Case Report

Electrophysiologic Findings in a Case of Severe Botulism

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Abstract
Botulism is a rare but severe and rapidly progressing neurological disease. Here, we describe a 44 year old woman developed nausea, vomiting, diarrhea and cranial nerve palsies after eating spaghetti with tomato sauce prepared at home. These were rapidly followed by descending limb muscle weakness and respiratory failure. Our patient’s clinical evaluation and the detailed electrophysiologic examination suggested the diagnosis of botulism. The patient was given only medical and supportive care and fully recovered approximately 2 months later.

Key words: Botulism, electrophysiologic evaluation

INTRODUCTION
Botulism is a severe neurological disease caused by the toxin of Clostridium botulinum (1,2,3). The toxin causes skeletal muscle paralysis by producing a presynaptic blockade to the release of acetylcholine at the neuromuscular junction (2,3). Also, it affects the parasympathetic and the sympathetic systems (4,5).

Up to date five clinical forms of botulism have been described: 1) Classic (food-borne), 2) Wound, 3) Infant, 4) Hidden (adult variant of infant form) and 5) Inadvertent botulism. Classic botulism is an intoxication caused by the ingestion of preformed toxin in food contaminated with toxin-producing bacteria. Infant, hidden and wound botulisms are infectious forms. Inadvertent botulism is the most recent form and occurs in patients who have been treated with botulinum injections (2,3).

The clinical presentation of classic botulism has a stereotypical pattern. After ingestion of contaminated food, within 2-36 hours, cranial nerve palsies develop and are followed by descending limb muscle weakness and, in severe cases, by respiratory failure (2,3). The sensory system and mentation are spared, although sensory abnormalities have been reported (6). Laboratory proof is the detection of toxin in the patients’ serum or stool. Electrophysiologic studies can be helpful in diagnosis when bioassay studies are negative or not done. The most consistent electrophysiologic abnormality is a small evoked muscle action potential in response to a single supramaximal stimulation. Posttetanic facilitation (PTF) can be found...
after rapid rates (50 Hz) or 10 s of exercise. The major treatment is medical and supportive care with attention to respiratory failure (2).

In this article, we describe a detailed electrophysiologic study of a severe case of classic botulism.

CASE PRESENTATION
A 44 year old woman was admitted to the hospital because of limb muscle weakness and respiratory failure. The day before, she ate spaghetti with tomato sauce prepared at home. Approximately 10 hours later nausea, vomiting and diarrhea started then she developed blurred vision, ptosis, diplopia, dysphagia and dysartria within 6-7 hours. These were rapidly followed by descending limb muscle weakness and respiratory failure. Neurologic examination showed bilateral ptosis and bilateral facial weakness. Her pupils were round, equal in size and both reactive to light. Palatal movements and gag reflex were lost. The muscle strength of upper and lower extremities were 3/5. Her deep tendon reflexes were normal. The sensory examinations were normal. Plantar responses were flexor. No pathologic reflex was found. Other symptoms were urinary incontinence and dry mouth. She needed intubation and mechanical ventilation.

She had no another known disease and no history of drug use. She did not have a family history of neuromuscular disorders. Laboratory examinations included blood count, erythrocyte sedimentation rate, renal and liver function tests, thyroid function tests and cerebrospinal fluid, which were all within normal limits.

Electrophysiologic studies
The electrophysiologic studies including nerve conduction studies (NCSs), quantitative electromyography (EMG) and repetitive nerve stimulation test were performed on an EMG equipment (Medtronic- Skovlunde-Denmark) on the second day of the patient’s illness. All recordings were performed in an air-conditioned room at a constant temperature of 25ºC. The patient’s skin temperature was kept between 31-32ºC.

a) Nerve conduction studies
Median, ulnar, peroneal and tibial motor NCSs including F-wave were performed bilaterally. Sensory NCSs included median, ulnar nerves bilaterally and sural nerve on the left side. All studies were performed using the standard techniques of supramaximal percutaneous stimulation with a constant current stimulator and surface electrode recording. F-wave latency is considered the shortest F-wave latency elicited by 20 consecutive stimuli minus the distal motor latency. Median and ulnar sensory nerve action potentials (SNAPs) were obtained orthodromically stimulating from digits I and III on the left side and from palm and digit I on the right side for median nerve, from digit V for ulnar nerve and recording at the wrist. Sural nerve SNAP was obtained antidromically. The results of NCSs were corrected for age and height according to reference values used in our laboratory (8,9).

