IF : 3.62 | IC Value 80.26

Volume-5, Issue-12, December - 2016 • ISSN No 2277 - 8160

Original Research Paper

Medicine

Acute Transvere Myelitis Following Japanese Enchephalitis Viral Infection : A Rare Presentation Of The Infection

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ABSTRACT *Japanese encephalitis (JE) is an important cause of epidemic encephalitis in southeast Asian countries. A huge population living in the endemic regions makes this disease a global health concern. JE is caused by Japanese encephalitis virus (JEV), a mosquito-borne virus, Single stranded positive sense RNA virus belonging to the genus Flavivirus (family Flaviviridae). WHO estimated that approximately 67,900 JE cases typically occur annually in the 24 JE-endemic countries, for an incidence of 1.8 per 100 000 overall. In India, many outbreaks of JE have been reported since 1955. [1] Apart from encephalitis, immune-mediated manifestation as acute transverse myelitis (ATM) have been rarely previously reported in JE. The case mentioned here is a male patient presented with acute-onset bilateral lower limb weakness followed by upper limb weakness without any signs of altered mentation. MRI cervico-dorsal Spine findings suggested Signal Intensity Alterations from the C2 to D10 levels. Corticosteroid pulse therapy for 5 days showed Improvements in power with complete recovery of other symptoms as well.*

KEYWORDS : ATM, Japanese Encephalitis Virus

INTRODUCTION

During recent years, majority of cases of the epidemic came from eastern Uttar Pradesh (Gorakhpur and adjoining areas). [2] The mortality of JE ranges between 20% and 40%. Serious neurological sequelae are seen in 33-50%. [3]

The reported clinical presentation of JE include altered sensorium, convulsions, headache, hyperkinetic movements and brain stem involvement features such as opsoclonus, gaze palsies and pupillary changes. However, immune-mediated demyelinating neurological manifestation as acute transverse myelitis (ATM) have been rarely reported previously in JE. We describe a patient who developed ATM following an infection with JE virus. This case report has its importance in view of potential therapeutic implications of this complication of JE, which was not expected previously.

CASE PRESENTATION

A 14-year-old Teenager presented with complaints of weakness in both lower limbs which started 3 days prior and progressed to weakness of both upper limbs the next day. The weakness in upper limbs was mild and distal, compared to lower limbs in which he had a complete paralysis. He had fever and headache about 2 weeks prior to the onset of weakness. There was no history of trauma, vaccination or similar attack previously. The patient did not develop any bladder or bowel incontinence.

Examination showed normal vital parameters and higher mental functions. Cranial nerves examination was normal. Muscle tone was normal in upper limbs and reduced in lower limbs. The power was MRC (Medical Research Council) grade 4/5 at shoulder, elbow and wrist and hand grip was weak bilaterally. In the lower limb the power was grade 2/5 at all joints. Deep tendon reflexes were present in upper limbs but absent in lower limbs. The abdominal and cremasteric reflexes were absent. Planters were bilaterally extensors. There was a sensory loss below C5 for all modalities of sensation.

INVESTIGATIONS

He had normal haematological, biochemical and thyroid parameters. Cerebrospinal fluid (CSF) analysis depicted a cell count of 65/mm^3 (all lymphocytes), protein—118 mg %, sugar—67 mg %. no organisms were visualised by direct microscopy using Gram-negative and acid-fast bacilli stains. The Serology for Dengue virus was positive. Dengue IgM Titres were Elevated in both CSF and Serum. The serology for Chikangunya virus, measles, mumps, Hepatitis virus, HIV, Epstein-Barr virus and cytomegalovirus was negative. The serum and CSF ELISA for JE carried out with JE IgM COMBO ELISA (Panbio, Australia) showed elevated IgM antibody titres in serum and CSF (serum- 27 PBU (Panbio units) against 11 PBU as the upper limit of normal), CSF- 37.1 PBU against 11 PBU as the upper limit). Serum aquaporin-E antibody for neuro myelitis optica (NMO) was negative. MRI of cervico-thoracic spine demonstrated signal intensity alterations, hyperintense on T2-weighted image and hypointense on T1-weighted image, extending from C2 to D10 spinal segments without any obvious post-contrast enhancement. Changes were also well depicted in axial images. MRI Brain did not reveal altered signals.

