

Japanese encephalitis presenting as transverse myelitis: An uncommon presentation

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ABSTRACT

Japanese Encephalitis Virus (JEV) is the main cause of viral encephalitis in South East Asia. Commonly, it presents as an acute encephalitic syndrome with fever, headache, seizures, and altered sensorium as clinical manifestations. However, there can be atypical presentations such as acute transverse myelitis (ATM) as the initial manifestation. Clinicians should be aware of such possibilities and myelitis due to the JE virus should be considered as a differential in children presenting with encephalomyelitis.

Key words: Acute transverse myelitis, Japanese encephalitis, Paraparesis

Japanese Encephalitis Virus (JEV) is the leading cause of viral encephalitis in South East Asia [1]. Globally, there are an estimated 68,000 clinical cases of JEV every year. The case fatality is as high as 30%, and long-term neurological or psychiatric sequelae can occur in 30–50% of patients [2]. In India, a country with a high burden of diseases caused by JEV, about 14–18% of reported cases of acute encephalitic syndrome are due to JEV [3]. The reported clinical manifestations include fever, headache, seizures, altered sensorium, hyperkinetic movements, and features of brain stem involvement such as opsoclonus, gaze palsies, and pupillary changes [4]. The unusual clinical presentations described in the literature are oromandibular dystonia, anterior horn cell involvement, acute flaccid myelitis, post-infectious transverse myelitis, and Guillain Barre Syndrome [5-8].


Acute transverse myelitis, as the initial presentation of Japanese encephalitis, is seldom reported. Herewith, we report a case of Japanese encephalitis in a child with paraplegia as the initial manifestation.

CASE REPORT

A 10-year-old previously healthy female child got admitted with fever and weakness of both lower limbs for 2 days. There was no history of back pain, girdle pain, sensory disturbances, or bladder involvement. No history of trauma, recent exanthem/vaccination either.

On examination, the child was febrile (100°C), conscious, and oriented with intact cranial nerves. She had flaccid paraplegia with absent deep tendon reflexes and mute planters. The upper limbs were normal. There was no sensory level or spinal tenderness. Kerning's sign was positive. Fundi were normal. Her pulse rate was 98/min, respiratory rate 24/min, and blood pressure 100/70 mm of Hg. A diagnosis of acute flaccid paralysis-Guillain-Barre Syndrome/acute flaccid myelitis/acute transverse myelitis was made and investigations were done.

Motor and sensory conduction were normal in all four limbs. As nerve conduction studies might be normal in Guillain-Barre Syndrome in the 1st week and because intravenous immunoglobulin would be useful in transverse myelitis as well, intravenous immunoglobulin was started. Sensorium worsened on the next day, and she developed decerebrate posturing. Glasgow's coma score (GCS) was E1VTM1 and the patient was intubated and ventilated. Meningitic dose of antibiotics – Ceftriaxone 100 mg/kg/day, acyclovir–30 mg/kg/day in three divided doses, and hypertonic saline-5 ml/kg/dose 3 times a day, were added considering acute encephalitic syndrome. A complete hemogram showed neutrophilic leukocytosis with a total count of 12450 cells/mm³, C-reactive protein was elevated (12 mg/l), and blood culture was sterile. Metabolic parameters were normal. Computed tomography (CT) brain was normal. Cerebrospinal fluid (CSF) analysis done on the 3rd day of admission, depicted no cells, protein-40 mg/dl, sugar-52 mg/dl, and sterile culture. She developed generalized seizures on the fourth day and needed anticonvulsants for control. As the child developed encephalopathy and seizures,

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acute myelomeningocele encephalitis versus acute disseminated encephalomyelitis (ADEM) was considered as differentials.

Meanwhile, she developed bilateral pneumothorax as a complication of mechanical ventilation which necessitated intercostal drainage on both sides. Magnetic resonance imaging (MRI) brain done on the 8th day showed T2, FLAIR hyperintensities in both thalami with diffusion restriction suggestive of flavivirus infection. There was no contrast enhancement (Fig. 1a-c). Serum tests were negative for malaria, dengue, typhoid, leptospira, scrub typhus, and COVID. CSF virology was negative for herpes, enterovirus, varicella, measles, Epstein Barr, Cytomegalo, and West Nile viruses. CSF gene Xpert for tuberculosis was also negative. However, both serum and CSF turned out to be positive for IgM against the Japanese B virus. She developed Gram-negative sepsis (acinetobacter) and appropriate antibiotics were added.

Her GCS improved to E4VTM4 on the 12th day and started recognizing her parents. She was able to move her upper limbs against gravity. But both lower limbs remained flaccid were flexic with zero power. MRI Spine screening revealed high signal intensity in the lumbosacral cord (Fig. 1d). A final diagnosis of encephalomyelitis due to the JE virus was established. Though her sensorium improved, she remained paraplegic. Hence, a repeat course of intravenous immunoglobulin was tried. However, there was no improvement and she succumbed to ventilator-associated pneumonia in the 3rd week of her illness.

