Neuromyelitis Optica

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Abstract

Neuromyelitis optica (NMO) is a rare, idiopathic, inflammatory disease affecting the spine and characteristically sparing the brain. It mimics multiple sclerosis (MS) in many aspects. A case report of 55 year old house-maker, with recurrent attacks of flaccid weakness with visual and bladder disturbances who was admitted in the Physical Medicine and Rehabilitation (PMR) ward for neuro-rehabilitation is presented. Her serological studies and magnetic resonance imaging of the spine and brain revealed a diagnosis of idiopathic demyelinating disease affecting the spine – Devic's disease.

Key words : Neuromyelitis Optica, Multiple Sclerosis, Functional Independence Measure, Optic neuritis.

Case report

A 55 year old home maker from Trivandrum was admitted in the PMR department with two days' history of gradual onset of weakness of right lower extremity. Later her weakness spread to the left lower extremity. She had decreased vision in both eyes. She also complained of tonic muscle spasms involving both lower extremities and increased frequency of micturition. However her bowel was continent. Ten days back she had backache with radiation to both lower extremities. She had similar weakness last year with weakness of all four limbs and bladder disturbance. She had recovered fully with medication and rehabilitation. History of bilateral optic neuritis was noted. There was no history suggestive of trauma, primary malignancy, collagen vascular disorders or any sexually transmitted diseases in the past.

On examination she had a Functional Independence Measure (FIM) score of 90 out of 126, with complete dependence in bladder and locomotion skills with partial dependence in toileting, bathing, lower body dressing and transfers. Her higher mental functions were normal. Cranial nerves other than the Optic Nerves were uninvolved. She could only count fingers at one meter with her right eye and had no perception of light in the left. Muscle bulk was normal in all four limbs. She had flaccid paraparesis with a power of grade 2 (Medical Research Council Grading) in the lower limbs. Deep tendon reflexes were diminished in both lower limbs with extensor plantar response bilaterally. She had decreased sensation to pinprick, vibration and light touch from the second lumbar segment downwards in both lower limbs. Power, sensations and reflexes were intact in both upper limbs. Superficial and deep anal sensations were preserved. She also had voluntary anal contraction with preserved anal wink.

Her investigations showed that rheumatoid factor, antinuclear antibody and C - reactive protein were positive. Cerebrospinal fluid analysis showed no pleocytosis or protein derangement. Cystometrogram revealed a safe capacity of 250 ml with sustained elevation of detrussor pressure to 22 cm of water. Spine radiographs were normal. Her MRI of the cervico dorsal spine revealed scattered areas of linear hyperintense signals at the fourth cervical to second thoracic levels involving the posterolateral cord and mainly centred in the white matter, suggestive of demyelination. MRI of the brain was normal, so were her electromyogram and nerve conduction studies. In view of non availability of test her NMO IgG auto antibody [1,2,3] was not done. Putting all the investigations and clinical findings, she was diagnosed to have neuromyelitis optica or Devic's disease[3,4]

Differential diagnosis[1] :

1 . Multiple sclerosis
2. Vasculitis due to autoimmune disease
3. Paraneoplastic syndrome
4. Vitamin b12 deficiency

