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A case of descending paralysis consistent with food borne botulism

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Introduction

Botulism was first reported in the 1820s in a Southern German town as a case series involving hundreds of patients with sausage poisoning. *Clostridium botulinum* is a gram-positive, rod-shaped, spore-forming, obligate anaerobic bacteria easily found on the surfaces of vegetables, fruits, and seafood, and exist in soil and marine sediment (1). There are seven serotypes (A to G) based on the antigenicity of toxin production. Currently there are eight distinct *C.botulinum* toxin types (A to H), in which A, B, E, and rarely F, G, and H, cause human disease (1, 2). Botulinum toxin is the most potent bacterial toxin and the most potent known poison. The minimum lethal dose (MLD) of Botulinum toxin in experimental mice is 0.0003 μg/kg (1). Once ingested, the toxin is primarily absorbed by the stomach and small intestine, as well as from the large intestine. This preformed neurotoxin by *Clostridium botulinum* targets multiple tissues including motor and sensory neurons and block the cholinergic innervation of neuromuscular junction in striated and smooth muscles as well as cholinergic innervation of the tear, salivary, and sweat glands (1).Thereby it inhibits the release of vesicles containing acetylcholine in the synaptic cleft as well as dopamine, serotonin, somatostatin, noradrenaline, and gamma-aminobutyric acid causing paralysis and autonomic disturbances (3).

Several forms of botulism are identified as syndromes by the mode of acquiring the Infection. They are infant botulism, foodborne botulism, wound botulism, iatrogenic botulism, adult intestinal colonization and bioterrorism-associated botulism. Nearly 70-75 % of cases account for infant botulism, 20-25% are foodborne, and 5-10% are wound-related (3). Botulism due to intestinal colonization is extremely rare (1).

Botulism appears after an incubation period of several hours to eight days (4). Classical presentation of the botulism syndrome is with absence of fever, symmetric neurologic deficits, normal sensorium and mental status, normal or slow heart rate and normal blood pressure and absence of sensory deficits with the exception of blurred vision (5).

Definitive diagnosis of botulism is confirmed by identification of toxin in serum, stool, vomitus, food sources or by isolation of *C. botulinum* from stool, wound specimens, or food sources (6). However, detection of toxins takes one to four days and anaerobic cultures of *C. botulinum* often take up to six days (1). This renders the importance of clinical diagnosis rather than waiting for laboratory evidence in management decisions of administering botulinum antitoxin which should be done as soon as possible, preferably within the first 24 to 72 hours of presentation (7, 8).

Case presentation

A 49-year-old previously healthy patient presented to a local hospital with vomiting and abdominal bloating for 2 days. He complained of abdominal

bloating around midnight on the first day and had 6 - 7 episodes of vomiting the next day morning, for which he had been treated by a general practitioner. He denied associated fever, headache or history of self-ingestion of toxic substances. But he could recall the fact that he had consumed canned fish for breakfast and lunch on the previous day of symptoms onset. Vomiting was settled with anti-emetic but continued to have abdominal discomfort throughout the day. Next day he was awakened with blurred vision, and noticed nasal type of speech with difficulty in swallowing. Furthermore, he felt upper limb weakness and difficulty in walking as well. Then he was admitted to a local hospital and transferred to a tertiary care center for further evaluation.

On examination, he was an averagely built person with BMI of 24 kg/m^2 and no altered sensorium. Glasgow coma scale was 15 out of 15. He was afebrile and neurological examination revealed bilateral mydriasis (Figure 1) with absent direct and consensual light reflexes and accommodation reflex as well. Further, he had bilateral complete ophthalmoplegia, bilateral partial ptosis and lower motor neuron type bilateral multiple cranial nerve neuropathy including facial, glossopharyngeal and vagus nerves. Motor and sensory branches of trigeminal, vestibulocochlear, accessory and hypoglossal nerves were intact. Neck muscle power was 4 out of 5 on the Medical Research Council (MRC) scale. There was a flaccid weakness in upper limbs with motor power 3+ out of 5 on MRC scale without sensory impairment. Lower limb also had a lesser degree of flaccid weakness with motor power 4+ out of 5 on MRC without sensory impairment. Bilateral plantar responses were downwards. Reflexes in upper and lower limbs were intact. Cardiovascular examination showed pulse rate of 60 bpm with blood pressure of 140/80 mmHg. No cardiac murmurs were audible. Respiratory system examination revealed a respiratory rate of 16/min, normal chest expansion and bilateral equal vesicular breathing were there with no added sounds. His forced vital capacity and single breath count was measured at bedside and both were normal. His abdomen was soft and non-tender on superficial examination and no organomegaly was found.

