Acute paralysis following "a bad potato": a case of botulism

[Paralysie aiguë après une «mauvaise patate» : un cas de botulisme]

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Purpose: Intensivists often encounter patients with respiratory failure as a result of neuromuscular disease, however, acute neuromuscular syndromes are less common. We present a case of food borne *Clostridium botulinum* and discuss the diagnostic and therapeutic considerations.

Clinical findings: A 35-yr-old healthy male presented with abdominal pain and blurred vision 12 hr after ingesting a "bad" potato. During the next 17 hr, the patient demonstrated a gradual descending paralysis which ultimately resulted in no cranial nerve function and 0/5 strength in all extremities. Sensation was intact. The patient required intubation and mechanical ventilation. His blood count, biochemical profile, computerized tomography and magnetic resonance imaging of the head were normal. A lumbar puncture revealed no abnormalities. Due to the rapid deterioration and presentation of 'descending' paralysis, botulism was suspected. The patient was treated empirically with botulinum anti-toxin. Samples of blood, stool and gastric contents were cultured for the presence of *Clostridium botulinum* and its toxin and these tests were positive for botulinum toxin A 12 days later. The patient's neuromuscular function gradually improved over a prolonged period of time. Six and one-half months after his initial presentation, the patient was discharged home after completing an aggressive rehabilitation program.

Conclusions: Botulism is a rare syndrome and presents as an acute, afebrile, descending paralysis beginning with the cranial nerves. If suspected, botulinum anti-toxin should be considered, particularly within the first 24 hr of onset of symptoms. Confirmation of the presence of botulinum requires days therefore the diagnosis and management rely on history and physical examination.

Objectif: Beaucoup de patients présentent une insuffisance respiratoire causée par une affection neuromusculaire, les syndromes neuromusculaires aigus étant plus rares. Nous présentons un cas d'intoxication alimentaire à *Clostridium botulinum* et discutons du diagnostic et du traitement.

Éléments cliniques : Un homme sain de 35 ans a consulté pour des douleurs abdominales et une vision floue 12 h après avoir mangé une «mauvaise patate». Pendant les 17 h suivantes, une paralysie descendante graduelle s'est installée et a finalement provoqué la perte de fonction des nerfs crâniens et réduit à 0/5 la force des extrémités. La sensation était intacte. Il a fallu intuber et ventiler le patient. La numération plaquettaire, le profil biochimique, la tomographie par ordinateur et l'imagerie par résonance magnétique de la tête étaient normaux. Une ponction lombaire n'a révélé aucune anomalie. Devant la détérioration rapide et la paralysie «descendante», nous avons pensé au botulisme. Un traitement empirique avec une antitoxine botulinique a été administré. Les échantillons de sang, de selles et du contenu gastrique ont été mis en culture pour vérifier la présence du *Clostridium botulinum*. La présence de la toxine botulinique type A a été révélée 12 jours plus tard. La fonction neuromusculaire s'est graduellement améliorée sur une longue période. Six mois et demi après sa première visite, le patient a pu partir après un programme de réadaptation complet et énergique.

Conclusion : Syndrome rare, le botulisme se présente comme une paralysie aiguë, afebrile et descendante affectant d'abord les nerfs crâniens. S'il est soupçonné, il faut penser à l'antitoxine botulinique, surtout au cours des 24 premières heures de l'apparition des symptômes. La confirmation de la présence de *Clostridium botulinum* demandant des jours, le diagnostic et le traitement reposent sur l'histoire et l'examen physique.

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PATIENTS who suffer from acute neuromuscular disease often require the support of the intensive care unit (ICU) for acute respiratory failure. Although intensivists often encounter chronic respiratory failure in patients with long standing neuromuscular problems such as Guillain-Barré syndrome, the acute syndromes may not be as familiar to these practitioners. We encountered such a case in our multidisciplinary ICU.