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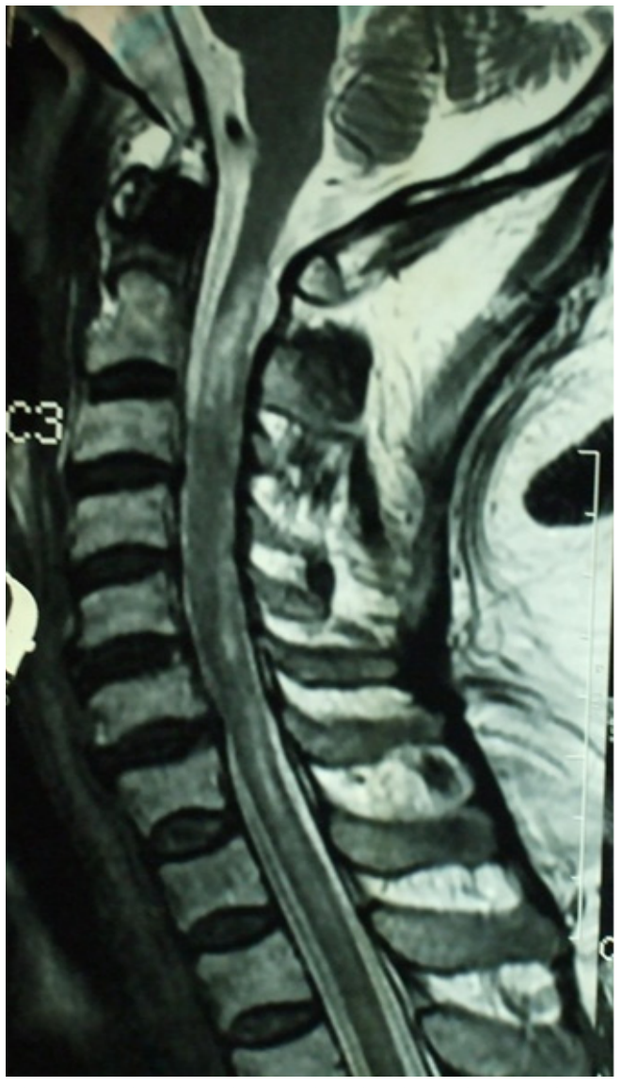

Fig 1. Cervico dorsal MRI showing hyperintense focus.

Diagnostic criteria for NMO have been proposed by Wingerchuck[4]. Diagnosis requires all absolute criteria and one major supportive criterion or two minor supportive criteria

Absolute criteria:

1. Optic neuritis
2. Acute myelitis and
3. No evidence of clinical disease outside the optic nerve or spinal cord.

Major Supportive Criteria:

1. Negative brain MRI at onset
2. Spinal cord MRI with signal abnormality extending over vertebral segments
3. CSF pleocytosis of > 50 WBC/ mm^3 or > 5 neutrophils/ mm^3,

Minor Supportive criteria:

1. Bilateral optic neuritis
2. Severe optic neuritis
3. Severe, fixed, attack-related weakness in one or more limbs.

Management: She was started on parentral steroids (methylprednisolone 1 gm i/v)[1,2,3] for 5 days and later changed to oral steroids which was gradually tapered over 3 months.Immunosuppresion with azathioprine[1,2,3] 50 mg was started and continued with fortnightly screening of routine blood examination and platelet count. Gabapentin was given for neuropathic pain. Neurovitamins were also given.

She was started on therapeutic exercise[7] with active range of motion to both lower limbs and reconditioning exercise to bilateral upper limbs. She was counseled about positioning and daily range of motion exercises at bedside to prevent complications of immobilization. She was put on tilt table for benefits of upright posture. On manual post voidal residual examination[5,6] significant residual urine of 100 ml was detected. She was catheterized and put on strict drinking schedule

Course in hospital : She responded to treatment. Her motor power improved and with further training for locomotor skills in parallel bars she was discharged with walker. Vision did not show any improvement. Intermittent clean catheterization was advised to her attendant as she was unable to do so due to decreased vision.

Follow up: She was followed monthly after discharge and on first follow up visit she was prescribed tripod cane for ambulation. On the next follow up she was ambulant without support with grade 4 power in bilateral lower limbs. FIM score was 113/126.

Discussion

Devic's NMO[1,2,3] is characterized by a unilateral or bilateral optic neutiis and transverse myelitis with variable interval between the two events. It is usual for the optic neuritis to precede the myelitis but in our patient myelitis preceded ophthalmic features of optic neuritis.In NMO[3] a positive antibody nuclear status can be positive without evidence of systemic connective tissue disease as seen in this case. It was also unusual to have normal CSF findings. NMO differs from multiple sclerosis by being restricted to spine and optic nerves only, more severe in attacks and in 30% cases associated with auto immune disease. On investigatory grounds CSF oligoclonal[1,2,3,4] bands are usually absent in NMO. MRI brain is non specific or normal in NMO whereas it shows multiple periventicular white matter lesions in multiple sclerosis. This diagnosis was made as this case fulfilled all absolute criteria, two major and two minor ones. It is important that the distinct signs and symtoms of NMO are recognized in order to permit early and

proper diagnosis, followed by appropriate rehabilitation and pharmacologic management.

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