CASE REPORT

Neuromyelitis Optica-A Distinct Disease Entity

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ABSTRACT

Introduction: Neuromyelitis optica is a demyelinating disorder involving optic nerve and spinal cord. It should be differentiated from other demyelinating disorders like Multiple sclerosis and Acute Disseminated Encephalomyelitis (ADEM). This case report is our attempt to discuss the dilemmas in diagnosis of NMO.
Case Report: We are reporting a rare case of 25 year female with classical clinical and radiological features of Neuromyelitis optica.
Conclusion: Neuromyelitis optica should be suspected in young females with optico-spinal involvement and treated effectively using steroids and immunosuppressants.

Keywords: Neuromyelitis Optica, AQP-4, optic neuritis, LETM

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INTRODUCTION

Neuromyelitis Optica (NMO), also known as Devic's Syndrome[1], is characterised by optic neuritis, myelitis and NMO antibody seropositivity. It is a distinct disorder known to be associated with connective tissue disorders and viral infections.[2] Treatment of acute episode of NMO includes steroid pulse therapy while azathioprine and Rituximab are used for maintenance therapy.[3] Our aim of reporting this case is to ensure early diagnosis and prompt initiation of treatment of NMO and discuss briefly about the features and management of this rare disorder.

CASE REPORT

A 25 year female patient presented to the Medicine OPD, Sawai Man Singh Hospital, Jaipur with chief complaints of bilateral lower limb weakness for five days, Painful loss of vision in both eyes and Sensory loss below the level of umbilicus for four days. No history s/o weakness in neck muscles & upper limb. History of fever 20 days back associated with exanthematous lesions over body ranging from papules, vesicles to healing crusts and scabs.
On general physical examination she was Conscious, Co-operative, Well oriented to time, place and person. Vitals were normal. There were fading rashes present on face, chest and abdomen suggestive of healed chicken pox. Higher mental functions were normal. Ocular examination revealed no perception of light in both eyes. Both pupils were dilated and fixed with absence of accommodation and light reflex. On Fundus examination, disc margin was blurred and disc oedema was present. Neurological examination revealed flaccid paralysis and loss of all modalities of sensation below T11. Bilateral planter reflexes were extensor.
The history and examination findings were suggestive of a demyelinating optico-spinal disease which further prompted us towards a differential diagnosis of Neuromyelitis Optica (NMO), Acute Disseminated Encephalomyelitis (ADEM) or an initial episode of Multiple Sclerosis.
Lab investigations showed normal CBC, Kidney function, thyroid function and plasma glucose. Anti nuclear antibody (ANA), Rheumatoid factor, Lupus anticoagulants were negative. CSF analysis was normal with negative oligoclonal band. CSF TB PCR was negative. Visual Evoked Potential (VEP) study performed by checkerboard visual pattern revealed non recordable waveform in both eyes.
In view of paraparesis, MRI of spine with contrast was done which showed Hyper intense lesion on T2 (Figure-1a), STIR (Fig. 1b), FLAIR sequence and isointense on T1 with cord expansion extending from T10 up to termination of cord further supporting Demyelinating disorder.
To ascertain differential of painful loss of vision, contrast enhanced MRI Brain with Orbit was advised

which showed chronic demyelinating plaques in right parietal-occipital deep white matter and left basal ganglia (Figure-2) Bilateral optic nerves were thickened and showed subtle altered signal intensity appearing hyper intense on STIR imaging (Figure-3). This pointed towards demyelinating CNS disorder with bilateral optic neuritis.

All this was suggestive of Neuromyelitis optica which was further supported by positive Serum NMO antibodies.

On the basis of history of chicken pox in the recent past and clinical features, VEP, radiological imaging and positive NMO antibodies, a diagnosis of NMO was made. For this acute attack of NMO, patient was treated with intravenous methylprednisolone 1 gm for five days followed by oral prednisolone 40mg once daily. Patient response started by day 4 in the form of Visual improvement. Patient could walk with support at time of discharge on day 11. The patient is now in our regular follow-up and doing well.

DISCUSSION

Neuromyelitis optica is an autoimmune inflammatory disorder characterised by optic neuritis and acute myelitis. This was first reported by Eugene Devic in 1894 after whom this entity is also known as Devic's Disease.[1] For long NMO was considered clinical variant of multiple sclerosis differing only in acuteness and intensity. However, detection of NMO IgG antibodies against water channel aquaporin receptors (AQP–4) present on astrocytes established this as a separate clinical entity.[4] In 2006, Wingerchuk et al explored likelihood ratios of several predictive models and finally proposed that in case of optic neuritis and acute myelitis, presence of atleast 2 of the 3 supportive criteria (Table-1) will have a sensitivity of 99% and specificity of 90%.[5]

Table 1 : Wingerchuk Criteria for NMO [5]
Absolute Criteria: 1. Optic Neuritis 2. Acute Myelitis
Supportive Criteria: 1. Contiguous Spinal Cord MRI Lesion extending over ≥ 3 vertebral segments 2. Brain MRI not meeting Diagnostic criteria for multiple sclerosis 3. NMO IgG Antibody seropositive status