His complete blood count showed WBC - 5,110 /L, Hb-15.4 g/dl, PLT - 330,000 /L. Blood picture did not revealed any abnormality. His renal and liver functions were biochemically within normal ranges (S, Creatinine - 64 mmol/L, BUN - 37 mg/dl, Na^+- 145mmol/L, K^+- 3.8 mmol/L, AST - 26 U/L, ALT - 27 U/L, ALP - 88 U/L, T.BIL - 16.5 mmol/L, D.BIL - 5.7 mmol/L, Albumin - 35 g/L, T. protein - 67 g/L). Inflammatory markers CRP was < 5 and ESR was -23 mm in 1st hour in normal range. Initially he underwent non contrast CT brain, later MRI brain and both were normal. Cerebrospinal fluid (CSF) study showed albuminocytological dissociation only [Color - clear, Appearance - colorless, Volume - 1.5 ml, Protein -153.7 mg/dl, Sugar - 69.4 mg/dl (RBS - 93), Polymorphs - nil, Lymphocytes - nil, Red cells - 4/mm^3, No molecular evidence of HSV1, HSV2, VZV, CSF culture- no growth]. Because of the clinical presentation and history of canned food ingestion botulism was high in the differential diagnoses and he underwent neurophysiological studies due to unavailability of botulinum neurotoxins (BoNTs) assay or anaerobic culture for definitive diagnosis. Repetitive nerve stimulation test for 20Hz showed incremental response which was supportive of botulism (Figure 2). The Electromyogram (EMG) and Nerve conduction study (NCS) were normal. Chest X-ray of this patient had no mediastinal widening suggestive of Thymoma. There was no fever throughout the hospital stay (Figure 3).

Botulism was the most probable clinical diagnosis in this case and it was decided to manage conservatively with supportive care in the absence of the definitive treatment, botulinum antitoxin. Frequent monitoring of blood pressure, pulse rate, saturation, single breath count at bedside and forced vital capacity done in order to identify the impending respiratory failure. Enteral feeding was done with the nasogastric tube and physiotherapy, swallowing exercises were continued. He had to undergo transient bilateral tarsorrhaphy in order to prevent damage due to bilateral facial nerve palsy. During hospital stay patients had constipation most probably due to botulism and managed with laxatives. Slow recovery was observed during one month of hospital stay and complete neurological recovery achieved within 3 months without respiratory failure needing intubation.

