

Case Report:

Progression of Acute Transverse Myelitis to Multiple Sclerosis

Dr. Abeetha S¹, Dr. Mitali Srivastava², Dr. Shivali Srivastava³

1- Post graduate, Department of Physiology, SMVMCH, Pondicherry- 605107

2- Post graduate, Department of Biochemistry, SMVMCH, Pondicherry- 605107

3- Post graduate, Department of Anatomy, SMVMCH, Pondicherry- 605107

Corresponding author: Dr. Abeetha S

Abstract:

Herewith we reported a case of 16 year old girl, with clinical as well as radiological findings of acute transverse myelitis, which later progressed to multiple sclerosis. The discussion entails findings in acute transverse myelitis and multiple sclerosis, differences between them and diagnostic criteria of afore mentioned diseases.

Keywords: Acute transverse myelitis, Elsberg phenomenon, Multiple sclerosis, Devick's Syndrome

Introduction:

Neurological disease in the young, a source of agony and burden for the patient as well as caretakers, is not very uncommon in India. Transverse myelitis is inflammation of the spinal cord in turn causing axonal demyelination¹. The term is self-explanatory, where 'transverse' refers to the lesion affecting the width of the spinal cord and 'myelitis' indicates inflammation. Recurrent episodes of transverse myelitis lead to scarring that interferes with neurotransmission².

The classical clinical findings seen in acute transverse myelitis are acute onset muscular weakness, low back ache, urinary retention followed by loss of bowel and bladder control, paraesthesia and at times, even paralysis, all of which indicate abnormal spinal cord function². Regardless of the treatment, patients sometimes recover with minor disability or may continue to live with lasting impairments. Acute transverse myelitis is mostly a one-time episode; however in certain cases it may also recur or progress to other deteriorating diseases like multiple sclerosis². A study by Calvo et al. conducted in Spain, concluded that 13% patients of idiopathic acute transverse myelitis progressed to multiple sclerosis over an

average period of 2.9 years. They also stated that onset of symptoms at an early age was strongly related with progression to multiple sclerosis³.

Multiple sclerosis is also a differential diagnosis of acute transverse myelitis, the others being neuromyelitisoptica and transverse myelitis due to infective and inflammatory etiologies such as herpes zoster, herpes simplex, systemic lupus erythematosus and neurosarcoidosis¹. In the following case report and discussion, an effort will be made to explain the progression of acute transverse myelitis to multiple sclerosis.

Case report:

A 16 year old female was brought in for consultation with complaints of headache and neck pain which was sudden in onset followed by weakness of both upper and lower limbs of the left side after a fall. These events took place 20 days before admission. However, slow but consistent improvement was noted by the patient. The patient gave history of difficulty in initiation of micturition over the last 20 days. On further elicitation of history, the patient revealed she had a loss of appetite and weight also. Patient was not known to have any similar history in the past. There was no history of loss of consciousness/ seizures/ ENT

bleed/ visual disturbances/ prior hospitalizations. The patient's family had no related history. During her course of stay in the hospital she developed sudden loss of bowel and bladder control 4 days after admission.

On examination, she was afebrile but pale. Her vitals were stable (PR 90/ min; BP 116/ 70 mm of Hg). The Central Nervous System examination revealed intact higher mental functions and cranial nerves. Motor system was found to be affected, with the power of left upper limb and both lower limbs being 1/5 and 4/5 respectively. Hypertonia was noted in all limbs except right upper limb. While deep tendon reflexes were also exaggerated in all limbs except right upper limb. Elsberg phenomenon was present. Impairment of pain and temperature over C3 and C4 dermatome was observed. Hyperaesthesia was also noted. Well sustained ankle clonus on the left and ill sustained on the right were also observed. Patient was provisionally diagnosed as a case of left hemiparesis/ Stroke in young for evaluation.

