Lower motor neuron paralysis with extensive cord atrophy in parainfectious acute transverse myelitis

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Abstract

We describe a young patient of acute transverse myelitis (ATM) who developed true lower motor neuron (LMN) type flaccid paraplegia as a result of anterior horn cell damage in the region of cord inflammation that extended from conus upwards up to the D4 transverse level. We infer that flaccidity in acute phase of ATM is not always due to spinal shock and may represent true LMN paralysis particularly if the long segment myelits is severe and extending up to last spinal segment.

Key Words

Acute disseminated encephalomyelitis, acute transverse myelitis, ascending myelitis, magnetic resonance imaging, parainfectious myelitis

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A 17-year-old male patient developed visual impairment, abnormal behavior, seizures and coma within a period of five days. Two weeks later, on regaining consciousness he was found to have complete sensory-motor flaccid paraplegia with bladder/bowel involvement and sensory level at D4. MRI showed multiple large sub-cortical white matter lesions in the brain along with continuous long segment lesion (C4 to conus medullaris) in the spinal cord [Figure 1], suggesting acute disseminated encephalomyelitis (ADEM).^[1,2] Visual loss recovered partially in next 6 months but the paraplegia remained complete and flaccid even after five years now. Present MRI showed normal brain but thread-like thin spinal cord extending from D3 downwards up to conus medullaris [Figure 1].

ADEM may cause cerebral as well as spinal cord lesions.^[1-4] Sometimes optic nerves may also get involved. Our patient had evidence of involvement of all the three sites. The spinal cord involvement in these patients is in the form of acute transverse myelitis (ATM). Flaccid paraplegia is known to occur in ATM and this flaccidity is generally attributed to spinal shock.

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However, at times it may not actually be a spinal shock but true lower motor neuron disorder due to involvement of anterior horn cells in the regions of inflammation in the spinal cord. In this situation the flaccidity would persist even after several weeks of follow-up.^[5] This type of acute transverse myelitis (ATM) described in one study as parainfectious ascending myelitis (PIAM) has continuous inflammation of the cord from conus medullaris to transverse sensory-motor level.^[6] The

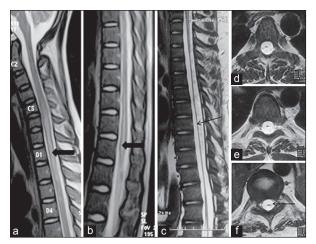


Figure 1: T2 weighted cervical (a) and dorso-lumbar (b) MRI of the patient showing continuous hyper-intense signal from C4 to conus medullaris (thick arrows). MRI done 5 years later shows thread-like atrophic spinal cord (thin arrow) extending from D3 to conus medullaris in sagittal T2 (c) and three different dorsal planes of axial T2 (d, e, f) weighted images

term spinal shock in this type of ATM remains a misnomer as there is a true extensive involvement of lower motor neurons (LMN) within the spinal cord.^[6] Contrary to common belief, some patients with PIAM-type ATM who develop flaccid paralysis attributable to spinal shock, never develop upper motor neuron signs during the course of illness and carry a bad prognosis. The present case is a rare form of ADEM where the cerebral and optic nerve lesions were mild and showed near-complete recovery but the spinal cord lesion (ATM) was severe and showed no recovery. In the present case, threadlike spinal cord five years after acute inflammation in the same region is a true demonstration of extensive LMN disorder in PIAM- type ATM.

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