

Expanded Case Summary 4: Botulism.

This case details the presentation and management of a patient admitted to the intensive care unit following a respiratory arrest with potential hypoxic brain injury. This case is notable as his presenting clinical features were consistent with suspected botulism. This complicated all aspects of his ICU care, particularly prognostication and providing information to relatives.

Clinical Problem

A young male intravenous drug abuser was referred to the medical assessment unit with a 3-day history of double vision, difficulty speaking, shortness of breath, productive cough and generalised weakness. He was found unresponsive in the acute assessment area and was successfully resuscitated from a hypoxic PEA cardiac arrest. It was unknown how long this patient was in cardiac arrest. He was transferred to ICU for further management and treatment of suspected botulism.

Word Count – 73

Management

On arrival to the intensive care unit we:

- Implemented lung protective ventilation with recruitment strategies to assist oxygenation. Neuromuscular blockade was utilised to support this.
- Commenced IV antibiotics to cover botulina infection, which included flucloxacillin, clindamycin, metronidazole, gentamicin and benzylpenicillin. CT pelvis/groin excluded soft tissue collection and abscess formation of recent injection sites.
- Commenced on botulism anti-toxin therapy.

Subsequent imaging (CT head) 12-hours post cardiac arrest was consistent with hypoxic brain injury. A lumbar puncture was performed which helped exclude inflammatory conditions such as a Gullian-Barre variant (for example, Miller Fisher Syndrome).

His family were made aware at an early stage of the potential for irreversible brain damage but the assessment of this was difficult due to suspected botulism infection.

Although his cardiorespiratory function improved with supportive care his neurology never fully recovered despite over six weeks of supportive care. At

best his eyes were opened spontaneously and he would extend to pain with up-going plantar reflexes. He retained spontaneous breathing. Clinically this was consistent with irreversible hypoxic brain injury. Ancillary testing was performed to augment this diagnosis. MRI brain was inconclusive however his EEG was consistent with hypoxic brain damage. Nerve conduction studies excluded neuromuscular disorders such as GBS.

After four weeks a tracheostomy was placed in order to facilitate weaning from mechanical ventilation and to allow the opportunity for potential discharge to a long term rehabilitation unit. He was discharged from ICU to a medical ward and following discussions with his family it was agreed that we would not escalate to ICU level care as he failed to make any neurological recovery during his prolonged ICU admission. His family were in agreement and understood the poor prognosis given poor recovery, severity of illness and evidence of hypoxic brain damage. Unfortunately, this patient died a few weeks later in the medical ward from a pneumonia despite antibiotic therapy.

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Discussion:

Background

Botulism is a condition characterised by descending flaccid paralysis caused by the neurotoxin produced by the bacterial organism *Clostridium botulinum*. This gram-positive anaerobic bacteria is commonly found in soil. Spores can be transmitted via food, intestinal proliferation or from wound contamination. As there has been recent epidemic of contaminated heroin, we must be vigilant when assessing unwell intra-venous drug abusers and be mindful of botulism.

Presentation

The neurotoxin produced by *C. botulinum* irreversibly binds to the presynaptic nicotinic acetylcholine receptors in the neuromuscular junction.

Traditionally the clinical features of botulism include:

- Diplopia/blurred vision
- Facial weakness
- Bulbar palsy with swallowing difficulties, dysphonia, dysarthria and airway obstruction
- Generalised malaise, especially arm weakness with preserved sensation
- Dyspnoea, respiratory muscle weakness, respiratory failure
- Autonomic instability.

The spectrum of illness can vary from mild prodromal illness to rapid airway obstruction, respiratory failure and death. Early recognition and treatment is therefore vital.

Differential diagnosis and Investigations

The differential diagnosis here included Guillain-Barre syndrome (particularly GBS variants such as Miller-Fisher Syndrome) which is an acute inflammatory demyelinating polyneuropathy. Myaesthesia gravis and organophosphate poisoning should also be considered.

Investigations should include:

- Wound microscopy/bioassay to isolate bacteria and neurotoxin
- EMG
- CT/MRI
- Lumbar puncture
- Nerve conduction studies

Management

- Antibiotic treatment and surgical drainage of abscess collections,
- Anti-toxin treatment to prevent further acetylcholine receptor antagonism. This does not affect receptors already bound by the toxin which highlights the importance of rapid recognition and treatment.
- General respiratory support, airway protection and cardiovascular support to treat autonomic instability
- Avoidance of drugs that may potentiate neuromuscular inhibition such as aminoglycosides and magnesium
- Regular assessment of cough and gag reflexes

Recovery is generally slow and may take months to fully regain neurological function. The mortality rate is between 5-10%, generally due to respiratory failure, airway obstruction and aspiration pneumonia. Early antitoxin treatment is associated with an improved outcome in a retrospective analysis of 134 cases.

Application to case

Our patient was particularly challenging. Upon arrival in ICU he received targeted and aggressive management, which was consistent with the recommended guidance. He received early antitoxin therapy, broad spectrum antibiotic cover as well as multi-organ support.

This patient's chances of survival were low as he had already suffered a cardiac arrest, significant aspiration pneumonia and hypoxic brain damage. The decision to site a tracheostomy was therefore an ethical dilemma. There was concern initially that siting a tracheostomy would be a mode of death in a patient who had no capacity to make any form of neurological recovery. However, after MDT and family discussions it was felt tracheostomy would be a safeguard in case any recovery was possible. In hindsight this may not have been the best course of action and perhaps this delayed the inevitable. However as there are cases of botulism that recover after a period of months it was not an unreasonable course of action.

I can find no case reports or literature which comment on differentiating botulism from hypoxic brain damage. Most case reports are positive and document the benefits and success of early recognition and treatment. An interesting article did comment that their diagnosis was delayed because the patient was a drug addict and intoxicated with benzodiazepines. Perhaps this was a contributing factor in our patient and why botulism was overlooked initially.

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Lessons learnt

This was a difficult case. It was frustrating that his family felt they received mix messages. Case-note review of family discussions appeared to be consistent.

It was particularly difficult as his cardiac arrest was preventable if botulism and its potential to cause rapid airway obstruction had been considered earlier. The diagnosis and treatment of botulism was only considered at ICU admission, by which point he was in multi-organ failure. This highlights the importance of vigilance and keeping a wide differential diagnosis particularly when treating intravenous drug abusers who are susceptible to multiple pathologies which may be masked by the sedative effects of drugs.

This case has supported my development as I have gained experience managing a rare condition, coupled with the complexities of excluding differential diagnoses which may alter patient management and undertaking difficult discussions with family. Continued discussions with family along with regular consultation with all members of the ICU team and MDT are important to reach a unified consensus on the best course of action.

Word Count – 167

References

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