
Case report

Neuromyelitis Optica - Devic's Disease - A Case Report

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Abstract:

Neuromyelitis optica also named Devic's disease is an acute combined optic neuritis and transverse myelitis occurring simultaneously or separated by months. We present a case of a 42 year-old man with recurrent bilateral optic neuritis and spinal cord involvement. Clinical MRI and cerebrospinal fluid data have fulfilled diagnostic criteria for Devic's Neuromyelitis optica (NMO). CSF examination showed mild pleocytosis and absence of oligoclonal band. MRI spine revealed demyelination of thoracic spinal cord. Visual evoked potential showed prolongation of P100 latency in both eyes. After Prednisolone treatment, his visual acuity began to improve on the 3rd Day and motor function improved on the 10th day.

Key words: optic neuritis, myelitis.

Case History:

A 43-year-old man from southern Tamilnadu was admitted to our hospital in June 2004 for loss of vision and paraplegia of 10 days duration. Visual loss was of painless sudden onset followed by flaccid weakness of both lower limbs with in hours. One month back he had one episode of

sudden loss of vision in both eyes, which recovered with 5 days of oral steroids. There was no history suggestive of connective tissue disorder, tuberculosis or trauma.

On admission his blood pressure was 130/80 mmHg, pulse rate 96/min and body temperature 37.1°C. His neurological examination revealed flaccid paralysis of both lower limbs

with loss of all modalities of sensation from T6 level. A bilateral positive babinski sign was noted. Cranial nerve examination revealed bilateral dilated pupils with absence of light and accommodation reflex. Perception of light was absent. Fundus examination showed bilateral optic atrophy. Other cranial nerves and cognitive functions were normal. He also had urinary retention necessitating bladder catheterization.

Laboratory work-up revealed normal hematocrit, white blood cell counts, platelets and increased erythrocyte sedimentation rate (48mm/hr). VDRL and HIV ELISA were negative. Anti nuclear antibodies (ANA) and antibodies to ds-DNA were not found. Analysis of cerebro spinal fluid revealed a clear Colour, 40 WBC'S per mm³, protein 20 mg/dl and sugar 80 mg/dl. Oligo clonal bands were negative (figure.1).

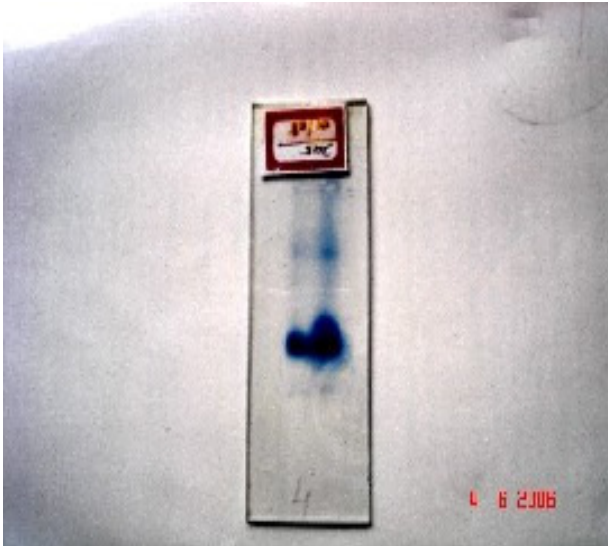


Figure.1 -CSF electrophoretogram showing the presence of major fraction of albumin & minor fraction of beta globulin & traces of pre albumin with no oligoclonal bands

Viral studies on serum and cerebro spinal fluid were negative. Staining and culture of CSF did not reveal any acid fast bacilli. Serum lactate levels were normal. An MRI examination of spinal cord obtained on the third day of hospital stay was suggestive of transverse myelitis (figure.2).

