A Treatable Cause of Cervical Myelopathy in a Young Female

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Abstract

Neuromyelitis Optica(NMO) or Devics disease is an inflammatory demyelinating disorder of the central nervous system involving the neurons of the optic nerve and the spinal cord. It is seen more commonly in females and follows a relapsing clinical course in more than eighty percent of cases. The incidence of the same is rising in the Indian subcontinent. We present here a case of a young female patient presenting with features of a subacute cervical myelopathy with longitudinally extensive transverse myelitis on imaging of the spine. Investigations revealed features of Neuromyelitis Optica. She was diagnosed and treated early which led to good long term outcome.

Key words: Neuromyelitis Optica, Demyelination, Longitudinally Extensive Transverse Myelitis

Introduction

Neuromyelitis Optica (NMO) is a chronic inflammatory autoimmune disease of the central nervous system associated with a characteristic pattern of astrocyte dysfunction and loss, leading to demyelination and neurodegeneration[1]. Originally known as Devic's disease, NMO mostly follows a relapsing course, and was long considered a variant of multiple sclerosis(MS). In 2004, discovery of a pathogenic NMO-associated IgG antibody, targeting the water channel membrane protein aquaporin-4(AQP 4), helped in differentiating NMO from MS[2]. After different clinical presentations were described for the disease, the term NMO spectrum disorder (NMOSD) was newly introduced in 2007. AQP4 is highly concentrated on astrocyte end-feet in various parts of the nervous system. Although pathogenic AQP4-antibodies (AQP4-ab) are found exclusively in patients with NMO, approximately 20–30% of NMOSD patients are seronegative for AQP4-ab [3] and other antibodies may be seen here. Typical clinical features of NMOSD include acute attacks of bilateral or rapidly progressive optic neuritis leading to severe visual loss or transverse myelitis with a relapsing course. Attacks most often occur over a period of hours to days, with varying degrees of recovery.

Spinal cord involvement in NMOSD typically presents with transverse myelitis, characterized by symmetric paraparesis or quadriparesis, bladder dysfunction, and sensory loss below the level of the lesion . Accompanying symptoms may include paroxysmal tonic spasms of the trunk or extremities, radicular pain, unsteadiness or Lhermitte sign[4].

Patients with NMOSD have a longer extent of spinal cord demyelination than patients with MS, generally involving three or more vertebral segments on magnetic resonance imaging (MRI), a condition termed longitudinally extensive transverse myelitis (LETM)[5].

We present here a young female with a subacute presentation of a cervical myelopathy who was found to have features of NMO on imaging.

Case report

This 30 year old female patient with no prior co morbidities presented with a one month history of intermittent episodes of numbness of all four limbs and difficulty in walking. The numbness involved all four limbs from neck downwards and was associated with

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imbalance on walking and a tendency to sway to either side. She then presented with a three day history of weakness of right upper limb which was predominantly distal in nature. There was no weakness of lower limbs or bladder involvement. There was no history of any preceding fever or respiratory tract infection. There was no significant past history or systemic symptoms. There were no symptoms of raised intracranial pressure .

Clinical examination revealed evidence of spasticity in both lower limbs along with bilateral extensor plantar responses, exaggerated deep tendon reflexes in all four limbs , ankle clonus and normal cranial nerve and sensations. Motor examination showed distal weakness in right hand. Lhermittes signs was positive. There was no clear sensory level on examination. Clinical localization was to the cervical spinal cord .

Imaging done-MRI of the cervical spine showed a long segment hyperintensity involving the cervical cord extending from the cervicomedullary junction to the lower border of C5(Figure 1). This was in keeping with Longitudinally Extensive transverse myelitis or LETM which was characteristic of demyelination secondary to NMO. There was peripheral enhancement on contrast. The other differential was of a cervical intramedullary neoplasm. MRI of the brain was normal. Serum NMO / Aquaporin antibody was strongly positive suggestive of NMO. CSF done showed raised proteins suggestive of an inflammatory process with normal cells and sugars.

A final diagnosis of an NMO was made and she was given intravenous pulse dose steroids followed by a long taper of oral steroids. In view of antibody positivity, she was started on long term immune modulation with Mycophenolate. She had complete clinical improvement with the same and was kept on regular follow up. Repeat imaging after three months showed resolution of the lesions (Figure 2).

