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EC NEUROLOGY
Case Series

Botulism – Pearls and Pitfalls Based on the Description of Clinical Cases

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Received: December 01, 2021; **Published:** January 11, 2022

Abstract

Botulism is a severe infectious disease caused by neurotoxins produced by the bacterium *Clostridium botulinum*. Its neurological symptoms are characterised by symmetrical descending muscle paralysis with ophthalmoplegia and coexisting autonomic dysfunctions. In severe cases, respiratory deficiency can be present. The final diagnosis is confirmed when the botulinum toxin is detected from the patient's blood or other body fluids. The prompt identification of neurotoxin is crucial for the right treatment with antitoxin and significantly improves survival rate of the patients.

The numbers of botulism cases may vary between the countries and are related to eating habits and traditions. The most common is the foodborne disease but depending on the country and society structure, other types of botulism could be developed more often.

In this paper two clinical cases of patients with botulism are presented. There was no obvious history of possible intoxication in any of them. The author's aim is to emphasize that botulism can mimic more common neurological diseases and should be always taken under consideration in suspicious cases of sudden muscle weakness.

***Keywords**: Botulism, Foodborne Disease, Anticholinergic Syndrome, Repetitive Nerve Stimulation, Antitoxin*

Introduction

Botulism is a disease caused by a neurotoxin produced by *Clostridium botulinum* – an anaerobic, spore forming bacteria, widely spread in environment, mainly in the soil and in upper layers of water reservoirs. The spores are resistant to high temperature and standard methods of food processing.

The disease in humans is caused by botulinum toxins A, B, E, F. The certain toxins have different aminoacid structure. In Europe, most of the infections are caused by neurotoxin B.

Botulism is a rare disease, which prevalence differs geographically. Due to WHO, there are about 1000 cases of botulism reported annually in the world. Foodborne disease's outbreaks are most common in USA, with an average of 110 cases a year.

In Europe, the incidence of botulism is generally low (0,02 cases per 100,000 population) [2], however it might be underestimated.

There are 5 types of botulism, giving the same clinical picture: foodborne, intestinal, infant botulism, botulism caused by wound infection and aerogenic. In foodborne botulism, the poison is ingested through inappriopriately processed food in which the bacteria survive and discharge the toxin. Usually it is canned food, home - made pickled or fermented vegetables, tinned meat or fish as well as improperly stored honey.

The occurrence of various types of botulism depends on the region of the world, society structure and reflects different eating habits and preservation methods used in different households [1].

For example, nowadays in most of European countries there is an upward trend in occurrence of wound botulism. It is related to the usage of intravenous black tar heroine by drug addicts. However, the cases of classic botulism are still the most common. It is due to the fact that even though the food processing industry is designed to destroy spores and inactivate the toxins, the products of 'high risk ' are still popular.

The clinical symptoms of botulinum toxin infection occur because of the blocked release of acetylocholine in neuromuscular junction of motoneurons and in the synapses of parasympathetic nervous system [3-5]. In consequence, after the incubation period of several hours to 8 days the typical symptoms are presented. They include symmetrical cranial nerve palsy, with characteristic ophthalmoplegia, and descending symmetrical flaccid paralysis which could affect also respiratory muscles and lead to respiratory arrest. Neurological symptoms are usually preceded by nausea, vomitting, diarrhoea. Botulism is an afebrile disease with always preserved consciousness and intact sensory nerves. The parasympathetic symptoms incude dilated pupils with no reaction to light, constipation, dehydration and ileus.

In evaluation of the patient nerve conduction study and repetitive nerve stimulation are the most helpful. Typically, the presynaptic block is present, with abnormalities reminding Lambert-Eaton syndrome (LEMS).

The antitoxin is the most effective when administered within the first 24 hours from the symptoms, however it can be started up to 72 hours.

Case Studies

Case 1

A 63-year-old patient, a forester, presented with diplopia and blurred vision, swallowing problems and fatigue. He was reporting pain and dryness of the mouth and throat and because of that he had not been eating or drinking for two days. He was constipated and lost appetite.

On physical examination the patient was dehydrated. Neurologically he had dysarthria, absence of pharyngeal and palatal reflexes, dilated unreactive pupils and bilateral eyelid drooping, more on the right. There was general muscle weakness in the limbs and deep tendon reflexes were absent.

The blood test, CSF test and MRI were normal. After admission to the ward the patient was deteriorating fast and after few hours he developed complete ptosis, ophthalmoplegia and respiratory insuficiency. Electrodiagnostic studies showed that motor nerve CMAP amplitudes were decreased and in the repetitive nerve stimulation there was 47% of increase in the amplitudes after 20 s of maximal muscle activation.

