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Severe oromandibular dystonia in a child following Japanese encephalitis treated with botulinum toxin

Abstract

Japanese encephalitis is an important cause of encephalitis in Southeast Asia. Survivors may suffer from various movement disorders leading to disability, presumed to be due to involvement of basal ganglia and thalamus. Oromandibular dystonia is a rare complication of Japanese encephalitis and treatment is unsatisfactory in severe cases. We report a child with JE who developed markedly severe oromandibular dystonia in subacute phase of illness. His Magnetic Resonance Imaging revealed involvement of basal ganglia and thalami. Oral antidystonic medications were used without much avail. In view of severe disabling oromandibular dystonia he was treated with botulinum toxin without adverse effects and had improved quality of life. Botulinum toxin may be considered as a therapeutic option in severe oromandibular dystonia following Japanese encephalitis.

Key words: Basal ganglia, Botulinum toxin, Dystonia, Encephalitis, Thalamus,

Introduction

Japanese encephalitis (JE) is caused by RNA (ribonucleic acid) virus, a mosquito born zoonotic disease which belongs to the *Flaviviridae* family. The survivors suffer from variable degree of neurologic deficits and diverse array of movement disorders in acute or subacute phase.[1] Treatment of oromandibular dystonia (OMD) following JE has been reported, with variable degree of success with use of multiple drugs in different combinations.[2] We report a child with severe OMD following JE, characterized by mouth opening and tongue protrusion that was unresponsive to oral medications but showed good response to botulinum toxin (BTx). The BTx can be thus worth considering in severe OMD secondary to JE.

Case report

A 11-year-old boy was admitted with high grade fever with alteration in sensorium for 15 days. In second week

of his illness he developed involuntary, persistent mouth opening and protrusion of tongue which led to impaired swallowing, drooling of saliva and inability to speak (Figure 1).

To overcome this, attendants would keep the food in mouth and push it inside while pressing the tongue to assist in swallowing. He also had dystonic posturing of left upper and lower limbs. Neurological examination revealed Glasgow Coma Scale (GCS) score of 12 (E4 M6 V2). His speech was incomprehensible and he needed assistance in walking. Pupils were normal in size and reaction. Kayser–Fleischer ring was negative on slit lamp examination. Motor examination revealed asymmetric (left > right) rigidity of all four limbs with intermittent dystonic posturing of left hand and feet. Severity of OMD was grade 4 (Range 0-4). Deep tendon reflexes were normal and plantar responses were flexor. HIs routine blood investigations were within normal limits. Serum ceruloplasmin level was normal. Cerebrospinal fluid (CSF) testing revealed 5 lymphocytes / mm^3, protein of 34 mg/dL and sugar of 80 mg /dL. CSF immunoglobulin (Ig M) was positive for JE by immunoglobulin M capture enzyme linked immunosorbent assay (MAC ELISA). CSF was negative for Epstein Barr virus, herpes simplex, varicella zoster virus serology, tuberculosis and cryptococcal infection. His serum ELISA for human immunodeficiency virus, hepatitis B and C were negative. Magnetic Resonance Imaging (MRI) of the brain was performed which revealed hyperintensities in bilateral thalami, caudate, globus pallidus, right substantia nigra, parietal lobe on T2 weighted image and fluid attenuation inversion recovery (FLAIR) sequence (Figure 2).

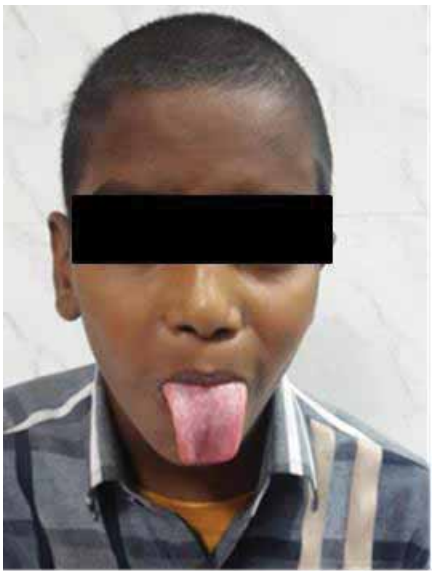

Figure 1: A boy with mouth open and tongue protrusion dystonia.