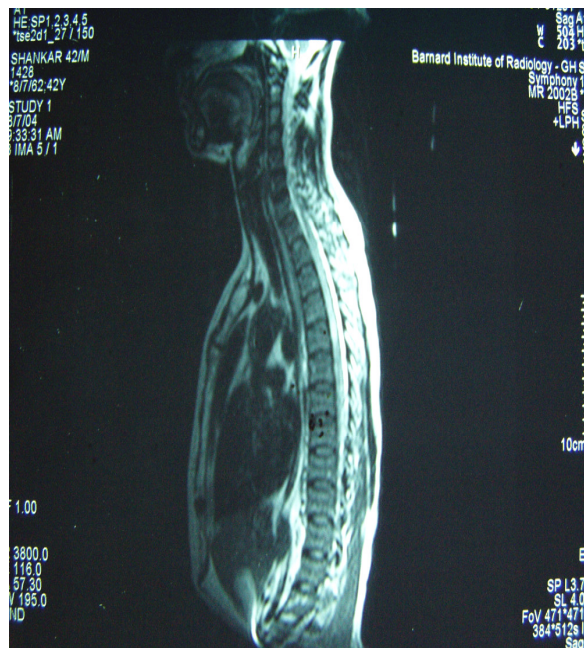


Figure.2- T2 weighted saggital MRI of the thoracic spine showing diffuse ill-defined hyper intensities involving mid and lower dorsal spine

MRI brain was normal. Visual evoked potential study (VEP) showed prolongation of P₁₀₀ latency in both eyes suggestive of demyelination. A diagnosis of Devic's disease was made and the patient was treated initially with intra venous methyl prednisolone (1gm/day for five days) followed by gradually tapered oral steroids. His visual symptoms began to improve by third day of treatment and motor symptoms by tenth day. By tenth day he was able to count fingers and move his lower limbs with gravity eliminated. During out patient follow up, periodic attempts to withdraw steroids were associated with further worsening of weakness. Patient was restarted on steroids and is presently on 30mg/day of oral prednisolone.

Discussion:

Devic's disease is defined as a syndrome characterized by acute optic neuritis in combination with acute transverse myelitis, demyelinating/necrotising lesions form in one/both optic nerve & myelitis may be simultaneous or separated by several months¹. First described in 1894, its incidence worldwide is around 5 per one lakh. The disease is much rarer in India. Singhal² BS has reported 14 cases from 1957-83. It is more common in Japan & East Asia. The name Devic's disease, Devic's syndrome, Neuromyelitis Optica is used interchangeably. It occurs in patients of varied ages (1 – 73 yrs). Monophasic illness has a mean age of 27 yrs at onset, whereas relapsing type tends to occur at a mean age of 43 yrs & has female preponderance.

The clinical picture of optic neuritis and myelitis develops over hours to days often preceded by headache, fever or myalgia. More

than 80% of patients develop bilateral optic neuritis. Severe degrees of neurological deficits are usual and degree of recovery is variable. Histopathology³ often shows demyelination across multiple levels with cavitation, necrosis, axonal spheroids in gray and white matter and loss of oligodendrocytes.

Diagnostic criteria for NMO have been proposed by Wingerchuck⁴ and Col. According to this criteria, diagnosis requires Three absolute criteria: 1) Optic neuritis, 2) acute myelitis, and 3) no evidence of clinical disease outside the optic nerve or spinal cord; as well as at least one of the following major supportive criteria: 1) negative brain MRI at onset, 2) spinal cord MRI with signal abnormality extending over 3 vertebral segments, 3) CSF pleocytosis of >50 WBC/mm³ or >5 neutrophils/mm³, or two of the following Minor supportive criteria: 1) bilateral optic neuritis, 2) Severe optic neuritis, 3) severe, fixed, attack-related weakness in one or more limbs.

Our patient has initially presented with optic neuritis, myelitis and MRI spinal cord abnormalities. Three absolute criteria, two major supportive criteria, and two minor supportive criteria were present. Laboratory tests have excluded other diseases, such as collagen and vascular diseases, auto-antibodies syndromes, and infections. Therefore, the signs and symptoms initially displayed by our patient are consistent with the diagnosis of NMO according to Wingerchuck's and col. criteria.

Oligo clonal bands⁵ in CSF are seen only in 17% of patients with NMO. There are few studies addressing NMO treatment. Patients with acute or sub acute onset often respond to Corticosteroids. The combinations of

prednisolone and azathioprine have reduced the attacks frequency in an uncontrolled series. Plasma exchange has been tried with good results. Interferons and immunosuppressive drugs efficacy has not yet been proved to be effective in preventing new attacks. Those patients destined for recurrent

myelitis and optic neuritis have a longer interval between the onset of myelitis and optic neuritis. The vast majority of patients with relapsing NMO are found to have very aggressive disease with frequent and severe exacerbations and a poor prognosis.

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