Rare presentation of botulism with generalized fasciculations

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Abstract
Botulism is a dreadful, life-threatening, neuroparalytic disease caused by gram positive bacteria *Clostridium botulinum*. Food borne botulism has been described following ingestion of preformed toxins in canned food or food products that have not been preserved properly. Botulinum toxin acts on neuromuscular junction and manifests as ophthalmoplegia, bulbar and limb weakness, and autonomic features along with respiratory compromise. The literature and case reports regarding neuroparalytic botulism in India are sparse. Generalized fasciculations have been rarely reported in literature as manifestation of food borne botulism. We present a 35-year-male presenting with usual features described, but with prominent generalized fasciculations which are rare, and which rapidly responded to treatment.

Key words: Botulism, food borne, fasciculations, neuromuscular, quadriparesis

Introduction
Botulism is a disease affecting neuromuscular junction, caused by potent neurotoxin of anaerobic bacteria *Clostridium botulinum*. The usual features are oculobulbar weakness, quadriparesis, and respiratory muscle paralysis.

Case Report
A 35-year-old male, presented with complaints of sudden onset pain abdomen of 8 hours duration following consumption of stale food (fried gram flour in buttermilk; *Dahi Vada* (Indian snack) kept inside a container since 4 days). He had one episode of vomiting; however there was no complaint of diarrhea. On 2nd day, he developed acute onset bilateral symmetrical drooping of eyelids with blurring of vision and difficulty in swallowing along with hoarseness of voice and dryness of mouth. He gradually developed progressive weakness of both upper limbs followed by lower limbs, with associated involuntary generalized twitching movements involving entire body. There were no complaints of seizures, altered sensorium, sensory symptoms, and bladder involvement. There was no history of medication intake, insect bite, or snake bite.

On admission, he was conscious and oriented. Vitals were stable with pulse rate of 68/min and blood pressure was 130/80 mmHg without postural fall. He was afebrile with mild tachypnea. Single breath count was 26. Higher mental functions were normal. Cranial nerve examination revealed bilateral dilated sluggishly reactive pupils, bilateral symmetrical ptosis with complete ophthalmoplegia along with decreased gag reflex. Motor system examination showed normal tone with bilateral symmetrical quadriparesis (muscle power, Medical Research Council (MRC) grade 4-/5). Deep tendon reflexes were normally elicitable with bilateral flexor plantar response. Patient also had generalized fasciculations involving anterior chest wall and bilateral upper and lower limbs [Video 1]. Sensory and cerebellar system examination was normal. There were no signs of meningeal irritation.

See the video: www.ijabmr.org
In view of history of symptom onset following stale food intake with presentation of dry mouth, bilateral ptosis with ophthalmoplegia, bulbar weakness, and lower motor neuron (LMN) quadriplegia, with normal deep tendon reflexes, possibility of neuromuscular junction affection; most likely food borne botulism was kept.

Hematological evaluation revealed hemoglobin (Hb)-14.8 gm%, total leucocyte count - 10,500/mm³, platelets-1.71 lakh/mm³, and erythrocyte sedimentation rate (ESR)-25 mm/l h. Biochemical parameters showed fasting blood sugar-74 mg/dl, urea-35 mg/dl, serum (S.) creatinine-0.9 mg/dl, S. sodium-136 meq/l, S. potassium-4.5 meq/l, S. calcium-9.2 mg/dl, S. magnesium-2.0 mg/dl, and serum creatine phosphokinase (CPK)-793 IU/l. Liver function tests were mildly deranged with serum glutamic oxaloacetic transaminase (SGOT)-95 IU and serum glutamic-pyruvic transaminase (SGPT)-66 IU. Thyroid profile was normal. Electrocardiogram, chest X-ray, and ultrasonography (USG) abdomen were normal. HIV ELISA was negative. Stool culture was sent.

Noncontrast computerized tomography of head with thin brainstem sections was normal. Motor and sensory nerve conduction study was suggestive of pure motor axonal affection of both upper and lower limbs with normal distal latency and conduction velocities. Three hertz repetitive nerve stimulation (RNS) test was negative for decremental response. High frequency (50 Hz) RNS also did not show any incremental response. Parenteral neostigmine test was negative.