Case report

A 35-yr-old male with no history of serious medical problems was well until 28 hr prior to presentation to our medical centre. The evening before, he had eaten supper with his family at a local restaurant. During his meal, he had taken a mouthful of a foil-wrapped baked potato and found it to be foul tasting and spat it out. The potato was immediately disposed of. He felt slightly nauseated but finished his meal. Approximately 12 hr later he awoke with abdominal pain and blurred vision. These symptoms progressed over the next three hours prompting him to go to the local Emergency Department where he was noted to have nystagmus, double vision, and slurred speech. A stroke syndrome was suspected and he was transferred to the regional hospital and was found to have the following pertinent findings: blood pressure 156/88 mmHg, heart rate 110 beats·min⁻¹, respiratory rate 22 breaths·min⁻¹, normal temperature and oxygen saturation 98% on room air. The neurological examination revealed multiple abnormalities including bilaterally reduced visual acuity, nystagmus and diplopia on extremes of gaze, dysphagia, dysphonia, extremity weakness in all limbs with a normal sensory examination and normal plantar response. The remainder of the physical examination was normal. He was then transferred to London Health Sciences Centre for further neurological assessment, and arrived approximately 28 hr from ingestion of the potato. Examination at this time revealed a Glasgow coma scale of 4. His blood pressure was 120/78 mmHg, heart rate 123 beats·min⁻¹, and respiratory rate of 35 to 40 breaths·min⁻¹ with minimal effort with an oxygen saturation of 93% on 100% oxygen via face mask. The arterial blood gases revealed a significant respiratory acidosis (pH = 7.12, PO₂ = 63 mmHg, PCO₂ = 94 mmHg, HCO₃ = 29 mmol·L⁻¹). The trachea was intubated and the lungs ventilated for airway protection and Type II respiratory failure, respectively. Upon re-examination of his neurological status, it was apparent that further deterioration had occurred. Within one hour of his transfer, he had no cranial nerve function and 0/5 strength in his upper and lower extremities. Pupillary reflexes were equal and minimally reactive to light, and

deep tendon reflexes were present. The initial complete blood count, biochemical profile and computerized tomography and magnetic resonance imaging scans of the head were all normal. A lumbar puncture demonstrated a normal opening pressure, cell count and glucose and protein content. Gram stain of the cerebral spinal fluid revealed no organisms.

Because of the rapid deterioration of a descending paralysis following the ingestion of a spoiled potato, botulism was strongly suspected. Cultures of blood, stool, gastric contents, and samples of food from the restaurant were sent for analysis for the presence *Clostridium botulinum* and its toxin. Unfortunately, the implicated potato was not available for testing. The decision was made to treat the patient for possible botulism poisoning and the botulism anti-toxin (Botulism Anti-toxin Trivalent (Equine) types A, B and E, Aventis Pasteur Limited, Toronto, ON, Canada) was administered after skin testing for hypersensitivity without any important adverse effect. The patient did not have any further neurological deterioration after the administration of the anti-toxin.

Results of the cultures and the toxin studies returned positive for both *Clostridium botulinum* and botulinum toxin A approximately 12 days after admission. The patient had a prolonged course in the ICU with a slow neuromuscular recovery. He was treated for an aspiration pneumonia during the first week of admission. He eventually required a tracheostomy and a gastro-jejunostomy feeding tube for respiratory and nutritional support, respectively. Upon discharge to his local hospital, the patient was able to open and close his eyes, had 1–2/5 strength in the upper extremities and could withdraw his lower extremities. Approximately six and one-half months after initial presentation, the patient was discharged home after completing an aggressive rehabilitation program in his local hospital. He did not have any requirements for long-term care. The local health department investigating this case reported no other cases.

Discussion

Clostridium botulinum is a gram positive, spore forming bacillus found naturally in soil and marine sediment and on the surfaces of fruits, vegetables and seafood.¹ Low acidity and oxygen tension are required for spore growth and toxin production. The spores are heat resistant and survive most conventional treatment processes. Estimates suggest approximately 100 to 110 cases per year of botulism occur in the United States (U.S.A.), primarily as outbreaks *vs* sporadic cases.¹ There are four common manifestations of the disease: foodborne, infantile, wound, and adult enteric

botulism. Infantile botulism is the most common overall² in the U.S.A., however, in Canada, foodborne botulism is most prevalent, especially among the Inuit.³ *Clostridium* species produce eight different toxins but toxins A, B, E cause the majority of disease; however, there are reports that toxins F and G also can result in disease.^{1,4} Toxin E is the most common toxin leading to infection in Canada. The toxins are made of a heavy and light chain and bind to the pre-synaptic terminal of peripheral cholinergic receptors.⁵ The heavy chain binds to the receptor, while the light chain undergoes endocytosis and is taken into the cell and blocks the release of acetylcholine. Once bound, the process is irreversible and the nerve terminal is rendered permanently inactive. Return of function occurs with growth of new terminals and formation of a new synapse. This process can take up to eight months.¹⁻³

The classical presentation of *Clostridium* botulism is one of an acute, afebrile, descending paralysis beginning with cranial nerve abnormalities as early as eight hours after exposure. There are no sensory deficits and deep tendon reflexes may be preserved early on. Depending on the route of infection, a prodrome of nausea, vomiting and diarrhea may occur. In infants, poor feeding, lethargy, a weak cry, decreased sucking and lack of muscle tone may be presenting signs.

