VIDEO CME

Multifocal Myoclonus in Dengue Encephalopathy

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Abstract

Introduction: Myoclonus is an involuntary, jerky body movement that is a sign of encephalopathy due to various underlying pathologies, one of which is dengue encephalopathy. Often, these movements are subtle and suppressed by the frequent use of sedatives and paralytic agents.

Case report: A 67-year-old male diagnosed with dengue encephalopathy presented with involuntary jerky movements of his face, which were confirmed as orofacial multifocal myoclonus by electroencephalogram (EEG) studies.

Conclusion: It is important to recognize myoclonic movements and look for an underlying cause.

Keywords: Dengue encephalopathy, Dengue neurological complications, Electroencephalogram delta waves, Electroencephalogram spike waves, Myoclonus, Orofacial dyskinesia.

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Introduction

Myoclonus is the second most common involuntary movement observed in intensive care units after tremors.[1] There are multiple causes of myoclonus, one of which is dengue encephalopathy. Neurological complications are seen in an estimated 4.2–13% cases of dengue fever and include direct neurotropic complications leading to encephalitis and myositis; systemic neurological complications or indirect effects due to metabolic complications resulting in encephalopathy, and cerebrovascular complications due to thrombocytopenia as well as platelet dysfunction and hypokalemic paralysis; and immune-mediated neurological complications, which include acute disseminated encephalomyelitis, Guillain–Barré syndrome, and opsoclonus myoclonus syndrome.[2,3] Multifocal myoclonus has been commonly reported in children, but rarely reported in adults. Here, we report a case of dengue encephalopathy with multifocal myoclonus.

Case Description

A 67-year-old gentleman, known hypertensive and diabetic, presented with history of fever for 1 week with thrombocytopenia. Prior to this, he had been diagnosed with dengue and symptomatically treated at his hometown with paracetamol and intravenous fluids, and received a transfusion of single donor platelets. After 4 days of stay in hospital, he had deterioration in his sensorium and was hence referred to a higher center.

On arrival, his vital parameters were normal with heart rate of 90 beats/min, respiratory rate of 28 breaths/min, blood pressure of 120/70 mm Hg, and oxygen saturation of 90% on room air. On examination, he was restless and disoriented, with a Glasgow Coma Scale (GCS) score of E2M5V2, for which he was intubated and ventilated with lung-protective ventilation. He showed signs of orofacial dyskinesia which worsened with pain stimuli, which was opined by neurology as orofacial multifocal myoclonus (as shown in the video). Both computed tomography and magnetic resonance imaging of brain were normal. Laboratory studies showed platelet count of 39,000 cells/mm,[3] hematocrit of 37%, total bilirubin of 3.96 mg/dL, aspartate transferase of 83, alanine transferase of 80, and serum creatinine of 2.07 mg/dL. Dengue serology was positive for IgM antibody. Blood gas analysis showed severe metabolic acidosis. Electroencephalogram changes (Figs 1 and 2) were seen which were suggestive of cortical myoclonus. Figure 1 shows periodic generalized spike wave discharges with background slow activity in delta range, and Figure 2 shows intermittent rhythmic delta activity and generalized spike wave discharges, suggestive of multifocal myoclonus. He was managed with fluid resuscitation and antipyretics. In view of persisting oliguria and worsening serum creatinine along with worsening metabolic acidosis, he was initiated on hemodialysis. Initially, the myoclonus required levetiracetam and sedation with midazolam to be controlled, and completely subsided spontaneously after about 5 days in intensive care unit. However, his sensorium did not improve, necessitating a tracheostomy. Following this, he remained comatose with GCS score of E1M3Vt, oliguric, on alternate day hemodialysis, and unable to be weaned off mechanical ventilation. He developed intercurrent infections which were treated with appropriate antibiotics, but unfortunately, expired 10 days later from septic shock.

