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Acute onset of bulbar and ocular paralysis
An isolated case, with differential diagnosis

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The diagnosis of botulism should be considered in all patients with the acute onset of cholinergic dysfunction and muscle weakness. The pupils are usually dilated, paralysis of ocular and bulbar muscles invariably occurs along with weakness of limb and axial muscles. This diagnosis is supported by the presence of a neuromuscular block and confirmed by the detection of toxin. Although the clinical presentation in this patient supported this diagnosis and a neuromuscular defect was present, no botulinum toxin was found. Not until Clostridium botulinum, type B organisms were cultured was the diagnosis of "probable botulism" made in this patient. Myasthenia gravis, atypical Guillain-Barré syndrome and a brain stem vascular lesion were also considered in the differential diagnosis. The anoxic encephalopathy following the cardiorespiratory arrest was an additional complicating factor.

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THE sudden onset of difficulty in speech and swallowing together with ocular weakness represents a neurological emergency. Often, these patients are diagnostic problems. The purpose of this case report is to discuss the various diseases which were considered in the differential diagnosis of a patient with these complaints.

Case report

This 68-year-old male was admitted to the Henry Ford Hospital on March 3, 1976, with a 24-hour history of vomiting, difficulty in speaking and swallowing followed by unsteadiness of gait, double vision and drooping of both eyelids. The patient reported that he lived alone.

Six days prior to admission, the patient was evaluated by one of us for a tremor of the upper limbs. It had been present for several years and was essential in type. No other abnormal neurological findings were detected. Past history was otherwise unremarkable.

On admission to the hospital, the patient was restless and vomiting. Blood pressure was 140/90. Pulse was 80 and regular. Temperature was 39°C. General physical examination was within normal limits. On neurological examination, he was alert and well oriented. The pupils were 4 mm in diameter and reacted sluggishly to light. Ocular movements were paralyzed. There was a complete bilateral ptosis. No abnormalities were noted on ophthalmoscopic examination. Bilateral facial weakness was apparent. Sensation over the face was intact. He was unable to swallow and

speech was dysphonic. The soft palate and tongue were weak. Strength was intact in the extremities. Respirations were labored. An intention tremor was apparent in both hands. Sensation was intact to pin prick. Deep tendon reflexes were present and equal. The plantar responses were flexor.

Routine studies of blood and urine were normal. Chest x-ray showed an infiltrate in the left lower lobe with a pleural effusion. Skull and cervical spine films were normal. The bilateral ptosis and ocular paralysis persisted following the intravenous injection of 10 mg of edrophonium chloride (Tensilon). A lumbar puncture (LP) was attempted but was unsuccessful due to his agitation.

Four hours after admission he developed a cardiorespiratory arrest. Rescucitation was successful and tracheostomy was performed. The patient, however, remained comatose and required respiratory assistance. Generalized convulsions and myoclonus developed. Although the seizures were controlled with anticonvulsants, the electroencephalogram (EEG) revealed electrical status with moderate amplitude spike and brief slow wave discharges from both frontal areas. Motor and sensory systems were difficult to assess. The deep tendon reflexes and plantar responses were absent.

One month later, along with improvement of consciousness, the patient was noted to be generally weak. The limbs were paralyzed except for a flicker of movement in fingers and toes. Ptosis was less apparent and the pupils, although dilated, reacted more briskly to light. The ocular movements were restricted in all gazes. Facial diplegia persisted. The deep tendon reflexes were absent.

Spontaneous respirations gradually improved and two months after cardiorespiratory arrest, assistance in breathing was no longer necessary. He continued to regain strength. The patient became able to sit with support to head and trunk. Strength in arms and legs improved. Generalized myoclonic jerks persisted. Although able to understand the spoken voice, his speech remained indistinct and he was unable to swallow. CT scan (computed axial tomography of the brain) revealed a few lucent areas in the right temporal lobe. Two weeks later, the CT scan was normal.