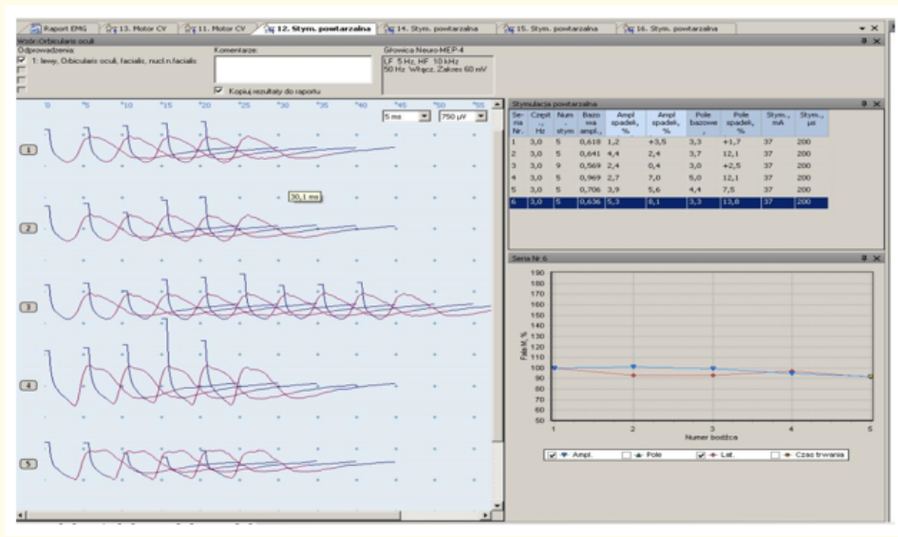

Figure 1: *Repetitive nerve stimulation No1.After 20s of maximal muscle activation: The 47 % increase in amplitudes.*

In the next step, the patient's blood sample was tested for the presence of botulinum toxin. The toxins B/E were detected and the treatment with antitoxin ABE was started. The patient needed intubation and ventillation at Intensive Care Unit, where he was hospitalised for nine days. After the discharge from ICU the patient underwent intensive rehabillitation and he left hospital after three weeks with no neurological deficits.

Case 2

A 58-year-old man was admitted to Neurology Ward because of slurred speech and dysphagia that appeared suddenly on the previous day. There was no history of food poisoning. The patient was admitted to Stroke Unit.

On admission the patient had elevated body temperture - 37,2 degrees and symptoms of pneumonia. Neurologically, there was slurred speech, absent pharyngeal and palatal reflexes, asymmetry of the face on the right and left-sided ptosis.He had dilated pupils with no reaction to light. Moreover, there was right- sided hemiparesis with brisk reflexes and with Babinski sign.

Chest X-ray showed bilateral pneumonia and in brain CT there were no acute lesions. MRI of the brain was planned, however the patient deteriorated rapidly and had to be intubated and ventilated mechanically. He was admitted to Intensive Care Unit with the diagnosis of brainstem stroke. After a week the sedation was withdrawn and the patient was extubated. He was conscious, with ophthalmoplegia and quadriplegia. All reflexes were absent.

Electrodiagnostic tests were performed and they showed very low amplitudes of CMAPs, slight decrease of amplitudes after 3 Hz stimulation and the increase in amplitudes of CMAPs after tetanic stimulus.

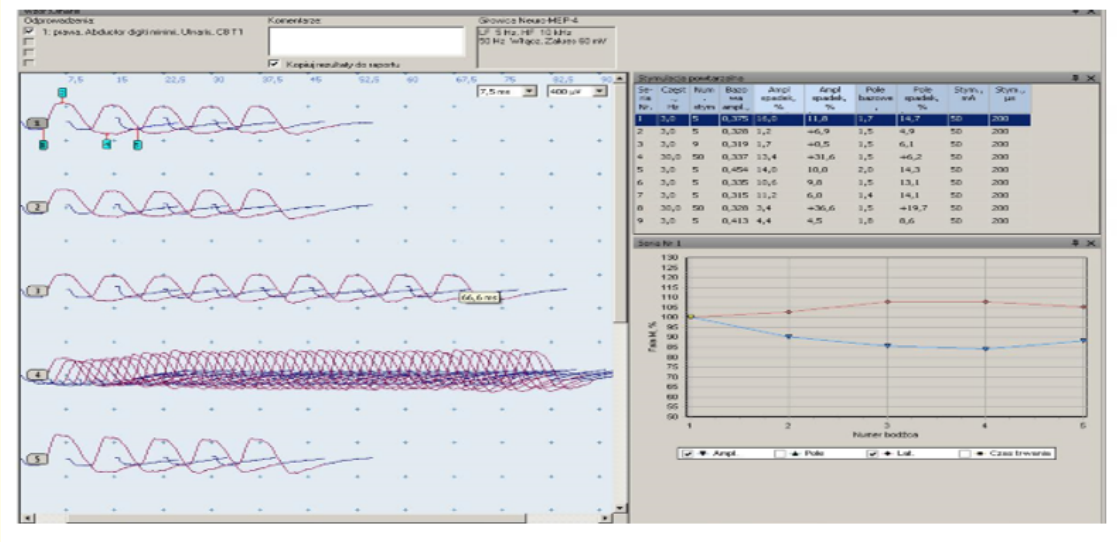

Figure 2: *Very low CMAP amplitudes. During the 3 Hz stimulation there was reduction of amplitueds 1-4 by 16 %. After tetanic stimulation there was 31-36 % of increase in amplitudes.*

