Respiratory failure due to achalasia cardia

Hakim Azfar Ali a,*, Ganesan Murali a, Berjees Mukhtar b

a Division of Pulmonary and Critical Care, Albert Einstein Medical Center, Suite 331 Klein, 5401 Old York Road, Philadelphia, PA 19141, USA
b Department of Internal Medicine, Albert Einstein Medical Center, Philadelphia, PA 19141, USA

KEYWORDS
Achalasia;
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Aspiration;
Pneumonitis;
Trypanosoma;
Pseudoachalasia

Educational Aims:
- To review the therapy incidence of achalasia
- To familiarize the reader with symptoms and diagnosis of achalasia
- To look at treatment of pulmonary aspiration in the setting of achalasia

Introduction

A 76-year-old male nursing home resident was sent to our hospital for evaluation of hypoxia and 3-week history of cough with production of large volumes of whitish frothy sputum. The patient a lifelong non-smoker was noted to have resting oxygen saturation ranging from 85 to 92% for the last 1–2 weeks. He had received azithromycin for 5 days without any benefit. He denied any fever, hemoptysis, chest pain or dysphagia. He had lost about 7 pounds over 3 months and was reported to be a "slow eater". His past history included a stroke with no residual deficits, epilepsy that was well controlled and schizophrenia. His medications included topiramate, escitalopram, quetiapine, phenytoin and omeprazole.

On examination he was alert and oriented. He appeared comfortable at rest breathing at 20 per minute. Other vitals were normal. Frothy whitish sputum could be visualized in his oro-pharynx. His jugular venous pressure was normal. On auscultation, he had bilateral crackles at the lung bases. Bilateral pedal edema was noted. The remainder of his examination was unremarkable.

His laboratory values included a WBC count of 8000/mm³, hemoglobin of 12.7 g/dl, a normal basal metabolic panel, and an albumin of 3 g/dl. The initial chest radiograph revealed prominence of interstitial markings.

He was given a 40 mg of IV furosemide in the ER with no improvement. His oxygenation worsened requiring a FiO2 of 50% over next 24 h. A CT scan of the chest revealed diffuse dilatation of the esophagus containing an air-fluid level without apparent wall thickening or point of obstruction (Fig. 1). The lung parenchyma showed a micronodular pattern with alveolar filling at the bases consistent with aspiration. A contrasted upper gastro-intestinal (GI) series
was ordered (Fig. 2). The esophagus appeared dilated, and the column of barium tapered into a typical "birds beak" at the gastro-esophageal junction, with significantly delayed emptying. Multiple tertiary contractions were visualized. This was consistent with achalasia.

An upper gastro-intestinal endoscopy did not reveal mucosal abnormalities or endo-luminal lesions. 100 units of botulinum toxin were injected sequentially in 5 different locations. Over the next few days, the patients' diet was advanced, his oxygen requirements improved and he was discharged back to the nursing home with 2 l of oxygen and outpatient follow up.

Discussion

Achalasia is a primary esophageal motility disorder characterized by failure of esophageal peristalsis and lower esophageal sphincter (LES) relaxation due to damage to the myenteric plexus. Initially described in 1674 by Sir Thomas Williams as a disease it was first termed achalasia (Greek for "lack of relaxation") by Hurst in 1927. Achalasia is a relatively rare disease affecting the genders equally with a prevalence of less than 1/10,000 and an incidence between 0.03 and 1/100,000 per year. The peak incidence is seen in the 3rd and 7th decade of life.1

Achalasia can be classified as primary when a direct etiology cannot be identified or secondary when there is a well-defined cause like Chagas disease. In either type, the pathogenesis involves degeneration of the plexus myentericus resulting in a lack of inhibitory impulses needed for coordination of lower esophageal sphincter relaxation and esophageal peristalsis.2 This may have a genetic, autoimmune or infectious (viral) origin but the exact cause remains to be determined.

The usual clinical presentation is that of progressive dysphagia both for liquids and solids, regurgitation and chest pain. Symptoms can often be misinterpreted as gastro-esophageal reflex disease leading to delayed diagnosis typically by 2–3 years. Symptoms may be worse in recumbent posture. More subtle symptoms include an increase in the time spent in eating (upon questioning). In patients with recent dysphagia (<6 months), weight loss and age >50 years, pseudachalasia secondary to a neoplasm should be ruled out by endoscopy and a CT scan.

Clinical symptoms related to aspiration have been well recognized in patients with achalasia. There is a strong association between dysphagia, esophageal disorders (Table 1) and the development of complications of aspiration.3 This is most likely secondary to the change in volume and frequency and bacteriology of the secretions.

