CASE REPORT

Botulism in Children: A Diagnostic Dilemma in Developing Countries

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Abstract

Botulism is a well-known disease of the neuromuscular junction. It is a rare but curable cause of paralysis in paediatric population. In addition to classical clinical signs and symptoms, the diagnosis of botulism requires laboratory confirmation of intoxication by various biological tests. These include demonstration of botulinum toxin in serum or isolation of the *Clostridium botulinum* from stool/gastric aspirates. However, it is not always possible to confirm intoxication due to unavailability of technical facilities, especially in resource limited countries like Pakistan. Under these circumstances, electrophysiological studies serve as an excellent diagnostic tool. These studies can provide quick diagnosis of botulism so that early administration of botulism immunoglobulin, if available, can reduce morbidity, mortality and length of stay in hospital. We report a case of botulism from Pakistan diagnosed on the basis of electrophysiological studies.

Key words: *Botulism. Children. Electrophysiology. Repetitive nerve stimulation.*

INTRODUCTION

Botulism is a rare systemic paralytic disorder, especially beyond the infantile age group. It is caused by Clostridium *botulinum*, which produces a neurotoxin responsible for the clinical manifestations.[1] In the United States; about 110 cases of botulism are reported each year. Most of these cases are seen in infants. In children, it is rarely reported suggesting that it is an uncommon condition beyond infantile age group.[2] In England and Wales, only 6 cases of infant botulism were reported by health protection agency between 1975 and 2006.[3] However, some recent studies have shown a rising trend in the incidence of infant botulism in UK.[4] There is very limited reported data about botulism in developing countries.

Establishing the diagnosis of botulism can prove to be very challenging. Sometimes, even classical cases may be misdiagnosed as neuromuscular disorders while on the other hand, some milder cases may go undiagnosed, if not suspected and worked up actively.[5] Several clinical mimics also need to be differentiated to rule out other etiologies as this can lead to false diagnosis and treatment.[6] Botulinum immunoglobulin (BIG-IV) is the treatment of choice and can significantly reduce the morbidity, hospital stay and expenses of the treatment, especially if given early in the course of disease.[7]

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CASE REPORT

An 8 years old child was admitted with acute history of dysphagia, ptosis of left eye and generalized weakness. No fever, seizures or altered mental status were reported. On physical examination, the child was afebrile, with normal vital signs. Neurological examination revealed decreased pupillary response to light; third and seventh cranial nerves involvement, weak gag reflex and absent deep tendon reflexes. An initial differential diagnosis of Guillain-Barre syndrome, brainstem encephalitis, poliomyelitis and botulism was made. Lumbar puncture showed a normal CSF; all other laboratory findings were normal as were the CT scanning and MRI findings of the brain. The child developed respiratory arrest on second day of admission hence, intubated and shifted to paediatric intensive care unit. Stool cultures were sent for polio while no biologic tests were available for botulism. Initial electrophysiological studies were normal so treated on the lines of Guillain-Barré syndrome on empiric basis. As the child did not respond to standard treatment and there was high clinical suspicion of botulism, electrophysiological studies were repeated. This time, the studies were highly consistent with botulism showing incremental response on repetitive nerve stimulation (Figures 1 and 2).

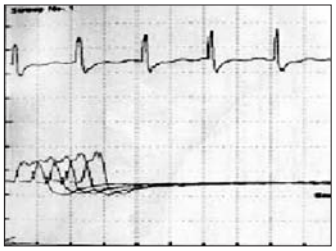

Figure 1: Repetitive nerve stimulation of ulnar nerve demonstrating 93% increment.

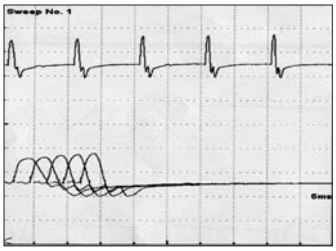

Figure 2: Repetitive nerve stimulation of median nerve demonstrating 53% increment.

No antitoxin was administered due to non-availability in Pakistan. Mechanical ventilation was continued. Later on, tracheostomy was done as the child needed prolonged respiratory support. The child remained on ventilator for about 6 weeks. Nasogastric feeding started along with other supportive measures. Child showed gradual recovery after 4 – 5 weeks, starting with improvement in ptosis. Full recovery of cranial nerves was achieved in 8 weeks. At discharge, the child was able to walk with support. On follow-up, complete clinical recovery was noted and tracheostomy was decanulated.

DISCUSSION

Botulinum toxins block the release of acetylcholine from presynaptic cholinergic nerve terminals at neuro-muscular junctions, thus leading to all its clinical manifestation. The signs and symptoms closely resemble to its differential diagnosis like Guillain-Barré syndrome, especially its Miller-Fisher variant, myasthenia gravis and other neurological diseases. For accurate diagnosis of botulism, demonstration of its toxin, either in stool or blood, is considered to be gold standard but these biological tests are not readily available in most developing countries including Pakistan. However, electrophysiological studies of botulism demonstrate characteristic defects of neuro-muscular transmission,[8,9] so that a suggestive clinical picture combined with these electrophysiological findings can be considered as the best method for the diagnosis of botulism in resource limited countries. In particular, electrophysiological testing is of great advantage for the early diagnosis of suspected cases even before the results of bacteriological tests, if available, so that antitoxin can be given earlier to reduce the morbidity and length of stay in the hospital. In this patient, blood and cerebrospinal fluid test results were normal along with CT scan and MRI of brain. Even initial EMG/nerve conduction velocity studies were normal but the patient demonstrated the classic features of botulism: progressive muscle weakness, cranial nerve palsies followed by sudden respiratory arrest. Since such a sequence of events may lead to serious consequences, early diagnosis and intervention is necessary.

Repetitive nerve stimulation (RNS) enables us to assess the status of neuromuscular junctions directly. Although increments in response to repetitive nerve stimulation are characteristic of both lambert-eaton myasthenic syndrome (LEMS) and botulism but in case of LEMS marked incremental response (> 200%) is seen while in botulism, only modest (50 – 100%) incremental response is seen.[10] In case of myasthenia gravis, a normal or decremental response is seen in response to repetitive nerve stimulation.[10] In this patient, repetitive nerve stimulation demonstrated 93% increment in ulnar nerve and 53% increment in median nerve (Figures 1 and 2) which are characteristic and diagnostic of botulism. Thus, an early and accurate diagnosis of botulism can be made with the help of electrophysiological studies.

There is limited data available about the cases of childhood botulism suggesting that it may be under-recognized, underreported or both. When bulbar palsies, hypotonia and weakness are present, clinicians should have a high index of suspicion for botulism. Electrophysiological studies are important diagnostic tool especially when stool culture and other biologic laboratory tests facilities are not available.

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