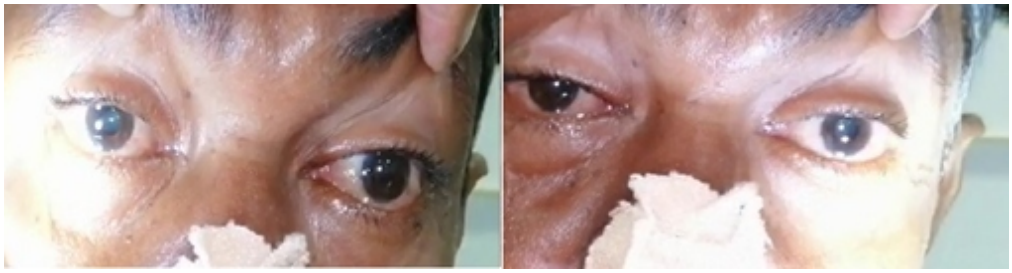

Figure 1: Bilateral mydriasis due to botulinum

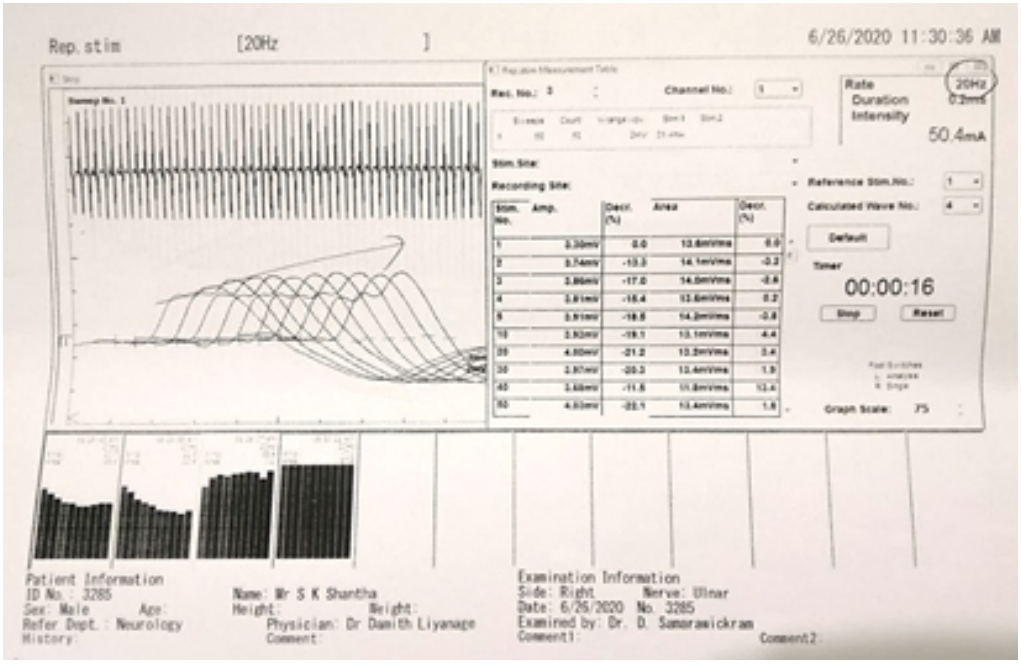

Figure 2: Incremental response in RNST at 20 Hz

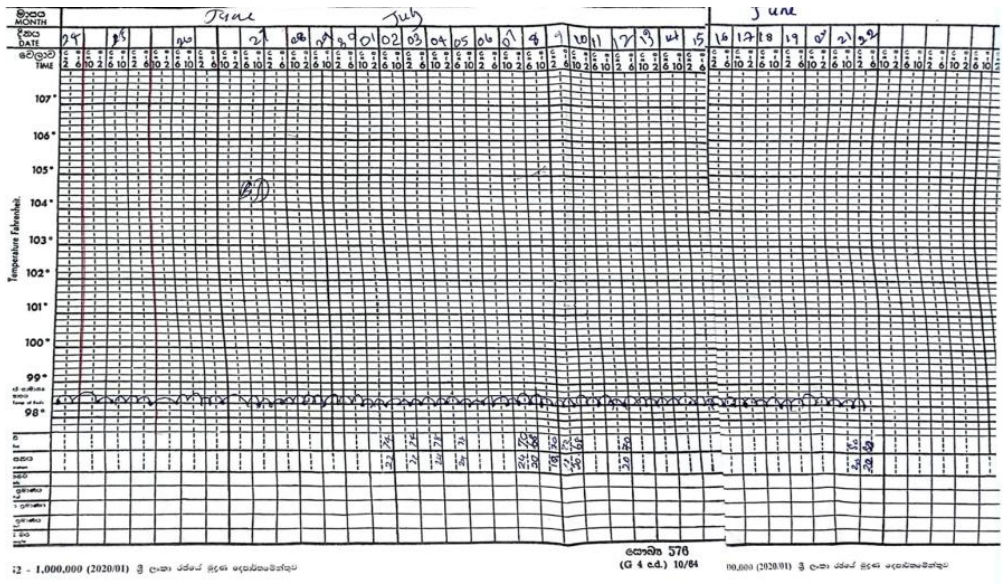

Figure 3: Temperature Chart showing apyrexia throughout the hospital stay

Discussion

Botulism is a rare disease and under-reported in Sri Lanka. Classical presentation of botulism is with symptoms of cranial neuropathy blurred vision, double vision, slurred speech, change in voice, dysphagia/ drooling of saliva and thick tongue sensation and at least one sign of neuropathy including ptosis, extraocular palsy, decreased tracking of objects, fatigability, facial paresis, loss of facial expression, fatigability while eating, fixed pupils, descending paralysis beginning with cranial nerves and absence of fever (1). In food borne botulism symptoms usually begin within 12 to 36 hours after ingestion of toxins and prodromal symptoms like nausea, vomiting, abdominal pain, diarrhea, and dry mouth with sore throat can be developed prior to the onset of neurological disability (8).