He was treated with sodium valproate, tetrabenazine, trihexyphenidyl, clonazepam without much improvement. After two weeks of trial with oral medication, he was treated with injection of 40 units of BTx in bilateral genioglossus and lateral pterygoid muscle (10 units each). There was significant reduction in the oromandibular dystonia from grade 4 to 2 at three months follow up, with improvement in swallowing. Hyperintensities in the thalami had also regressed in size on repeat brain MRI at three months follow up.

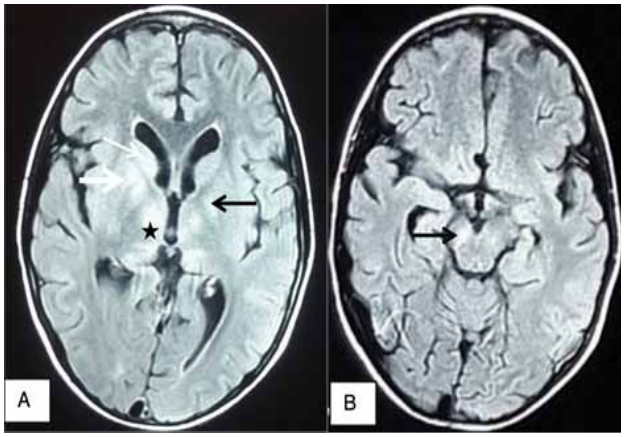

Figure 2: A. FLAIR image MRI brain (axial view) of patient showing hyperintensities in bilateral caudate (thin arrow), putamen (thick arrow), globus pallidus (black arrow) and thalami (asterisks) B. Hyperintensity in right substantia nigra (arrow).

Discussion

OMD is a focal dystonia whereby repetitive or sustained spasms of the masticatory, facial, or lingual muscles result in involuntary, and possibly painful jaw opening, closing, deflecting, retruding, or a combination of the above.[3] Various hereditary, neurodegenerative, metabolic-toxic, pharmacologic, demyelinating, neoplastic and infectious (e.g. encephalitides) etiologies are implicated.[4] In a study of 165 patients from neurocritical care setting, 33 had new onset cervical dystonia (CD) or OMD. In 21 of these 33 patients, identified etiologies included the following: intracerebral haemorrhage in 13, ischemic stroke in 6, status epilepticus in 1, and unclear etiology in 1. Spontaneous recovery was noted in 7 patients. Four patients with OMD and 7 patients with CD received BTx therapy. Two of the 4 patients with OMD (50%) and 4 of the 7 patients with CD (57%) treated with BTx improved.[5] In a study of 17 patients (JE in 14, nonspecific encephalitis in 3) with OMD, various oral medications were used in different combinations. The OMD regressed completely in 6 patients, improved by 1 grade in 2 patients, and remained unchanged in 7.[2] Our patient had markedly severe OMD

that was not responding to two weeks trial with oral medication, hence BTx was administered without any adverse effects and with significant reduction in the OMD and improved swallowing at three months follow-up.

The inhibitory basal ganglia output has been considered to be dysfunctional in patients with dystonia. However, in a recent review it has been proposed that dystonia is attributable, not to dysfunction of single brain region but rather to a motor network dysfunction.[6]

In our patient, there was clear involvement of both thalami and basal ganglia during the encephalitic process, and this may explain the manifestation of OMD. The involvement of above structures in various combinations has also been reported in JE.[2] In a study of 148 patients with JE, the dystonia was more common in children (43.18%) than adults (18.2%). The presence of thalamic lesion was significantly associated with development of dystonia.[7] Treatment of dystonia is symptomatic and BTx therapy is generally recommended for focal dystonia.[3] In our review of the literature, we were unable to find any report on the use of BTx in OMD following JE. Although the dystonia may improve with time in some cases, the use of BTx in the early phase may help in early symptomatic relief. In our patient's case, the use of BTx may have helped in providing early symptomatic relief, but may not be solely responsible for recovery.

Conclusion

Patients with JE may have severe OMD. BTx can be used as a symptomatic therapy with improved quality of life and without significant adverse effects. While treatment of the neurological complications of JE such as OMD can be actively pursued, from a broader perspective, prevention of JE is the more ideal approach to the management. In JE, avoidance of mosquito bite and immunization against JE remain an effective mode of prevention.

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