Improvement continued and three months after the onset of the illness he was discharged to a nursing home. When he returned to the clinic one month later for evaluation, the ocular movements were normal and the ptosis had disappeared. The pupils were of normal size and reacted briskly to light. Difficulties in speech and swallowing persisted, so tube feedings were still necessary. Movements of the soft palate and tongue were adequate, however, and the gag reflex was active. Weakness and wasting of muscles in the extremities were present. The generalized myoclonus persisted. The deep tendon reflexes were generally sluggish and plantar responses flexor.

Nerve conduction studies were performed five days after admission. Left median nerve latency from wrist to thenar eminence was 7.7 meters/second and a conduction velocity of 58 m/sec was obtained in the forearm. The left median nerve at the wrist was also stimulated supramaximally with frequencies ranging from 2 pulses/sec to 50 pulses/sec. The initial stimulus evoked a 2 millivolt action potential from the thenar eminence. With the next few stimuli, the amplitude progressively declined and this lower amplitude was maintained with successive shocks. This decrement was 1 millivolt with the slower rates of stimulation. Following cessation of the stimuli for a period of five seconds or more, the evoked action potential returned to the initial amplitude of two millivolts.

Four months later, these studies were repeated. Normal latencies and velocities were obtained for both peroneal, left ulnar and left median nerves. Supramaximal stimulation of the left median nerve at the wrist evoked a 10 millivolt action potential from the thenar eminence. This amplitude was maintained with a stimulus frequency of 5 pulses/sec for a period of one minute. Electromyogram (EMG) recordings with a monopolar needle electrode were also performed and several muscles in the left upper and lower limbs were studied. At rest, these muscles were electrically silent. During voluntary contraction, the motor units were normal in amplitude and duration, but a full interference pattern was not obtained.

Five days after admission, serum and feces from the patient as well as specimens of food from his home were obtained. Cultures of uncooked sausage were reported as positive for Clostridium botulinum, type B, but no toxin was detected. Serum, sent to the Center for Disease Control, Atlanta, Georgia, for a mouse-toxin neutralization test, was reported as negative for botulinum toxin. A stool specimen was inadequate for culture since it contained enema material. Six weeks after admission, another stool specimen was submitted to the Center for Disease Control. These cultures were reported five weeks later as positive for Clostridium botulinum, type B, but no toxin was detected.

Discussion

The symptoms of botulism usually develop from 8 to 36 hours after the ingestion

of contaminated food and have been documented by several investigators.[1-4] Blurred and double vision with drooping of eyelids are more common than difficulty in speech or swallowing. Generalized weakness with dryness of eyes, nose and throat are often conspicuous. These patients are alert and paresthesias are absent. Nausea, vomiting, abdominal pain and diarrhea may occur and are attributed to other bacterial contaminants. The classical complaint of constipation is uncommon.

In our patient, the clinical presentation of ocular and bulbar paralysis with dilated pupils was suggestive of botulism and sufficient to examine serum and feces, as well as food from his home, for culture and toxin identification. Although botulinum toxin was not detected in these specimens, Clostridium botulinum, type B organisms were cultured from feces and food specimens. These laboratory findings were discussed with Eugene Gangarosa, M.D., from the Center for Disease Control. Since botulinum toxin was not identified, he felt this to be a case of "probable botulism, type B."

Other cases of botulism have been reported in which toxin has not been detected in serum.[5-10] Dolman[6] has mentioned that "absence of detectable circulating toxin does not exclude botulism". In some of these patients, however, toxin was found in feces[7,10] while in others no toxin was demonstrated. In view of these findings, it has been emphasized that the administration of antitoxin should not be withheld or discontinued on the basis of negative laboratory findings, especially if the clinical and epidemiological evidence strongly suggests botulism.[6] Treatment with antitoxin was not given to our patient since clinical improvement had occurred prior to the detection of Clostridium botulinum, type B in the feces.