The differential diagnosis of a patient who presents with acute neuromuscular dysfunction is broad (Table). The history and physical examination are important in helping discern the etiology of a patient's illness, as are the appropriate investigations. Initial investigations in patients who present with acute neuromuscular disease should include complete blood count, electrolytes, creatinine, blood urea nitrogen, glucose, cultures of appropriate body fluids, electromyography, chest radiography, computerized tomography and magnetic resonance imaging of appropriate anatomy and analysis for botulinum toxin if suspected by the history and presentation. Toxin type is identified by injecting mice with the source of botulism toxin and various type specific anti-toxins are administered to determine which prevents death.⁶ ELISA and PCR testing to replace this method remain controversial.⁷ Electromyography is also helpful in the diagnosis, with the most common finding with botulism poisoning being small evoked potentials in response to a single maximal stimulus.⁸

Treatment of botulism is centered around early administration of the botulinum anti-toxin. It is an equine trivalent anti-toxin made from horse globulins that are modified by enzymatic digestion. The anti-toxin is effective against toxins A, B and E. The anti-

TABLE* Differential diagnosis of acute neuromuscular dysfunction

<i>Condition</i>	<i>Distinguishing features</i>
Guillain-Barré syndrome	History of infection; paresthesias; ascending paralysis; areflexia; high CSF protein; EMG
Myasthenia gravis	Recurrent paralysis; + edrophonium test; EMG
Stroke	Asymmetric paralysis; imaging; risk factor
Lambert-Eaton myasthenia syndrome	History of lung cancer; increased strength with activity; EMG
Tick paralysis	Paresthesias; ascending paralysis; exposure
Sepsis/meningitis	History and physical; CSF
Poliomyelitis	Febrile; asymmetric paralysis; CSF

CSF = cerebral spinal fluid; EMG = electromyography.

*Adapted from Arnon *et al.*⁴

toxin binds only to free circulating toxin and has no effect on nerves already affected. Thus, the goal of administration is to reduce the number of peripheral nerves affected by the circulating toxin. The product monograph suggests that one vial should contain enough anti-toxin to be effective in patients exposed, but the suggestion is made that if symptoms progress after the initial dose, further doses may be of benefit.⁹ The main side effects are those of hypersensitivity and anaphylaxis, both of which are more common with repeat dosing. Thus, a bedside challenge prior to administration is required. This requires an intradermal injection, as per tuberculin skin testing, and if a wheal or flare occurs at 30 min, anti-toxin administration is not recommended. Also of importance is awareness of a delayed serum sickness, which could develop up to 14 days after treatment. Despite the risk of side effects, it is clear from retrospective reviews that the anti-toxin is beneficial to patients and mortality is substantially reduced with its administration, especially within a 24 hr onset of symptoms.¹⁰ Also, in two series of cases of wound botulism, the earlier the administration of anti-toxin, the more favourable the clinical outcome.^{11,12} Our patient received anti-toxin relatively early at 24 hr after the development of symptoms. Despite having severe manifestations of botulism at that time, anti-toxin was still indicated and may have prevented further neurological impairment and shortened the duration of his illness. An experimental heptavalent (A,B,C,D,E,F,G) anti-toxin has had success in the treatment of botulism but it is not as readily available.¹³

Other therapeutic modalities have been considered but little evidence supports their use. Antibiotics for wound botulism is recommended by some for adjunctive therapy with anti-toxin and wound debridement. Aminoglycosides should not be used in any capacity, as they have been shown to potentiate the neuromuscular blockade, especially in infants.¹⁴ Other considerations include the use of laxatives, enemas and other cathartics in foodborne botulism, provided there is no significant ileus present. Induced vomiting and gastric lavage may be considered if ingestion is recent; no evidence exists supporting the use of activated charcoal. In addition, patients may require significant supportive care including ventilation, nutrition, physiotherapy and occupational therapy. Patients are expected to survive their illness, but persistence and vigilance on the part of the health care team is required.

Botulism, although a rare illness, is treatable and reversible but requires the physician to be thoughtful of it as a possible etiology of disease. Its presentation is dramatic and its effects are severe and debilitating. Effectiveness of treatment depends on early recognition and administration of anti-toxin. *Clostridium botulinum* is one of many potential biological weapons that are of global concern and, in these unsettled times, it may be an illness requiring our attention. *Clostridium botulinum* is a reportable disease and all cases should be referred to Health Canada Botulism Reference Service for Canada. Early communication may limit the spread of a very serious disease process.

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