Discussion

Myoclonus is an abnormal movement characterized by brief, sudden, abrupt twitching involving the face, extremities, and trunk caused by an abnormally increased excitability of neurons leading to muscle contractions called positive myoclonus or a sudden brief loss of muscle tonus followed by a rapid recovery of tonus called negative myoclonus.[4] As in the present case, myoclonic movements

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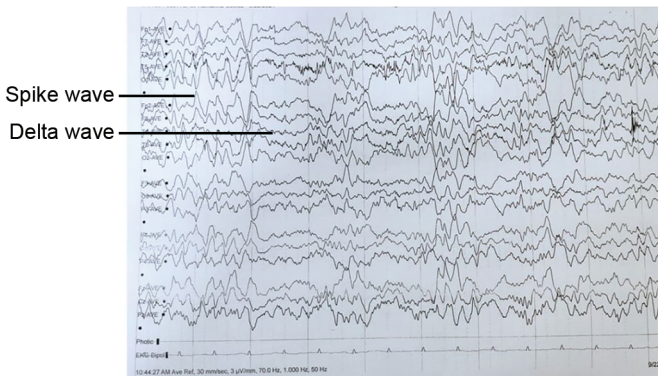

Fig. 1: Periodic, generalized spike wave discharges with background slow activity in delta range

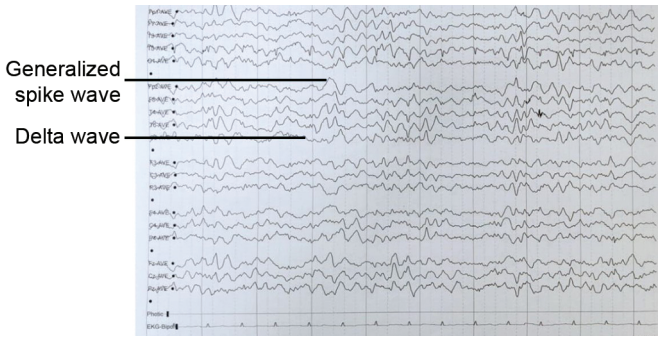

Fig. 2: Intermittent rhythmic delta activity and generalized spike wave discharges

may be subtle and often suppressed by sedatives or paralytic agents used commonly in patients in the intensive care units.

Myoclonus is classified according to the presentation as follows:[5]

- Cortical: appears spontaneously, or with movement, shock-like movement, highly sensitive to stimulus, irregular in rhythm.
- Subcortical: appears spontaneously, periodic, less shock-like movement, can be generalized and segmental, can be sensitive to stimulus.
- Spinal: occurs spontaneously and involves muscles innervated by spinal segments.
- Peripheral: rare, caused by lesions of peripheral nerves/plexus.

The underlying pathology in cortical myoclonus may involve dysfunction of the inhibitory interneurons of the motor cortex, which are responsible for maintaining balanced cortical activity.[6] This leads to reduced short-interval intracortical inhibition of the motor cortex (M1). This reduction has been attributed to the dysfunction of GABAergic system within the motor cortex.

An EEG is useful in localizing the source of the myoclonus. In cortical myoclonus, multifocal or generalized spike-and-wave or poly-spike-wave discharges are seen. In noncortical myoclonus, jerk-linked discharges are absent. The patient in this case report fitted the criteria of cortical myoclonus based on clinical presentation and EEG findings.

Myoclonus is a frequent complication of encephalopathy due to various conditions including hypoxia, infection, drug toxicity, metabolic disorders, and neurodegenerative disorders,[7] as enumerated in Table 1.

Treatment in isolated focal myoclonus should focus on treating the underlying cause. The myoclonus itself may require to be suppressed if it is sustained for long periods of time or if it causes instability or ventilator dyssynchrony; for this sodium valproate and levetiracetam are the drugs of choice. The generalized myoclonus seen in hypoxic-ischemic encephalopathy is associated with poor neurological outcome.

Table 1: Causes of myoclonus in critically ill patients

- Intoxication/drug related encephalopathy
  - Anesthetic agents—etomidate, propofol
  - Opioids—fentanyl, morphine
  - Antidepressants—tricyclic antidepressants, lithium
  - Heavy metals poisoning, cocaine, alcohol withdrawal, etc.
- Vascular—hypoxic encephalopathy
  - Myoclonus with severe hypoxic-ischemic encephalopathy
  - Lance–Adams syndrome
- Metabolic encephalopathies
  - Uremic, hepatic encephalopathy, rarely hyper and hypoglycemic encephalopathy
- Encephalitides
  - Viral, EBV, West Nile virus, autoimmune, paraneoplastic
- Traumatic brain injury
- Posterior reversible encephalopathy syndrome
- Traumatic spinal cord injury, spinal cord tumors
- Progressive encephalomyelitis with rigidity and myoclonus
- Peripheral nerve lesions

Conclusion

Recognizing myoclonus in critically ill patient and determining the neuroanatomical origin and treatment of the underlying cause plays a key role in the management.

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Video

Myoclonus video is present in the website of jacutecare.com

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