Our patient satisfied all the absolute and supportive criteria for NMO. She had bilateral optic neuritis which was confirmed by VEP and MRI of orbit. There was also clinical history of acute paraparesis which on MRI spine showed Longitudinal Extensive

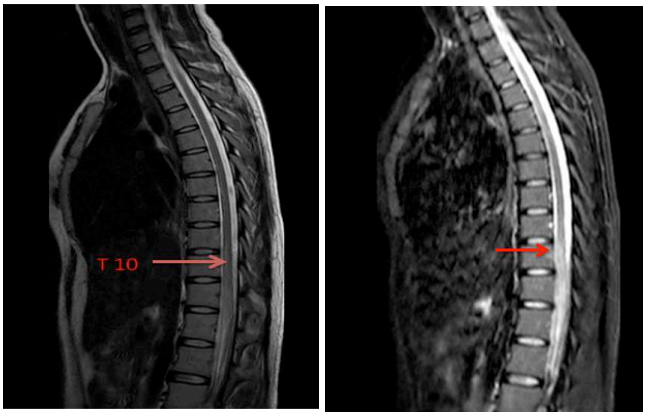

Figure-1: MRI Spine showing Hyperintensity with cord expansion T10 to conus (a: T2 Image; b: STIR Image)

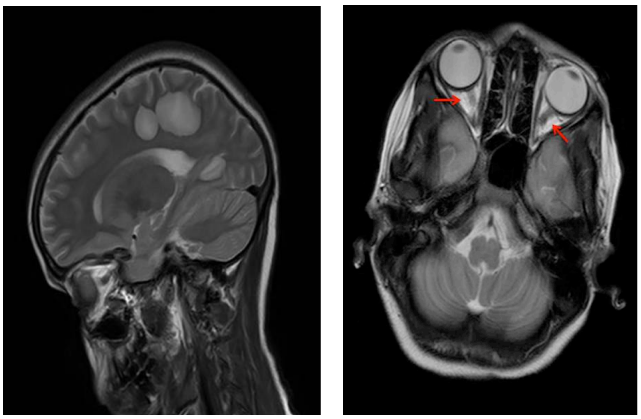

Figure-2: MRI Brain (T2 Saggital image) showing Hyperintense Parieto – Occipito – Basal ganglia lesions; Fig3: MRI Orbit (T2 axial image) showing Bilateral Optic Nerve thickening and hyperintensity.

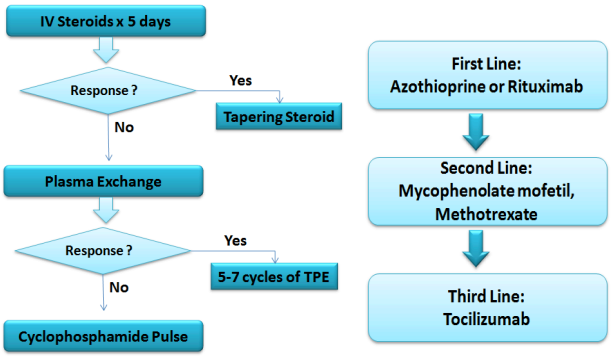

Figure-4: Treatment of NMO (a:Treatment of Acute Attack; b:Maintenance Therapy) Compiled from Trebst et. al. 2014[3]

Transverse Myelitis (LETM) involving spinal cord from T10 to further up to conus.

AQP4, present on the astrocyte membranes, is the first identified antigenic target in demyelinating disease. Antibodies directed against these receptors are found in 60-90% of patients of NMO.[6] Seropositivity is higher in females and those with relapsing disease. Antibody levels correlate with disease severity.[7] Sensitivity of these anti-AQP4 antibodies for detection of

NMO is ~ 75% and specificity is more than 90%.[6] Our patient was also positive for these antibodies.

NMO is known to be associated with other autoimmune diseases like systemic lupus erythematosus, Sjogrens syndrome, Myasthenia gravis, Sarcoidosis.[8] Onset of disease may be preceded by acute infections such as Varicella zoster virus, Epstein–Barr virus, HIV.[2] In our case preceding history of chicken pox was present.

Differential Diagnosis includes Multiple sclerosis, Acute Disseminated Encephalomyelitis (ADEM). In our case, brain imaging showing occipital lesion and absence of periventricular involvement along with bilateral optic neuritis and LETM favours diagnosis of Neuromyelitis Optica.

Treatment of acute attack comprises of intravenous methyl prednisolone 1gm for consecutive five days. Patients who fail to respond to steroids should be given therapeutic plasma exchange (TPE). Maintenance therapy includes immunosuppressants like Azathioprine and Rituximab (Figure-4).[3]

CONCLUSION

Neuromyelitis optica is a rare demyelinating disorder. It should be suspected especially in young females presenting with features of optic neuritis and myelitis with background history of viral illness. It must be differentiated from multiple sclerosis and ADEM because of different therapeutic interventions. If diagnosed early, NMO can be treated effectively. Relapse can be prevented with use of immunosuppressants.

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