Because it is an acute flaccid paralysis, differential diagnoses that should be excluded are Guillain-Barré syndrome (GBS) [starting with sensory complaints, areflexia and ascending neuropathy], and its variants like Miller Fisher syndrome (MFS) [typical triad of symptoms - opthalmoplegia, ataxia, areflexia and can have extremity weakness and fixed dilated pupils, positive Anti GQ1b 85-90%], Bickerstaff's brainstem encephalitis (BBE) [typical triad of symptoms - opthalmoplegia, ataxia, decreased consciousness with hyperreflexia, positive Babiniski's sign, deep sensory impairment, anti GQ1b positive in 70%], Fisher-Bickerstaff's syndrome (overlap of clinical finding of MFS and BBE - opthalmoplegia, ataxia, clear consciousness, preserved tendon reflexes) (2, 3, 5-7). GBS and its variants can have elevated protein in CSF analysis and it may be rarely observed in botulism as in our case. On the other hand the CSF protein levels in the first 10 days of GBS are usually normal (8). Other differential diagnosis are myasthenia gravis (insidious onset, history of fatigability, preserved reflexes, lack autonomic symptoms, positive ice pack test at bedside and positive edrophonium test), Lambert Eaton myasthenia syndrome (insidious onset, cranial nerve sparing, respiratory failure rare, associated with cancer), stroke syndrome (structural changes on CT or MRI, and most of the time presenting with unilateral weakness), poliomyelitis (acute onset, asymmetrical weakness, fever, elevated CSF cells and it was not reported in Sri Lanka after 1993), tick paralysis (visible tick, outdoor activities, ascending weakness) (2, 3, 5-7).

Neurophysiological studies frequently present supportive evidence for diagnosis, but the normal findings do not exclude the clinical diagnosis as it can happen in the early phase of the illness (9). Repetitive nerve stimulation (RNS) at frequencies of 2 to 5 Hz will deplete the available stores of acetylcholine from the neuromuscular junction and decrease CMAP amplitudes resulting in decremental response. Frequencies of 20 to 50 Hz cause accumulation of calcium in the presynaptic terminal and cause increased release of acetylcholine resulting in an incremental response (5, 9). Our patient had an incremental response at 20 Hz of RNST and it was keeping with the clinical diagnosis of botulism (Figure 2). Nerve Conduction Study (NCS) will demonstrate normal conduction velocities with normal sensory nerve action potentials and decreased compound muscle action potential (CMAP) amplitudes if the presynaptic block is severe enough (9). In Electromyography (EMG) studies it is expected to see low amplitude short duration motor units (9). But in our scenario both NCS and EMG were normal.

Definitive diagnosis of botulism can be made by toxin identification in serum, stool, vomitus, or food sources or by isolation of *C. botulinum* from stool, wound specimens, or food sources (1, 3, 6, 9). But it takes one to four days to detect toxins and six days for growth and identification of the organism anaerobic culture media (1). In our case we could not perform those investigations due to unavailability of specific laboratory tests.

Treatment of botulism should address gastro-intestinal decontamination, the administration of the specific antidote and the support of respiratory function if necessary. Gastrointestinal decontamination in food borne botulism should only be considered in cases of ingestion of possible contaminated food is recent and if the constipation due to the anticholinergic effect as it may cause a permanence of the contaminated food in the gastrointestinal tract (9).

Antitoxin is the main therapeutic option for botulism and it binds to circulating neurotoxins and prevents their binding to the neuromuscular

junction (1, 8). If the clinical suspicion for botulism is high antitoxin should be administered as soon as possible even before the definitive test results are available (1, 7). In some cases of Botulism, patients may require ventilator support as it can cause respiratory failure with progressive descending paralysis specially in type A toxin which is the severe of all other types (3).

Conclusions

Botulism is always a clinical diagnosis and results of specific diagnosis tests should not delay the start of prompt management. Realizing its natural course, meticulous monitoring, supportive care and early identification of impending respiratory failure will reduce the mortality. Botulinum antitoxin should be given as soon as possible if it is available but it is not required in every case.

Informed consent has been obtained from the patient to publish this case report with photographs.

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