ACG CASE REPORTS JOURNAL

CASE REPORT | ESOPHAGUS

Rapidly Progressive Muscle Paralysis and Acute Respiratory Failure Following Endoscopic Botulinum Toxin Injection

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ABSTRACT

Botulism toxin injection (BTI) is a well-known and relatively safe endoscopic treatment for achalasia. We report a case of a 90-year-old female diagnosed with achalasia who subsequently underwent BTI with symptomatic relief. The therapy was complicated by systemic botulism, however, leading to progressive muscle paralysis with diaphragmatic involvement requiring mechanical ventilation support. This is the first reported case of BTI for achalasia causing systemic botulism.

INTRODUCTION

Botulinum toxin injection (BTI) for achalasia is a well-tolerated procedure with few side effects, such as transient chest pain and heartburn; rarely, cases of mediastinitis, esophageal mucosal ulceration, and pneumothorax have been reported.[1-4]

CASE REPORT

A 90-year-old female presented with epigastric pain and coffee-ground emesis. An esophagogastroduodenoscopy revealed a tortuous and dilated esophagus with large amounts of retained undigested food matter. A barium swallow study showed markedly decreased lower esophageal peristalsis with a characteristic bird's beak appearance, consistent with achalasia. Given the classic findings on barium swallow study, manometry was not performed. The patient underwent endoscopic BTI of 80 units botulinum toxin in the areas around the lower esophageal sphincter. She tolerated the procedure well, and there were no intraprocedural complications. Following the procedure, the patient noted significant symptomatic improvement and was able to tolerate a diet. Within hours of the procedure, however, the patient was noted to have profound neuromuscular weakness of her upper extremities. Neurologic exam revealed 0/5 deltoid, 3/5 biceps, 2/5 triceps, 4/5 wrist extension, and 4/5 intrinsic hand muscle strength bilaterally. There was decreased muscle tone and absent reflexes in the upper extremities bilaterally without fasciculations, and sensory exam was entirely intact. Lower extremity as well as cranial nerve neurologic exams were unremarkable, and the patient remained conversant and alert.

At this time, the differential diagnosis included atypical Guillain-Barre syndrome (GBS), cervical cord compression, and systemic botulism as a complication of endoscopic BTI. Magnetic resonance imaging of the cervical spine ruled out cord compression, and a lumbar puncture ruled out GBS by absence of characteristic albuminocytologic dissociation. Computed tomography of the head did not yield any acute intracranial pathology or evidence of cerebrovascular accident. Given the time course of her symptoms, it was determined that the patient was suffering from botulism as a result of endoscopic BTI. The Centers for Disease Control (CDC) was notified, and heptavalent equine antitoxin was administered via infusion (0.5 mL/min initially, then increased to 2 mL/min) 2 days after initial symptom onset. Serum samples for botulism were collected prior to antitoxin administration and were sent to the

ACG Case Rep J 2016;3(4):e166. doi:10.14309/crj.2016.139. Published online: November 23, 2016.

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CDC. However, despite antitoxin therapy, the patient's condition progressed over the next several days, leading to generalized muscle paralysis with involvement of her diaphragm, ultimately requiring mechanical ventilation in the intensive care unit. She remained there for 4 months without improvement in clinical status, with her course further complicated by ventilator-associated pneumonia, urinary tract infection, and decubitus ulcers. The patient's condition was compounded by critical illness polyneuropathy, diagnosed by electromyography, for which she remained ventilator-dependent until withdrawal of care by her family.

DISCUSSION

Botulism is a rare neuroparalytic condition resulting from a neurotoxin expressed by *Clostridium botulinum*. The clinical features of this disease are typically acute onset of symmetrical descending weakness with the absence of fever, sensory deficits, and unresponsiveness.[5] Diagnosis is made by clinical features as well as serum and fecal testing, although it is difficult to grow the bacteria in complete anaerobic conditions, which yields a high rate of false-negatives. The treatment of botulism is administration of antitoxin followed by supportive care as well as intensive physical therapy.[5] Overall, with prompt recognition and treatment, botulism has a relatively low mortality rate of <5–8%.[5]

This exceedingly rare case of severe botulism following endoscopic BTI raises important concerns over the safety profile of the procedure. While previously thought to have virtually no risk of causing generalized paralysis due to the low dose of toxin used for treatment, physicians must now be cognizant of the possibility of this catastrophic complication to help patients make informed decisions. Moreover, rare occurrences of iatrogenic botulism seen with BTI in higher doses used for other disorders are often mild and typically occur in patients with underlying neuromuscular disorders, which was not the case in our patient.[2] We hypothesize that the inadvertent injection of toxin into a nearby blood vessel at the esophageal sphincter caused systemic circulation of the toxin, which resulted in severe botulism. The low muscle bulk in this 90-year-old female may be a contributing factor.

DISCLOSURES

Author contributions: All authors contributed equally to manuscript writing, editing, and approval. G. You is the article guarantor.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received March 29, 2016; Accepted June 17, 2016

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