DISCUSSION

Japanese Encephalitis is caused by the JE virus, a mosquito-borne flavivirus. The estimated global incidence is 1.8 per

100000 [1]. It causes acute encephalitis syndrome characterized by fever, headache, convulsions, altered sensorium, and brainstem signs with pyramidal and extrapyramidal features. Atypical presentations such as acute flaccid myelitis, post-infectious transverse myelitis, and Guillain-Barre Syndrome have been reported in the literature [6-8]. Inflammation-mediated damage to the spinal cord is not a common feature of Japanese encephalitis. There are very few case reports to date [9,10].

Although Guillain-Barre syndrome was considered as a differential at admission due to flaccid quadriplegia and areflexia, it was excluded on day 2 as the child developed encephalopathy and then seizures. Development of encephalopathy and seizures ruled out the possibility of acute flaccid myelitis as well. Spinal cord involvement can occur in ADEM, along with encephalopathy and seizures, but the MRI brain was not suggestive of ADEM. Neuroimaging revealed bilateral thalamic involvement characteristic of flavivirus infection. CSF IgM-specific ELISA for JE virus was positive confirming the diagnosis of Japanese encephalitis in our patient. Serum and CSF for other flaviviruses - Dengue and West Nile viruses were negative. Bilateral thalamic hyperintensities can also occur in deep venous sinus thrombosis, which was ruled out by normal magnetic resonance venography.

Nandan *et al.* have described a 13-year-old adolescent boy who presented with fever, sensory-motor paraparesis which progressed to quadriplegia, and acute onset altered sensorium in whom neuroimaging showed abnormal cord intensities in the entire spinal cord. Our child had a similar clinical presentation, but cord involvement was restricted to conus medullaris [9]. Mohta *et al.* have reported a case of a young male with altered sensorium and quadriplegia and were diagnosed to have JE with longitudinally extensive transverse myelitis whose MRI spine showed altered signal intensities in the cervical cord (C2–C6). The patient did not show improvement and succumbed to ventilator-acquired pneumonia [10]. Our patient too had a similar downhill course despite immunotherapy and symptomatic care.

The role of steroids in idiopathic transverse myelitis is well documented, but there are few such records in the case of acute infectious myelitis [10]. Treatment for Japanese encephalitis is mainly supportive. Studies show that IVIG may prove to be useful in Japanese encephalitis due to its immune-modulation effect [11]. Although we tried two courses of intravenous immunoglobulin, we did not find improvement in the clinical status of our patient. Underlying gram-negative sepsis resisted a trial of high-dose steroids or plasmapheresis. The mortality of JE ranges between 20% and 40% [12]. Our child remained on ventilatory support and succumbed to ventilator-associated pneumonia in the 3rd week of illness.

CONCLUSION

Transverse myelitis can be a rare initial presentation of Japanese encephalitis. Clinicians should be aware of this uncommon presentation of a common disease, especially in an endemic area.

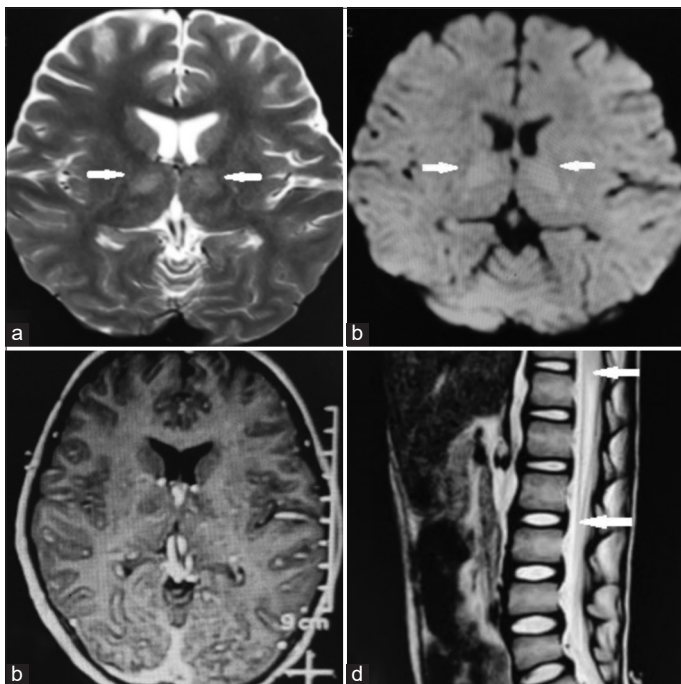


Figure 1: (a) MRI brain axial T2W image shows hyperintensities in bilateral anterior thalami. (b) Diffusion-weighted imaging shows diffusion restriction. (c) T1 axial contrast imaging shows no contrast enhancement. (d) MRI spinal cord sagittal T2W image shows high signal intensity in lumbosacral cord

Clinical progression to develop encephalopathy, involvement of bilateral basal ganglia, and thalami in neuroimaging could aid in the diagnosis.

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