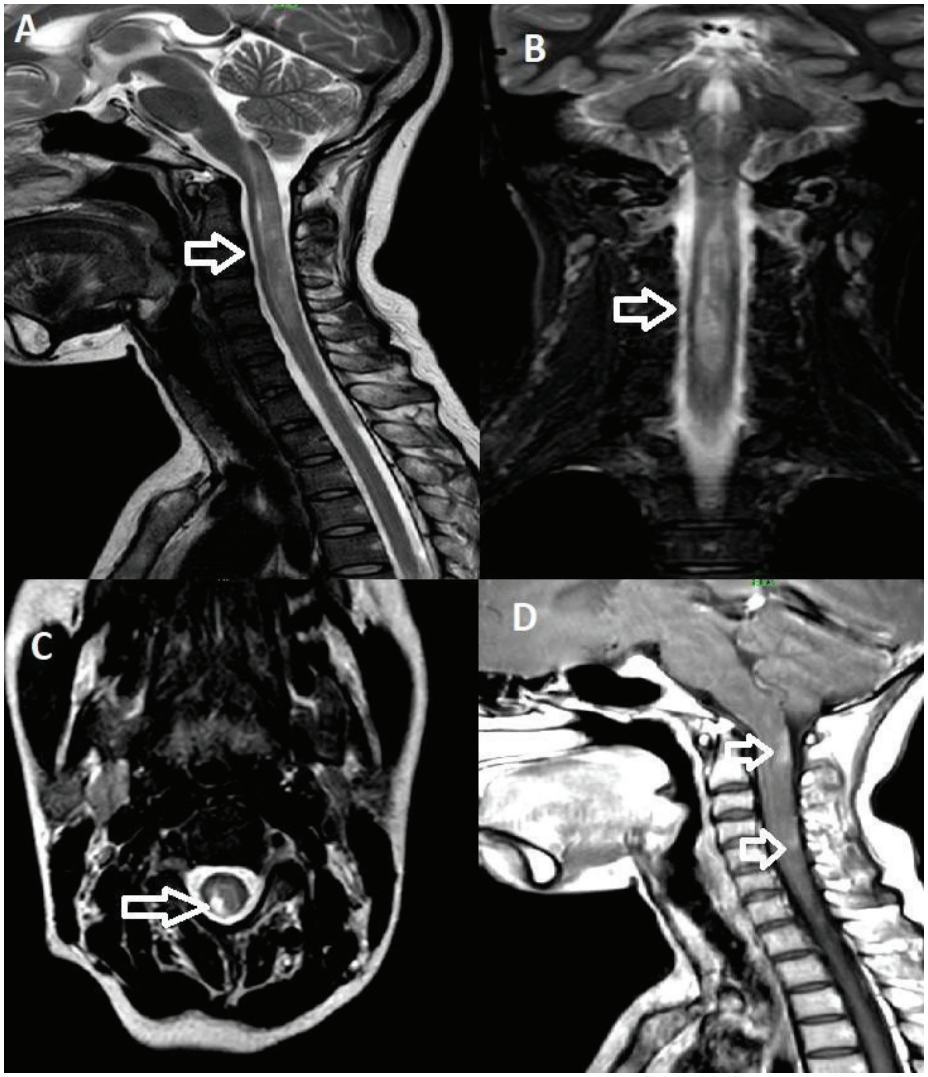

Figure 1 :MRI of the spine a.T2 sagittal image showing longitudinally extensive transverse myelitis B. T2 coronal image showing the same C.T2 Axial image showing the cord hyperintensity(arrow) D. Post contrast image showing patchy enhancement