On the left side, the amplitudes of median, ulnar and tibial compound muscle action potentials (CMAPs) were moderately reduced and the amplitude of peroneal CMAP was normal. The distal latencies and conduction velocities (CVs) of median, ulnar, tibial and peroneal CMAPs were normal. Peroneal F-wave was absent and the numbers of the median, ulnar and tibial F-waves were reduced. The sensory conduction studies of median, ulnar and sural nerves were normal on the left side. On the right side, distal latencies and CVs of the median and ulnar CMAPs were normal whereas the amplitudes were moderately reduced. The numbers of F-waves were reduced. The CMAPs of tibial and peroneal nerves could not be obtained. The sensory conduction studies of median, ulnar and sural nerves were normal on the left side. Table 1 shows the results of NCSs.

b) Quantitative EMG
The abductor digitii minimi (ADM), first dorsal interosseus (FDI), biceps brachii, deltoid, trapezius and tibialis anterior muscles were examined on the right side. The motor unit potentials (MUPs) were collected with concentric electrodes and MUP data analyzed with multi MUP analysis (7). Thirtty MUPs were collected after doublets were deleted and the remaining MUPs, a minimum of 20, were analyzed.
Table 1: The results of nerve conduction studies.

<table>
<thead>
<tr>
<th>Nerves</th>
<th>Motor</th>
<th>Sensory</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Median</td>
<td>Ulnar</td>
</tr>
<tr>
<td>Side</td>
<td>R</td>
<td>L</td>
</tr>
<tr>
<td>Latency (ms)</td>
<td>3.3</td>
<td>3.2</td>
</tr>
<tr>
<td>Amplitude (mV, µV)</td>
<td>1.6</td>
<td>0.6</td>
</tr>
<tr>
<td>CV (ms)</td>
<td>54.2</td>
<td>55.5</td>
</tr>
<tr>
<td>F-wave latency (ms)</td>
<td>20.9</td>
<td>20.9</td>
</tr>
<tr>
<td>F-wave frequency</td>
<td>7/20</td>
<td>8/20</td>
</tr>
</tbody>
</table>

NR: No response, D: Digit, P: Palm

Values for amplitude are expressed as mV for motor studies and µV for sensory studies

Bold values are abnormal (outside ± 2 SD of the reference material in the laboratory).

The ADM, biceps brachii, deltoid and trapezius muscles showed normal MUPs whereas the tibialis anterior and the FDI muscles revealed normal recruitment of polyphasic, low amplitude, and short duration MUPs. Also, there were a moderate number of fibrillation potentials and positive sharp waves on the tibialis anterior and the FDI muscles.

c) Repetitive nerve stimulation test

After establishing a supramaximal CMAP bilaterally with surface electrodes over trapezius muscle bilaterally and ADM muscle on the right side, repetitive nerve stimulation was performed. Ten single square wave pulses of 0.3 ms duration at 3 Hz at 15 to 25 mA were used for each stimulation run. After the first stimulation run, the patient was instructed to maintain 20 seconds of maximal voluntary muscular contraction and postexercise stimulation was done to obtain an incremental response. For each muscle the patient had at least 5 runs at 1, 2, 3, 4 and 5 minutes after exercise. Automated decrement calculations were recorded and PTF was measured after exercise.