TREATMENT

The diagnosis of ATM was made and the patient was treated with intravenous methyl prednisolone 1 g daily for 5 days. The patient received physiotherapy followed by gait training on recovery of power for rehabilitation. The patient was also given nutritional supplements and supportive medicines.

OUTCOME AND FOLLOW-UP

The patient did not show any improvement in power during the first 2 days of treatment. Towards the 3rd day, the patient showed improvements in power. Suspecting anterior horn cell involvement, an electromyography (EMG) was done, which showed no

spontaneous activity. The patient regained normal power and other myelopathic features were reversed at 3 months follow-up. The repeat MRI study demonstrated complete resolution of the lesion.

DISCUSSION

The important presenting features of JE are altered sensorium, convulsions, headache, hyperkinetic movement disorders, features of brain stem involvement as opsoclonus, gaze palsies and pupillary changes, dystonia, decerebrate rigidity, paralysis and seizures. [4] Unusual features of JE include respiratory paralysis, oromandibular dystonia, acute flaccid paralysis and hemiplegia with dysarthria. Differential Diagnosis are meningitis, Febrile convulsions, Reye's Syndrome, Rabies, Cerebral Malaria, Toxic Encephalopathy. However, there are few previous case reports of immune-mediated demyelinating neurological syndrome as ATM following JE. [5]

Transverse myelitis results from inflammation of complete thickness of the spinal cord leading to loss of motor, sensory and autonomic functions below the involved spinal cord segment. The diagnosis of ATM in our patient was made on the basis of a typical clinical and radiological picture. Preceding JE virus infection was mild, manifested by fever and headache 2 weeks prior to the onset of weakness. It was confirmed by positive IgM antibody in the serum and the CSF to JE virus which has very high sensitivity and specificity. [6] The aetiology of ATM is varied, including viral infections, autoimmune disorders such as systemic lupus erythematosus, vasculitis, multiple sclerosis and following vaccination. Various viral infections associated with ATM include herpes simplex virus, Varicella zoster virus, Epstein-Barr virus, cytomegalovirus, enteroviruses, HIV, influenza and rabies virus. [7] ATM associated with viral infection is mostly the result of immunological injury to spinal cord rather than direct viral invasion.

The phenomenon of acute flaccid paralysis has been identified in JE patients. This manifests as asymmetric weakness, predominantly involving the lower limbs. This was described to be due to anterior horn cell lesion. Anterior horn cell involvement in JE has been proved on the basis of prominent fibrillation and neurogenic changes in the wasted muscles on EMG. However, our patient did not have any such evidence of anterior horn cell involvement on EMG. Thus, his weakness cannot be explained by anterior horn cell involvement and was purely due to ATM. As JE infection has been rarely previously reported to cause ATM, it is difficult to provide a probable explanation for this complication on the basis of current literature. It is likely that the neuronal damage and disruption of blood–neuron barrier secondary to inflammation lead to exposure of neuronal antigen to immune responsive elements, further mounting an antineuronal autoimmune response, which leads to neuronal damage manifesting as ATM. Furthermore, a delay of 3 weeks between JE virus infection and ATM favours an immune-mediated insult to the spinal cord. However, further studies into the pathogenesis of neurological manifestations in JE are needed to explain such presentation.

To conclude, our case report suggests that in a case of JE, one should be vigilant for early diagnosis of possible immune-mediated demyelinating syndrome such as ATM, for consideration of an early institution of immunomodulator therapy to prevent adverse consequences.

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