After the results of electrodiagnostic tests, significant for botulism, the patient's blood sample was checked for the presence of neurotixins. Botulinum toxin B was detected and the patient was treated with antitoxin (started on the ninth day from the beginning of symp-

toms). Nevertheless, there was no significant improvement and finally the patient needed tracheostomy and feeding tube (PEG). He was discharged from hospital with general muscle weakness and was admitted to Rehabilitation Ward for many weeks.

Outcome

In both of cases there was no notable history of specific food eaten and actually, it is hard to tell whether they were foodborne infections or other kind of botulism. Both patients had pneumonia from the beginning of neurological symptoms, most probably due to aspiration.

Repetitive nerve stimulation (RNS) in both patients was abnormal, suggesting the presynaptic block. However, the increase of amplitudes after the tetanic stimulus was lower than 60% (the rise of 47% in the first case, the rise of 36% in the second case).(5)In typical presynaptic blocks, rapid rates of stimulation will produce a marked facilitatory response in excess of 60%.

In the patient number 1, tick paralysis and Guillain-Barre syndrome were suspected at first (the patient was working in the forest). Because the necessary tests were conducted quickly, the treatment with antitoxin was started very early – from the second day of the symptoms. This man was hospitalised for 27 days in total, having spent 9 of them at Intensive Care. He was discharged from hospital in good condition, with no neurological abnormalities.

The patient number 2 had an atypical course of intoxication and the asymmetry of the symptoms was very confusing. He was diagnosed with a stroke and electrodiagnostic tests and botulinum toxin detection were done late. Because of that the treatment was introduced in the eighth day from the beginning of the symptoms and the outcome of this patient was much worse. He spent 47 days in hospital, most of them at Intensive Care. He was discharged from hospital with quadriplegia, tracheotomy and PEG, requiring long rehabilitation.

Discussion

Botulism is still an actual epidemiological and clinical problem. The most important is to remember about this disease in differential diagnosis of a patient with acute muscle weakness. The diagnosis based on typical history and symptoms seems to be easy, but sometimes its course is untypical and the patient might not report eating suspected food.

For instance, by definition botulism is an afebrile disease. However, as shown in this paper, the patients might have high body temperature from the beginning of the symptoms and it could be due to coexisting infection. Aspiration pneumonia is one of the most common complications of botulism.

Rapid diagnosis is crucial for the treatment and the outcome of the patient. This is because the treatment with antitoxin should be started as soon as possible, preferably up to 72 hours from the first symptoms. Delays in this procedure may lead to respiratory insufficiency, prolonged hospitalisation and its consequences and may cause severe disability or even the death of the patient.

From epidemiological aspect, it is important to prevent botulism outbreaks by making people aware of it. The mass poisonings from commercial sources are extremely rare nowadays and most of infections are related to inappropriate food processing at the household. The problem is global though because of the open European borders and the facility of travelling and exporting the goods between the countries. The persistent tradition of preparing home-made tinned food in some households may be the cause of emergence of botulism in countries in which it was absent for years. For example, in 2006 in Ireland the first in 20 years case of foodborne botulism was diagnosed in a Polish person who injested the toxin from the canned food sent to him from Poland for Christmas [6].

Conclusion

Botulism is a rather rare case of muscle paralysis and it is often forgotten in clinical practice. This is due to the fact that the typical symptoms are not always clear and may mimic other common neurological diseases like stroke, acute polyradiculoneuropathy or neu-

roinfection. Moreover, as the foodborne intoxication is the most common and the one that everyone remembers when thinking about botulism, it is crucial to know that other ways of infection are possible. Very often, botulism is being ruled out hastily at the beginning of diagnostic process, just after negative eating history.

The most important when dealing with this disease is to be familiar with the symptoms and avoid unnecessary, time-consuming and expensive diagnostics tests like MRI. Flaccid paralysis can be fully reversible if the right diagnosis is set up without delays. On the other hand, the disease could be avoided if people take precautions on their eating habits. Therefore, informative campaigns about botulism should be transmitted both within the community and in medical circles.

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Volume 14 Issue 2 February 2022