May 2017 Critical Care Case of the Month

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History of Present Illness

A 54-year-old Hispanic male who was incarcerated 3 days prior to hospital admission was brought into the emergency room from prison for alcohol related withdrawal seizures.

Physical Examination

Upon arrival to the ER, the patient was noted to be hypoxic with copious thick secretions in his mouth. He was intubated for airway protection, started on propofol and fentanyl drips as well as intravenous thiamine and folic acid.

Radiography

A chest radiograph was performed (Figure 1).

Figure 1. Portable anterior-posterior (AP) radiograph of the chest.

Which of the following are **true** regarding management of this patient?

1. Phenytoin should be administered for prevention of seizures
2. Prophylactic antibiotics for aspiration pneumonia should be administered
3. Thiamine and folic acid should be administered
4. 1 and 3
5. All of the above

Correct!

3. Thiamine and folic acid should be administered

Benzodiazepines are preferred and are considered first-line treatment in patients with alcohol withdrawal (1). These agents reduce the symptoms of withdrawal including seizures and help to prevent symptom progression. Phenytoin is ineffective in preventing alcohol withdrawal seizures (1).

Due to the likelihood of nutritional deficiencies in patients with alcohol withdrawal, , thiamine and folic acid should be routinely administered (1). Folic acid supplementation and thiamine 100 mg daily are recommended to prevent the development of Wernicke's encephalopathy.

For aspiration pneumonitis, early prophylactic antibiotics are not recommended (2). This practice is believed to lead to the selection of more resistant organisms.

Sedation was discontinued and the patient became responsive. On the third day a spontaneous breathing trial was initiated which the patient passed. However, shortly after extubation, he required reintubation for tachypnea and hypoxemia. He was noted to have a poor cough and thick oral secretions. Despite again passing a spontaneous breathing trial, he again failed extubation on day five. A repeat chest x-ray was performed (Figure 2).

Figure 2. Repeat portable AP radiograph of the chest.

Which of the following **should be performed?**

1. Bronchoscopy
2. Thoracentesis of the left pleural effusion
3. Ultrasound of the left chest
4. 1 and 3
5. All of the above

Correct!

4. 1 and 3

There is opacification of the left chest on the radiograph. There is also volume loss on the left side. Note the deviation of the trachea to the left. A significant pleural effusion is unlikely with volume loss present.

The most likely cause of the volume loss in this patient's situation is retained secretions in the left lung. The left lung is more difficult to suction than the right since the right mainstem bronchus is more in line with the trachea while the left mainstem bronchus is more at an angle making passage of the suction catheter into the left lung more difficult. Bronchoscopy was performed and copious amounts of secretions were removed from the left lung.

The patient was noted to have left-sided ptosis and decreased power 3/5 in his proximal upper and lower extremities. Reflexes were intact. Magnetic resonance imaging of the brain and brainstem were unremarkable. The possibility of diaphragmatic weakness was raised and ultrasound of the left diaphragm was performed (Figure 3).

Figure 3. Still image from diaphragmatic video. The echogenic edges of the diaphragm are labeled with solid white arrows, as is the characteristic central echogenic line (empty arrow) of the diaphragm. The liver is labelled with an "L".

Diaphragm thickening fraction (DTF) assessed by ultrasound was calculated using the following formula: Thickness at end inspiration - Thickness at end expiration / Thickness at end expiration. This patient's DTF was found to 55%.

Which of the following are **true**?

1. Diaphragmatic pacing should be considered
2. The diaphragm as assessed by ultrasound is normal
3. The high DTF predicts respiratory failure after extubation
4. 1 and 3
5. All of the above

Correct!

2. The diaphragm as assessed by ultrasound is normal

The cause of the retained secretions is unclear but diaphragmatic ultrasound and DFT are normal. The diaphragm normally thickens during inspiration. A DFT < 0.36 predicts failure of extubation (3).

Which of the following **should be done next?**

1. Contact the patient's family
2. Negative pressure ventilation
3. Percutaneous tracheostomy
4. 1 and 3
5. All of the above

Correct!
4. 1 and 3

The patient has failed several extubation attempts and percutaneous tracheostomy should be performed. This was done and the patient was quickly weaned from mechanical ventilation.

Attempts to contact the patient's family required permission from the prison warden where the patient was incarcerated and so was delayed. Eventually they were contacted and mentioned that the patient had a diagnosis of oculopharyngeal muscular dystrophy (OPMD).

Which of the following are **true** regarding OMD?

1. OPMD has been associated with respiratory failure because of retained secretions and aspiration
2. OPMD is associated with acetylcholine receptor antibodies
3. OPMD has not been reported in Hispanics
4. 1 and 3
5. All of the above

Correct!

1. OMD has been associated with respiratory failure because of retained secretions and aspiration

The work up of this patient with recurrent respiratory failure without severe pulmonary disease suggested a neuromuscular component. The episodes of poor airway clearance, atelectasis, contributed to this suspicion. The patient was alert and able to follow commands, and without a lateralizing deficit, suggesting a more peripheral cause of neuro-muscular abnormality. Diaphragmatic weakness was excluded as a cause via ultrasound, myasthenia gravis and Guillain-Barre were found to be less likely due to the exam and the laboratory finding of a negative acetylcholine receptor antibody.

Oculopharyngeal muscular dystrophy (OPMD) is an increasingly recognized autosomal dominant, late onset neuromuscular disorder primarily characterized by ptosis, pharyngeal muscle weakness and proximal skeletal muscle weakness (4). It is most prevalent in the French-Canadian population from Quebec; however, in recent years there has been an unexpected large OPMD population of Hispanic offspring identified in New Mexico (5).

We report a rare case in which OPMD complicated mechanical ventilation management in a 54-year-old Hispanic gentleman admitted to the ICU for respiratory failure secondary to alcohol withdrawal seizures. To our knowledge, this is the first case report of OPMD in the Hispanic population resulting in tracheostomy placement. The first case report of OPMD complicating airway management was reported by Christopher *et al.* (4) in an 83-year-old male of French-Canadian descent after elective surgery.

Despite frequent and aggressive airway clearance therapy, OPMD resulted in re-intubation, tracheostomy placement, aspiration, extensive neurological workup and a prolonged hospitalization course in our patient. Early recognition and tracheostomy should be advocated in such patients to prevent these complications.

References

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