After 20 seconds of exercise, an increment of % 62.5 in the amplitude of CMAP was observed over the trapezius muscle at the right side and an increment of % 61.5 over the trapezius muscle on the left side, whereas no significant incremental response was observed over the ADM muscle. The PTF persisted for 6 minutes on the left and lasted for 2 minutes on the right trapezius muscle. No decremental response to slow rates of nerve stimulation was observed. Figure 1 shows the repetitive nerve stimulation recording of the right trapezius muscle.

The evaluation of cranial nerve pallsies rapidly followed by descending limb muscle weakness and respiratory failure were thought us an acute and severe neuromuscular disorder. The electrophysiologic findings including small evoked muscle action potentials, polyphasic, low amplitude, short duration MUPs and repetitive nerve stimulation test included PTF after 20 second exercise localized the disorder to the presynaptic region of the neuromuscular junction. Having a history of eating spaghetti with tomato sauce prepared at home and followed by nausea, vomiting and diarrhea 10 hours later suggested us the diagnosis of botulism. The patient was given only advance medical and supportive care and fully recovered approximately 2 months later.
**DISSCUSSION**

Botulism is an emerging disease that develops in a stereotypical pattern, with cranial nerve palsies followed by descending weakness and in severe cases respiratory failure, within 2-36 hours after ingestion of contaminated food. Autonomic signs include constipation, dry mouth, postural hypotension, urinary retention and pupillary abnormalities. Also; nausea, vomiting and diarrhea may occur early in the disease. Our patient’s clinical evaluation suggested the stereotypical pattern of botulism.

Electrophysiologic findings can provide presumptive evidence of botulism in patients with this typical pattern and the expected findings are as follows: 1- normal sensory conduction studies, 2- small motor action potential amplitudes with normal CVs and distal latencies, 3- infrequently, decrement in motor action potential amplitude to slow rates of nerve stimulation 4- PTF, usually between % 30-60, after rapid rates (50 Hz) or 10 sec of exercise is seen and lasts for 30-60 sec or persist for several minutes in cases of PTF of % 40 or more, 5- low amplitude, short-duration and polyphasic MUPs with spontaneous denervation potentials, 6- increased jitter and blocking by single-fiber EMG.

In our case we found small motor action potential amplitudes, polyphasic, low amplitude, short-duration MUPs with spontaneous denervation potentials and a PTF of approximately % 60 in two muscles after 20 seconds of exercise. These findings indicated the neuromuscular junction, especially the presynaptic region. Lambert Eaton myasthenic syndrome (LEMS), is an important diagnostic challenge at this point. A PTF of more than % 40 is known to be significant in botulism and may be absent in severely affected muscles whereas PTF is found in all limb muscles and often twofold or more than botulism in LEMS. Also, PTF of % 40 or more can persist for several minutes in botulism as seen in our case and lasts for only 30-60 sec in LEMS. The other similar acute neuromuscular diseases such as Guillian-Barre syndrome characterized by ascending weakness, demyelinating neuropathic findings and elevated CSF protein, Miller-Fisher variant characterized by absent deep tendon reflexes and diphtheritic neuropathy with tonsillar exudates and neuropathy, may come to mind in the differential diagnosis. Myasthenia gravis is ruled out due to the severe progressing pattern of the patient’s disease beginning acute after nausea, vomiting and...
diarrhea, accompanied by autonomic signs and the electrophysiologic findings consisting of small, short-duration MUPs with spontaneous denervation potentials, PTF after 20 sec exercise and lack of decremental response. The clinical evaluation of the disease and the electrophysiologic findings, locating the lesion to the presynaptic region of the neuromuscular junction, helped us to exclude these probabilities and diagnose the case as classic botulism.

Botulism is a rare but severe and rapidly progressing neurological disease, which is important to diagnose, as early as possible and beginning medical treatment. We believe that detailed electrophysiologic studies have an important role in the early diagnosis of botulism, especially when the bioassay studies can not be performed.

REFERENCES

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