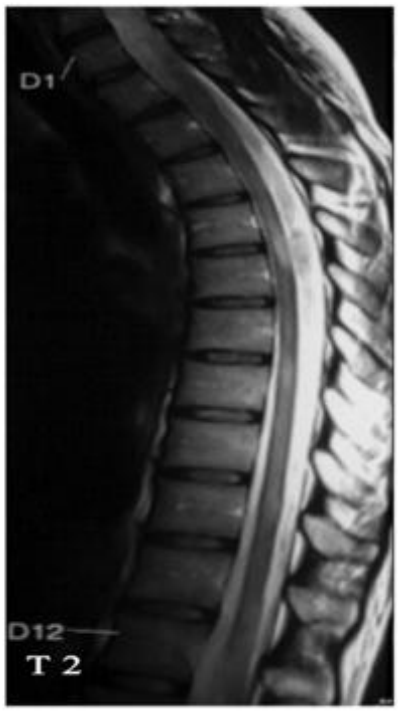
CASE 1



CASE 2



CASE 3



Results

- In our study, 2 cases had features of recurrent myelitis, one with brain stem involvement and one with optic neuritis.
- In our study it was found that, females are affected more(4 out of 5).
- I/V Methylprednisolone was given for acute treatment.
- Rituximab, azathioprine and other immunosuppressant may be used for preventing relapses.
- In our study those who presented with myelitis and brain stem features responded well to treatment, and who presented with optic neuritis responded partially.

Conclusion

- High level suspicion should be there for NMO spectrum disorders and NMO IgG antibodies should be done if patient having features of myelitis /optic neuritis /long segment involvement of spinal cord.
- If diagnosed, should be given I/V methyl prednisolone in acute phase and immunomodulators should be given to prevent relapses.
- It has a poor prognosis in relapses but early diagnosis, treatment with steroids in acute phase and using immunomodulators to prevent relapses can improve the outcome

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