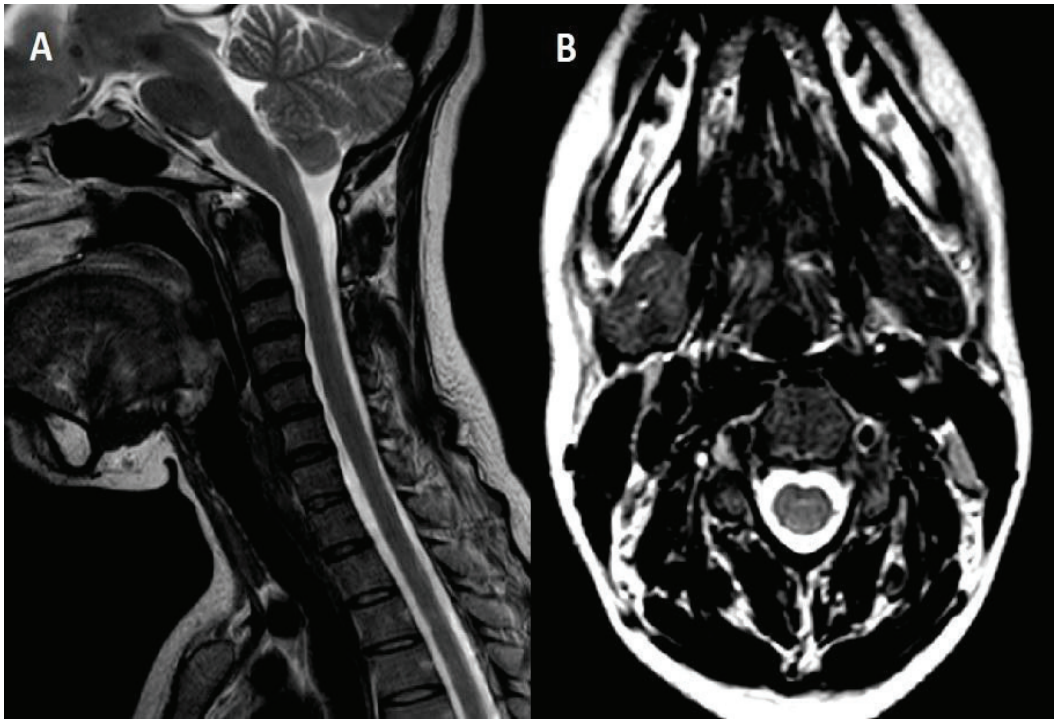

Figure 2- Post treatment images after 3 months A.T2 sagittal and B. T2 axial images showing resolution of signal changes

Discussion

Neuromyelitis optica is an autoimmune demyelinating disease that mainly involves the spinal cord, brain and optic nerves[6]. The cardinal clinical features of this disorder are optic neuritis and longitudinally extensive transverse myelitis which can occur either simultaneously or as separate events[7].

In 2006, the international consensus diagnostic criteria for NMOSD were postulated. There were two definitive criteria i.e., transverse myelitis and optic neuritis, and two of the following: brain MRI that is not typical for multiple sclerosis or AQP4-IgG positivity, or a large spinal cord lesion that exceeds three spinal segments. In the year 2015, these criteria were revised by an expert panel. According to the new criteria, one main clinical finding along with Aquaporin 4 antibody positivity is considered enough for diagnosis. However, the other common differential diagnoses have to be excluded by appropriate investigations[8]. The main clinical findings are as follows: (1) optic neuritis, (2) acute transverse myelitis, (3) area postrema syndrome, (4) acute brainstem syndrome, (5) acute diencephalic syndrome, (6) symptomatic cerebral syndrome. The antibody specific to the AQP4 water channels associated with NMOSD is considered to be an accurate serum marker for pathology [9].

The typical radiological feature of an NMO cord lesion is usually a contiguous spinal cord lesion three or more segments in length with centrally predominant distribution.

Our patient had evidence of a LETM and NMO was one of the main differential diagnosis considered. Her blood results showed strong positivity for Aquaporin 4 antibody and hence she was started on immune modulation.

Neuromyelitis Optica is a disease with relapsing course in most patients and needs to be treated promptly in both acute and chronic phases. In an acute attack, pulse steroid therapy is considered the first-line therapy of choice. If the patient does not respond to steroid therapy, plasmapheresis, intravenous immune globulin (IVIG) as well as cytotoxic drugs including Azathioprine, Mycophenolate and Rituximab may be used[10] . In our case, clinical improvement was observed after the administration of pulse steroids (1g/day of methylprednisolone for five days). Thereafter the patient was discharged from the hospital with a tapering schedule of prednisolone and Mycophenolate as maintenance. Her repeat imaging at three months showed resolution of lesions. She is on regular follow up.

Conclusion

NMO is an important cause of cervical cord demyelination in young females. Long segment involvement of the cord is a characteristic feature which should alert the clinician to the appropriate diagnosis. Early diagnosis and treatment leads to good long term outcomes. Because of the relapsing nature of the disease, long term immune modulation may be needed in patients with antibody positivity.

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