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A Case Report of Neuromyelitis Optica Spectrum Disorders

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Abstract
Neuromyelitis optica (NMO) and Neuromyelitis optica spectrum disorders (NMOSD) are inflammatory disorders characterized by immune-mediated demyelination and axonal damage predominantly involving optic nerves and spinal cord. Previously NMO was considered as a variant of multiple sclerosis (MS), but now it is considered a separate entity(1). Here we report a case of middle aged female presenting with quadriplegia, urinary retention, sensory abnormalities which suggested myelopathy but without any evidence of optic neuritis, suggesting a diagnosis of neuromyelitis optica spectrum disorder. The diagnosis was confirmed by positive aquaporin 4 antibody.

Introduction

NMO is more frequent in women than men (>3:1) with the median age of onset of 32 to 41 years, but still case reports of NMO occurring in children and adolescents have been reported(2). The incidence and prevalence of NMO and NMOSD were high in black population. The recurrence rate is quite high in NMO cases which is around 70%(3). Children are more likely to be NMO IgG seronegative .Typically, the average age of onset is about 10 years later than that of MS(4).

Case History and Examination

A 52 Year old female presented with subacute onset, gradually progressive, proximal and distal weakness of both UL & LL with bilateral decreased sensation below the nipple and Urinary retention since 15 days duration. She had h/o constipation, vomiting, hiccups also. There was no H/o of cranial nerve, cerebellar, posterior column symptom, fatigue, fever, head and neck injury or seizure. She is non diabetic, normotensive, attained menopause 10years back.

On examination, the patient was Conscious, oriented, afebrile with stable Vitals. Central nervous system examination revealed a spastic quadriparesis with 4/5 power, exaggerated deep tendon reflexes and Bilateral plantar extensor. All modalities of sensation were reduced below the level of T4 with intact cranial nerves and normal cerebellar signs. Other system examinations were normal.

Basic investigations showed raised total count-18900 with normal ESR, renal and liver fuction test. CSF analysis showed Total count-10cells/µL, L-8%, M-2%; Glucose-60mg/dl, protein-54mg/dl, chloride-116mg/dl; negative for atypical/abnormal cells; oligoclonal bands were negative. MRI BRAIN was normal except for a small demyelinating lesion in area prostrema of medulla

(Fig:1) . MRI Spine showed Long segment cord hyperintensity with mild swelling in cervical cord proximally extending into lower medulla & distally till C5 (Fig:2a); few other short segment hyperintensity in lower cervical & upper thoracic cord till D5 (Fig:2b). Since the patient's MRI spine showed longitudinal lesions extending more than 3 spinal segments, NMO was thought of and anti-aquaporin 4 antibodies was done ,which turned to be positive.