Patient was managed conservatively keeping in view the possibility of foodborne botulism presenting as features of neuromuscular blockade. He was treated with cathersics, intravenous antibiotics, and short course of intravenous steroids (hydrocortisone). Respiratory and hemodynamic parameters were closely monitored. Patient showed gradual improvement of ptosis, ophthalmoplegia, limb weakness, and dysphagia. Respiratory effort improved and fasciculations subsided [Video 2]. Repeat biochemical and hematological parameters were normal. Stool culture was sterile. He was discharged after 2 weeks.

**DISCUSSION**

Botulism is a rare neuroparalytic disease caused by potent neurotoxin of anaerobic bacteria *Clostridium botulinum*. This is an anaerobic gram positive bacterium that forms subterminal spore. The bacterium proliferates under anaerobic condition, leading to germination of spores and produces a powerful toxin. Till date, eight different types of toxin (A, B, C1, C2, D, E, F, and G) have been described, which are zinc related endopeptidases that cleave polypeptides in cell membrane docking proteins: SNAP-25 and synaptobrevin, leading to blockade of presynaptic release of acetylcholine at neuromuscular junction and involvement of postganglionic parasympathetic nerve endings and peripheral ganglia.[1] Most human cases of food borne botulism are described with toxin A, B, and E. The botulinum toxin has been described as most potent neurotoxin and clinical manifestations depend on dose and toxin type. Home canned food, preserved food, alkaline food, and seafood are commonly implicated in food borne botulism.[2]

The incubation period of food borne botulism is 12-36 h after toxin ingestion. Clinical manifestations in initial phase are nausea, vomiting, and dryness of mouth. Eventually signs and symptoms of ocularbular muscle weakness including diplopia, ptosis, ophthalmoplegia, dysarthria, and dysphagia along with blurring of vision supervene.[3] Dilatation of pupils is found in less than 50% of patients.[5] These abnormalities of cranial musculature are followed by descending pattern of weakness involving upper limbs, lower limbs, and in severe cases respiratory muscle involvement leading to dyspnea and eventually respiratory failure. Sensory system and mentation are spared. Deep tendon reflexes are usually preserved. Autonomic symptoms and signs include dry mouth, constipation, pupillary abnormalities, and postural hypotension.

In the present case, patient presented with above mentioned features along with prominent limb and chest wall fasciculations. Very few cases of botulism with associated muscle fasciculations have been reported.[4,5] The cause of fasciculation can be postulated to be due to denervation changes or due to unstable neuromuscular junction.

The differential diagnosis of botulism includes: Myasthenia gravis, Lambert-Eaton myasthenic syndrome, tick paralysis, Miller Fisher variant of Guillain-Barré syndrome (GBS), diphtheritic neuropathy, brainstem stroke, and magnesium intoxication.[3] However in our patient, in view of abdominal symptoms, features of neuromuscular blockade, absence of history of insect bite, and normal brain imaging; possibility of food borne botulism was considered.

Electrophysiological studies can provide presumptive evidence of botulism. The most common electrophysiological abnormality being reduced compound muscle action potential (CMAP) amplitudes with normal motor and sensory nerve conduction. RNS may reveal an incremental response (facilitation) to repetitive stimulation at high frequencies, with a decremental response at lower frequencies.
Routine laboratory tests are not beneficial in confirming the clinical suspicion of botulism. Definitive laboratory diagnosis of botulism is established by detection of toxin in patient’s serum, stool, gastrointestinal contents, or isolation of *Clostridium botulinum* in stool. However, sensitivity of detection decreases rapidly with time. Chances of obtaining positive results are less than 13% in serum samples taken after 2 days and only 36% of stool samples are positive after 3 days. Hence, such clinical presentation with history of food borne illness should be strongly considered for possibility of botulism and needs to be treated urgently.

Treatment consists mainly of intensive medical and nursing care with careful attention to respiratory status. Botulinum antitoxin may be administered, however there is controversy due to lack of efficacy in many cases and risk of anaphylaxis. Steroids, plasmapheresis, and intravenous immunoglobulin have been described as adjunctive treatment.

Our patient was managed with intravenous steroids, antibiotics, supportive care, and regular respiratory status assessment, with gradual and complete recovery and disappearance of fasciculations.

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