The electrophysiological findings in botulism are due to a defect in the release of acetylcholine and resemble the Eaton-Lambert syndrome.[3,5,11,12] The amplitude of the muscle potential to a supramaximal stimulus is smaller than normal, it increases with repetitive supramaximal stimuli and post-tetanic facilitation occurs. In the Eaton-Lambert syndrome these neuromuscular abnormalities are found in all muscles while, in botulism, the defects may be confined to a limb or entirely absent, especially in mild cases with primarily autonomic involvement. In other patients with botulism, the neuromuscular block is sufficiently profound to prevent facilitation.

Although the low amplitude of the evoked potential in our patient was compatible with botulism, the further decrease in amplitude with successive stimuli was similar to the pattern obtained in myasthenia gravis. This is an unusual response in botulism but has been observed with slower rates of stimulation and in type B botulism with primarily autonomic involvement.[3,9] In keeping with other patients with botulism, nerve conduction was normal. Fibrillations have also been recorded in these patients and are attributed to the toxin-induced state of denervation.[13] They were not observed in our patient.

These electrophysiological investigations were repeated four months after the onset of his illness. Along with clinical recovery, the amplitude of the evoked muscle action potential returned to normal and no alteration was noted with repetitive stimuli. Nerve conduction remained normal.

Although the pupils are usually dilated and either fixed or react sluggishly to light in patients with botulism, normal pupils have been described in this disease.[3] Furthermore, fatigue has occurred in some mild cases of botulism; strength as well as ptosis have improved following the administration of edrophonium chloride.[3] In these cases of botulism, myasthenia gravis must be considered. Furthermore, in our patient, repetitive stimulation gave a myasthenic response, but the acute onset of paralysis with pupillary involvement are not encountered in this condition. Regardless, a test dose of edrophonium chloride was given with no improvement of strength.

Six days prior to the acute illness, the patient was evaluated for an essential tremor which had been present for several years. On this occasion, gait was not ataxic, ocular movements and pupils were normal and the deep tendon reflexes were present. The syndrome of ophthalmoplegia, ataxia and areflexia should be entertained in the differential diagnosis since this type of Guillain-Barré disease is frequently confused with botulism. It was first described by Collier[14] and subsequent reports of such clinical presentations have been made.[15,16] The pupils are usually spared in this form of polyneuritis, but dilated and fixed pupils have been noted as well as normal spinal fluid protein and normal nerve conduction.[17,18] These latter findings are more commonly encountered in botulism and conceivably the diagnosis may have been incorrect in some of these patients. This is especially likely to occur since isolated and mild forms of food-borne botulism have been reported.[4]

A brain stem vascular accident was also entertained in the differential diagnosis, in view of the sudden onset of ocular and bulbar paralysis. The patient was, however, alert and there were no long tract motor or sensory signs at the time of admission to hospital. Likewise, the seizures and myoclonus were not due to cerebrovascular disease but rather related to the cerebral anoxia which undoubtedly occurred during the period of cardiorespiratory arrest.

In view of the degenerative changes which occur at the neuromuscular junction in botulism, recovery of muscle strength is delayed for several months.[19] In our patient, however, four months after onset the pupils and ocular movements were normal, ptosis had disappeared yet profound bulbar and extremity weakness remained. This persistent weakness was probably cerebral in origin and related to the anoxic episode, rather than botulism.

The EEG changes, consisting of high voltage slow waves both generalized and focal in nature, have been described in three patients with botulism.[20] The EEG abnormalities in our patient along with CT scan changes were attributed to the cerebral anoxia.

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Erratum

The information at bottom of p. 3 of the Spring, 1977, Vol. 25, No. 1 issue is incorrect. Requests for reprints of the paper on *"The clinical variations of hereditary spastic paraplegia in four families"* should be addressed to Dr. Robert D. Teasdall at Henry Ford Hospital, and not Dr. Sidney Goldstein, as incorrectly given. Dr. Teasdall is head of the Division of Neurology. Dr. Goldstein is head of the Division of Cardiovascular Diseases.