Since the patient was stable, she was immediately taken up for MRI (Magnetic Resonance Imaging) with screening of spinal cord, which showed hyperintense signal changes in C1 to C3 cervical spine indicating demyelination. Mild volume loss of the thoracic spinal cord was also noted. Thoracolumbar screening showed L3- L4 left posterolateral and L4- L5 mild diffuse disc bulge. Taking into consideration the opinion of the radiologist, lumbar puncture for CSF (Cerebro Spinal Fluid) analysis was done under aseptic precautions. The results did not reveal any significant findings. Oligoclonal bands were not found in the CSF sample. The CSF total protein was found to be low i.e. 7.3mg/ dl (normal: 15- 45 mg/ dl), while CSF sugar was within normal limits i.e. 54 mg/ dl (normal 40- 85 mg/ dl). Opinion was obtained from a neurologist

who stated that the patient had neurogenic bladder and suggested that the patient might be suffering from a spinal cord lesion or even initial stages of multiple sclerosis. The patient was then referred to an ophthalmologist for opinion to rule out Devick's syndrome (Neuromyelitisoptica). No ocular manifestations of neurological disorders were noted. With the available reports and opinions of consultants of various departments, the patient's diagnosis was modified to Quadriparesis/ Demyelinating acute transverse myelitis.

The mainstay of treatment remained systemic and oral steroids as well as multivitamins and physiotherapy. Methyl prednisolone 1 gm in 500 ml NS was given intra venous over a period of 1 hour on the day of admission followed by 500 mg IV OD for 5 days. From the 6th day the patient was started on tablet prednisolone, 50 mg OD for 6 days, which was subsequently tapered to 40 mg for the consecutive 2 days. The patient was discharged at request but advised to come for follow up a week later for tapering the steroid dose. Multivitamins used for her treatment were Methylcobalamin IM OD, B Complex IM OD and Vitamin E capsules 400 mg HS for 2 weeks.

Discussion:

The incidence of demyelinating diseases in our country is 0.22%, and two thirds of these patients suffer from demyelinating diseases other than classical multiple sclerosis⁵.

Few of the diagnostic criteria for acute transverse myelitis are as follows⁵:

- Sensory, motor, or autonomic dysfunction attributable to spinal cord lesion
- Bilateral signs and/or symptoms (though not necessarily symmetric)
- Clearly defined sensory level
- Exclusion of extra-axial compressive etiology by neuroimaging

- Inflammation within the spinal cord demonstrated by CSF pleocytosis or elevated IgG index or gadolinium enhancement

As stated earlier, early onset acute transverse myelitis has a higher chance of progressing to multiple sclerosis, which can be diagnosed by various criteria, like McDonald's criteria. Neuro-imaging as well as presentation of a clinically isolated syndrome suggestive of multiple sclerosis such as unilateral optic neuritis, internuclearophthalmoplegia, partial myelopathy are included in this criteria⁵.

Transverse myelitis limits itself to the spinal cord without involvement of central nervous system. Demyelination, axonal loss and involvement of gray matter are also seen. In transverse myelitis there is uniform, central and symmetrical involvement of the spinal cord, whereas it is patchy and peripheral in multiple sclerosis⁸.

Neurons are unique as they do not divide after birth (with the exception of bipolar olfactory neurons), which is why, inflammation or damage to the spinal cord, causes formation of scar tissue that interrupts the network of neuronal communication from the point of damage. Affection of the spinal cord at a particular level involves loss of function not only of the corresponding segment, but also of those below it.

In a study by Alvarenga et al. it was stated that MRI of the spinal cord showed inflammation in 92% of the patients suffering from acute transverse myelitis due to various etiologies. 61% of the patients had extensive spinal cord damage. In majority of the patients (94.4%) MRI Brain was normal. CSF analysis revealed oligoclonal bands in 38% of those who were tested⁶. In the present case report, though the MRI brain was normal, oligoclonal bands were not found in the CSF

analysis, which if present, indicate a poor prognosis.

In yet another study by Harzheim et al. it was recorded that in 22% of patients, acute transverse myelitis was the foremost manifestation of possible multiple sclerosis. In such cases involvement of a single segment of spinal cord was more common, while multi segmental involvement was seen in acute transverse myelitis due to other causes. They also stated that signal changes were seen in MRI spinal cord of 96% of the patients. Peripheral nervous system involvement was seen in 27% of patients with acute transverse myelitis, but not in patients progressing to multiple sclerosis⁷. In the patient under discussion MRI Brain with screening of spinal cord showed hyper- intense signal changes in C1 to C3 cervical spine and peripheral nervous system involvement was absent.

