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Electrodiagnosis in Cranial Botulism

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Abstract

Botulism is an acute neurologic disorder that causes potentially life-threatening neuroparalysis resulting in weakness and flaccid paralysis. In addition to the autonomic nervous system, botulism may involve only the cranial nerves, thus posing special diagnostic problems. Almost all cranial nerves are not easily testable. The anatomical and physiological characteristics of the spinal accessory nerve makes it very suitable to electrophysiological diagnosis of cranial botulism.

Although with a working diagnosis of botulism, treatment should not be delayed pending neurophysiological tests, electrophysiological studies including spinal accessory nerve may assist in diagnostic differentiation between cranial botulism and other disorders with similar clinical presentations such as Miller-Fisher and myasthenia gravis.

Keywords

botulinum toxin; repetitive nerve stimulation; single supramaximal stimulaton; spinal accessory nerve

Introduction

Botulism is an acute neurologic disorder that causes potentially life-threatening neuroparalysis due to a toxin produced by *Clostridium botulinum*. Toxin binding blocks acetylcholine release, resulting in weakness and flaccid paralysis. Clinical botulism develops in a stereotypical pattern: cranial nerve palsies are followed by descending weakness of the limbs and, in severe cases, respiratory paralysis. However milder cases with only cranial nerve involvement may occur [1].

Electrophysiological studies can be of help to the clinician in establishing the diagnosis of botulism. The most consistent electrophysiological abnormalities are [2]:

1) A small compound evoked muscle action potential (CMAP) in response to a single supramaximal stimulus (SSS) of the appropriate nerve.

2) Post-tetanic facilitation.

3) A decremental response of CMAP to a slow rate of stimulation.

A recent study validated SSS of a nerve before and after a 10 sec maximal voluntary contraction of the muscle as being a simple test that can be used to confirm the diagnosis of botulism [3]. For example, the cut-off percentage increment value of 25% has been considered both specific and sensitive in the

diagnosis of botulism in the case of a SSS applied to the ulnar nerve at the wrist [3]. These expected electrophysiological abnormalities, may not always be present especially in mild botulism. This may lead to electrodiagnostic difficulties and sometimes a delay in establishing a diagnosis [4]. In this regard, we need to remember that the botulism antitoxin is unlikely to be available onsite in most centers.

Case Report

Here we describe the electrophysiological profile in a case of botulism with abnormalities of cranial nerve muscles, absence of limb weakness and peripheral autonomic system involvement. The patient was a 33-year old man with an eleven-day history of rapidly progressing blurred vision, constipation, xerostomia, dysphagia, dysgeusia, dysarthria, associated with diarrhea, abdominal pain and urinary retention in the last six days. The patient referred that he eaten homemade ham the day before the symptom onset. Neurological examination showed bilateral mydriasis with very sluggish pupillary response to light and palpebral ptosis. There was a moderate weakness of upper trapezius and sternocleidomastoid muscles (Medical Research Council scale: 4). Muscle strength in the limbs and tendon reflexes were normal. He denied ingestion or application of medications with anticholinergic properties [5].

Motor and sensory nerve conduction studies (spinal accessory, ulnar, radial, peroneal, sural and tibial nerves) were performed and compared with those of our laboratory. Surface recording electrodes (Ag/ AgCl) were used. The spinal accessory nerve was stimulated just above the midpoint of the posterior border of the sternocleidomastoid muscle, where the nerve becomes superficial; the active recording electrode was located on the trapezius muscle approximately 5 cm lateral to the C7 spinous process on a line between this structure and the acromion and the reference electrode on the acromion; finally, a ground was placed between the stimulating and recording electrodes.

The nerve conduction studies were all normal except for the small CMAP amplitude of the spinal accessory nerve (Table 1). On SSS, the CMAP amplitude of the spinal accessory nerve was 36% larger than before exercise at diagnosis, whereas the SSS applied to ulnar nerve gave normal values. After therapy both baseline CMAP and SSS of the spinal accessory nerve yielded normal results. Repetitive nerve stimulation (RNS) was performed in the ulnar and spinal accessory nerve at diagnosis and 2.5 weeks after therapy. A significant decremental response was only evident in the spinal accessory nerve, disappearing in the follow up (Table 2). Standard needle EMG was performed and first dorsal interosseous, deltoid and upper trapezius muscles were examined. There was no evidence of spontaneous activity in any of these muscles and the pattern of recruitment was normal; in the trapezius there were polyphasic motor unit potentials.

The cerebrospinal fluid examination showed no abnormalities. Neither anti-GM1 nor anti-GQ1b antibodies (IgG) were present in serum of the patient. Magnetic resonance imaging of the brain did not show any abnormalities. Laboratory tests were not performed because of the long time which had elapsed from the onset of symptoms [6]. The patient was treated with antitoxin and symptoms fully disappeared after three weeks. The antitoxin was not available onsite and it was transported from an institute which was 250 km away.

Discussion

We report this case to highlight that occasionally botulism may involve only cranial nerves and the peripheral autonomic nervous system, thus posing special diagnostic problems. Almost all cranial nerves are not directly testable, and those which are testable usually show low amplitude (this is especially true in pre-synaptic syndromes) which reduces the sensitivity in detecting anomalies; in addition, their CMAPs are often contaminated by stimulus artifact. The superficial location of the spinal accessory nerve posterior to the sternocleidomastoid muscle allows easy access to stimulation and its high CMAP amplitude makes this nerve very suitable for the electrophysiological diagnosis of cranial botulism. This is a crucial point for two main reasons:

1) A spinal accessory nerve study may assist in diagnostic differentiation between cranial botulism (especially in cases with mild systemic symptoms) and other disorders with similar clinical presentations such as myasthenia gravis and Miller-Fisher syndrome when patients present with mild alterations of peripheral nerve conduction.

2) It can be used for the rapid assessment of patients with suspected botulism when serologic and toxologic confirmation cannot be quickly obtained.

It is important however to highlight that with a working diagnosis of botulism, treatment should not be delayed pending electrophysiological tests. The U.S. Food and Drug Administration has approved botulism antitoxin to treat patients showing signs of botulism with suspected exposure.

Tables

Table 1: Resting CMAP and single supramaximal stimulation results

	At diagnosis	2.5 weeks post therapy	Control values mean (range)
Resting CMAP (ADM)	9.6 mV	9.7 mV	9.7 (5 to 13) mV
Postexercise facilitation (ADM)	6%	5%	3.3 (-13 to 17) %
Resting CMAP (upper trapezius)	4.1 mV	8.2 mV	10.6 (4.9 to 16) mV
Postexercise facilitation (upper trapezius)	36%	2%	5% (-5 to 12) %

ADM: abductor digiti minimi; CMAP: compound muscle action potential. The control values are those of our laboratory.

Table 2: Repetitive nerve stimulation results

	At diagnosis	2.5 weeks post therapy	Normal values
RNS decrement at 2 Hz (ADM)	0 %	1 %	<10 %
RNS decrement at 2 Hz (Trapezius)	-17 %	-1 %	<10 %

RNS: repetitive nerve stimulation. ADM: abductor digiti minimi

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