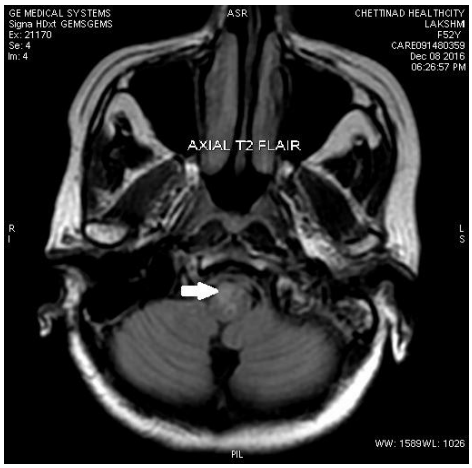

Figure:1 MRI-Brain

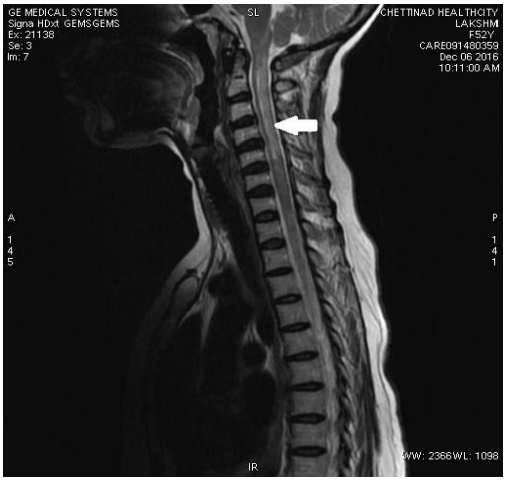

Figure:2a - MRI-SPINE

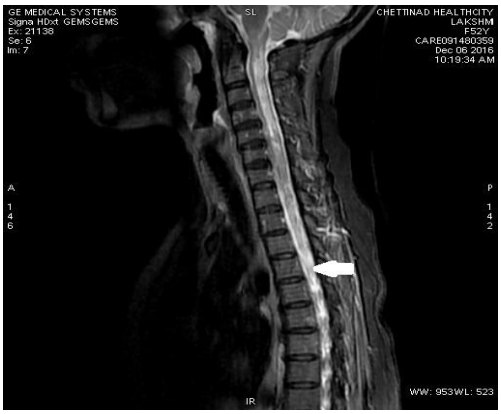

Figure:2b-MRI SPINE

Fundus exam and Visual evoked potential– normal. Hence the diagnosis of neuromyelitis optica spectrum disorder was made. Patient was managed with steroids which was gradually tapered and steroid sparing agent along with physiotherapy.

Discussion

Neuromyelitis optica is an aggressive inflammatory disorder characterized by recurrent attacks of optic neuritis (ON) and myelitis .It can manifest at any age and found more frequent in women than men (>3:1). Attacks of ON can be bilateral (rare in MS) or unilateral; Myelitis can be severe and transverse (rare in MS) and is typically longitudinally extensive, involving three or more contiguous vertebral segments[5]. The incidence of NMOSD in women is up to 10 times higher than in men, but cases are described in children and older adults. NMO has varying diagnostic criteria (Table-1), which are listed below. NMO and NMOSD criteria, later it was updated with six clinical core charecteristics (Table-2). Each of it, was later classified on the basis of AQP4 igG Antibody status of the individual.

Diagnostic Criteria for Neuromyelitis Optica (Table- 1): [6]

Table-1:

Required:

- ✓ Optic neuritis
- ✓ Acute transverse myelitis

Supportive (2 of 3 criteria required):

- ✓ Longitudinally extensive cord lesion extending over 3 or more vertebral segments
- ✓ Brain magnetic resonance imaging normal or not meeting criteria for multiple sclerosis
- ✓ Aquaporin-4 seropositivity

NMO and NMOSD criteria recognize six core clinical characteristics, which are (Table-2) [7]

Table-2:

1. Optic neuritis
2. Acute myelitis
3. Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting
4. Acute brainstem syndrome
5. Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
6. Symptomatic cerebral syndrome with NMOSD-typical brain lesions

As aquaporin 4 igG antibody was positive with transverse myelitis (one among the six clinical core characteristics),a Diagnosis of NMOSD with AQP4 igG was made with the following criteria (Table-3)[8]

Table-3:

- ✓ At least one core clinical criteria
- ✓ A positive test for AQP4-IgG using the best available detection method (cell-based assay strongly recommended)
- ✓ Exclusion of alternative diagnoses

In patients with acute or recurrent attacks of NMO or NMOSD, suggest initial treatment with high-dose intravenous methylprednisolone, plasma exchange, long-term immunosuppression treatment; natalizumab, or fingolimod is not effective and may be harmful[9].

Conclusion

Diagnostic criteria for NMOSD require the presence of at least one core clinical characteristic. It is not mandatory that all patients with NMO should have optic neuritis. Sometimes in early stages patient may present without optic neuritis (as in our case).since it causes the Long-term disability and high mortality. So in such cases early diagnosis and management would help in preventing the progression of disease.

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