The patient had sensory, bilateral motor as well as autonomic signs and symptoms (neurogenic bladder) with a clearly defined dermatome i.e. C3, C4. Elsberg phenomenon was also seen in the patient. It can be described as weakness of the limbs occurring in order of ipsilateral arm, ipsilateral leg followed by contralateral leg and finally contra lateral arm. It is often referred to as a 'U' shaped or clockwise involvement of limbs, and is classical of cervical myelopathy⁵. There was no ocular involvement in the patient, which helped to rule out Devick's disease (Neuromyelitisoptica).

The patient initially had all features suggestive of acute transverse myelitis, later it was noted that it was probably the first attack of multiple sclerosis, as she had a clinically isolated syndrome (partial myelopathy) and lack of peripheral nervous system involvement. However, radiological investigations showed no abnormal findings in the brain, which is possible in the initial stages of multiple sclerosis. In a study by Palace J, it has been stated that a normal

MRI brain following a clinically isolated episode has an 11% risk of developing multiple sclerosis over the consecutive 10 years⁹.

Immediate initiation of treatment and absence of oligoclonal bands indicated a good prognosis, which is perhaps why the patient improved slowly but steadily.

In spite of various available criteria, it is challenging to make a clear-cut diagnosis of transverse myelitis progressing to multiple sclerosis. The patient under discussion apparently

suffered from her first episode of multiple sclerosis, which closely mimics acute transverse myelitis. While, acute transverse myelitis is usually a one-time episode, multiple sclerosis is a deteriorating disease which recurs and eventually debilitates the patient unhinging their daily life to a great extent. Thus, it is essential to diagnose and differentiate the two conditions from each other. In case of suspected multiple sclerosis, the patient can be asked to come for follow ups there by clinching the diagnosis at the earliest.

References:

- 1) Jacob A, Weinshenker BG. An Approach to the Diagnosis of Acute Transverse Myelitis. *Semin Liver Dis* 2008;28(1):105-20.
- 2) National Institute of Neurological Disorders and Stroke [Internet]: Bethesda:National Institutes of Health; 2014 [updated 2014 April 16; cited 2014 June 5]. Available from: http://www.ninds.nih.gov/disorders/transversemyelitis/detail_transversemyelitis.htm
- 3) Calvo AC, Mañé MA, Martínez, Alentorn-Palau A, Escuer JB, Pinel LR et al. Idiopathic acute transverse myelitis: outcome and conversion to multiple sclerosis in a large series. *BMC Neurology* 2013;13(1):135.
- 4) Mathew NT, Mathai KV, Abraham J, Taori GM. Incidence and pattern of demyelinating disease in India. *Journal of the Neurological Sciences* 1971;13(1):27–38.
- 5) Miller DH, Weinshenker BG, Filippi M, Banwell BL, Cohen JA, FreedmanMS et al. Differential diagnosis of suspected multiple sclerosis: a consensus approach. *Multiple Sclerosis* 2008;14:1157–74.
- 6) Alvarenga MP, Thuler LCS, Neto SP, Vasconcelos CCF, Camargo SG, Alvarenga P et al. The clinical course of idiopathic acute transverse myelitis in patients from Rio de Janeiro. *Journal of Neurology* 2010;257(6):992-8.
- 7) Harzheim M , Schlegel U, Urbach H, Klockgether T, Schmidt S. Discriminatory features of acute transverse myelitis: a retrospective analysis of 45 patients. *Journal of the Neurological Sciences* 2004;217(2):217-23.
- 8) Awad A, Stüve O. Idiopathic Transverse Myelitis and NeuromyelitisOptica: Clinical Profiles, Pathophysiology and Therapeutic Choices. *Current Neuropharmacology* 2011;9:417-28.
- 9) Palace J. Making The Diagnosis Of Multiple Sclerosis. *J NeurolNeurosurg Psychiatry* 2001;71(1):3-8.

Date of submission: 29 March 2014

Date of Publication: 09 June 2014

Source of support: Nil; Conflict of Interest: Nil