Case Report

Acute bulbar palsy plus syndrome: A rare variant of Guillain–Barre syndrome

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ABSTRACT

Guillain–Barre syndrome (GBS) is the most common cause of acute flaccid paralysis worldwide both in adult and pediatric population. Although flaccid paralysis is the hallmark of this disease, there are some rare variants which may be easily missed unless suspected. Here, we present a very rare variant of GBS - acute bulbar palsy plus syndrome in a pediatric patient. A 13-year-old female child presented with right-sided lower motor neuron type of facial palsy and palsy of bilateral glossopharyngeal and vagus nerve of 2 weeks duration. On detailed neurological examination, motor and sensory system were normal, but the deep tendon reflexes were absent universally. Nerve conduction study showed demyelinating motor neuropathy. Based on typical clinical course and electrophysiological studies, the diagnosis was made. To the best of our knowledge, this is the first pediatric case of unilateral facial palsy with bulbar involvement without any motor abnormality.

Key words: Acute bulbar palsy plus syndrome, Bulbar palsy, Guillain-Barre Syndrome, nerve conduction study

Introduction

Guillain-Barre syndrome (GBS) is the commonest cause of acute flaccid paralysis in children. but the rarer forms of this disease can be easily missed if not evaluated properly as many of them lack any motor anomaly.

Case Report

A 13-year-old female child presented to our outpatient department with complaints of facial deviation, dysphagia, especially for liquid, nasal regurgitation of food, nasal intonation of voice for the last 14 days. Symptoms were acute in onset, progressed initially for 3–4 days, followed by a static course. She did not have weakness of any limb. She had upper respiratory tract infection 3 weeks back that improved without any treatment. There was no history of any injection, insect bite, animal bite, head injury, ear discharge, and loose motions. Vaccination status of the child was unknown. Mother was not sure of oral polio vaccine or other vaccination. Her previous medical history was not significant.

On the day of first contact, the child was fully conscious, alert, and responding. Physical examination showed facial deviation to the left, drooling of saliva, and nasal intonation. Pulse rate was 70/min, regular rhythm, good volume; blood pressure was 100/60 mmHg in sitting decubitus. On examination, right-sided lower motor neuron type of facial nerve palsy was noted along with 9th, 10th cranial nerve (CN) palsy. Rest all CNs were normal. Limb muscles power were normal, of grade 5/5, but deep tendon reflexes were universally absent and plantar responses were downgoing bilaterally. Sensory
system examination was within normal limits. Throughout the course of disease, sensory and cerebellar systems were normal and there was no sign of meningeal irritation. There was no bladder and bowel incontinence. Examination of other systems also did not show any abnormality. Fundus examination was normal. Nerve conduction study was done which showed reduced amplitude and decreased conduction velocity of right median motor nerve. Right peroneal motor nerve also showed reduced amplitude. Study of sensory nerves was normal. Rest other tested nerves were normal. Routine blood investigations including complete hemogram were within normal limits. Workup for poliovirus was negative. Daily physiotherapy was started under supervision. As symptoms were not progressive, attendants were explained about danger signs and she was followed up twice weekly, every time with detailed neurological examination. Disease did not progress further and slow improvement was noticed at first in facial nerve within next 4 weeks. Serological tests for antibodies were not done due to financial constraints. On the basis of typical clinical course and electrophysiological study, the case was diagnosed as acute bulbar palsy plus syndrome – a localized variety of Guillain–Barre syndrome (GBS).

Discussion

GBS is an immune-mediated postinfectious peripheral neuropathy which is typically characterized by acute onset of symmetrical ascending limb weakness and generalized areflexia. It can be triggered by infectious, inflammatory, or systemic diseases. There are few rare regional variants of GBS such as polycranial neuritis and acute bulbar palsy plus syndrome where patients present with pharyngeal and palatal weakness with or without other CN involvement in the absence of any limb weakness or ataxia as in our case. Although CN palsies are common in GBS, multiple cranial neuropathies as a variant of GBS are very rare and account for only 5% of patients[1] and even rarer in pediatric population. Case with pure bulbar palsy with unilateral facial palsy in GBS in pediatric population has not been reported earlier as per our literature search. Because of rarity of features and most often GBS is not considered in differential as it does not progress to limb weakness and lack of supportive evidence, there are only very few reports of pure cranial neuropathy with areflexia and normal motor and sensory functions.[2,3] A similar case was reported in an adult female of 19 years from Malaysia who had spontaneous improvement without any intervention.[4] Although bilateral facial nerve palsy is known in GBS, unilateral palsy is extremely rare and only single case report of an adult male we came across in literature search.[5] A recent study analyzed 11 adult cases of acute bulbar palsy plus syndrome and concluded it a variant of GBS.[6] Nerve conduction study provides strong supportive evidence in the diagnosis of GBS, especially the atypical one. Delayed distal latency, slow nerve conduction velocity, temporal dispersion of waveforms, conduction block, prolonged or absent F waves, and prolonged or absent H-reflexes are all findings that support demyelination of peripheral nerves in GBS. Abnormality in the conduction studies in the form of demyelinating neuropathy or axonal variety is almost universally detected in cases of GBS[7] mostly after a minimum of 5 days from the onset of disease.[8]

Conclusion

This case represents a rare form of GBS. This also highlights the fact that GBS should be considered in cases of isolated multiple cranial palsies for early intervention and close monitoring of any progression or complication.

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Conflicts of interest

There are no conflicts of interest.

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