Several acute or chronic pulmonary syndromes (Table 2) may occur after aspiration, depending on the amount and nature of the aspirated material, the frequency of aspiration, and the host’s response to the aspirate. The most well recognized is chemical injury from the acid contents of the stomach leading to a lung injury pattern termed aspiration pneumonitis, usually self-limited.4

The aspirate may be colonized bacteria that can lead to aspiration pneumonia in the right setting. The recognition of the different syndromes associated with aspiration is important to prevent inappropriate antibiotic use. The distinction between a pneumonitis and pneumonia is primarily clinical. A pneumonia presents with persistent or worsening clinical and radiological picture beyond 48 h.

\[\text{Table 1} \quad \text{Esophageal disorders associated with pulmonary aspiration}\]

| 1. Achalasia cardia (primary or secondary) |
| 2. Scleroderma |
| 3. Esophageal/pharyngeal diverticula |
| 4. Esophageal neoplasia |
| 5. Gastro-esophageal reflux |
| 6. Hiatal hernia |
Table 2  Pulmonary aspiration syndromes

<table>
<thead>
<tr>
<th>Syndrome</th>
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<tbody>
<tr>
<td>Acute aspiration pneumonitis</td>
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<tr>
<td>Acute aspiration pneumonia</td>
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<tr>
<td>Chronic aspiration pneumonitis</td>
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<tr>
<td>Diffuse aspiration bronchioloitis</td>
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<td>Isolated bronchospasm</td>
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<td>Airway obstruction</td>
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<td>Lung abscess</td>
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<td>Exogenous lipid pneumonia</td>
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<tr>
<td>Chronic interstitial fibrosis</td>
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<tr>
<td>Atypical mycobacterial infections (M. fortuitum)</td>
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</tbody>
</table>

An elderly male with dyspnea, frothy sputum and hypoxia

after aspiration. The majority of reports describe dyspnea with patchy bilateral alveolar opacities.

Atypical mycobacteria particularly Mycobacterium fortuitum can cause disease in the setting of dysphagia.5

The mechanical effects of a large food bolus may lead to bronchial obstruction and atelectasis.

Any of the syndromes associated with aspiration may present with similar pulmonary symptoms including recurrent wheezing, apnea, chronic cough, bronchorrhea, hypoxemia and refractory asthma. Bronchorrhea is defined as the production of more than 100 ml per day of watery sputum. Bronchorrhea may occur in either bronchioalveolar or metastatic cancer growing in a bronchioalveolar pattern and has been reported with aspiration.

A long-term or recurrent aspiration can lead to chronic interstitial changes/fibrosis clinically represented as intermittent cough and waxing and waning pulmonary lesions. The term diffuse alveolar bronchiolitis (DAB) has been proposed to define a clinical entity that is characterized by a chronic inflammation of bronchioles caused by recurrent aspiration of foreign bodies.6 DAB was originally recognized in the elderly, but can occur in younger patients with achalasia or GERD with similar manifestations.

Esophageal manometry is the gold standard to diagnose achalasia. It classically reveals esophageal aperistalsis with a non-relaxing lower esophageal sphincter (LES) with pressures frequently above 8 mm Hg.7 Typical radiographic findings on a barium swallow study include the smooth tapering in the distal esophagus with the typical “bird’s beak” or “champagne glass” appearance proximal dilatation and lack of primary peristalsis noticed during fluoroscopy.

The treatment of pulmonary aspiration in the setting of achalasia includes management of the pulmonary issues and treatment of achalasia. Antibiotics are not routinely indicated in aspiration pneumonitis but are the mainstay of treatment in aspiration pneumonia. Empirical therapy with broad-spectrum agents is recommended. Anaerobic coverage is not routinely indicated. Sampling of the lower respiratory tract for culture may allow targeted antibiotic therapy and subsequent de-escalation. All cases need supportive care with oxygen and support of ventilation with decommodation. Preventive measures for further episodes of aspiration include treatment of the underlying condition and in the case of critically ill or nursing home patients elevation of the head of bed.

Medical therapy such as calcium channel blockers, nitrates and phosphodiesterase inhibitors aimed at reducing lower esophageal sphincter pressure is usually ineffective. Patients who are good surgical candidates should be offered either pneumatic balloon dilatation or laparoscopic myotomy. Long term (i.e. 5–10 years) clinical response rates to balloon dilatations range from 40 to 80%.8

Symptom improvement after myotomy ranges from 83 to 100% for the first year and 67–85% for sustained (i.e. >10 years) remission.9

In the only randomized trial comparing outcomes 95% of patients treated with surgical myotomy had a good long-term result compared with 65% of patients treated with pneumatic dilatation10 with lower risk of subsequent interventions.

If there are contraindications to surgery the patients can be offered endoscopic injection of botulinum toxin A (botox) in the lower esophageal sphincter which that acts by presynaptic cholinergic blockade. The best results have been noticed in patients with vigorous achalasia. Although the majority of patients notice important improvement after the first treatment, patients should be followed up regularly in order to identify disease progression early and avoid the development of the end-stage disease when esophagectomy may become the only treatment option.

CME Section

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Educational questions

Answer the following questions

1. Which of the following is not true for the incidence of achalasia?
   a. Usually appears only in the elderly (over age 70)
   b. Is equally likely to occur in men and women
   c. Occurs in about 1 in every 10,000 adults
   d. Usually has a gradual onset
2. The progressive symptoms of achalasia usually include:
   a. dysphagia for liquids then solids
   b. regurgitation
   c. chest discomfort
   d. all of the above
3. Achalasia is usually diagnosed:
   a. early in the course
   b. often misdiagnosed as reflux disease
   c. by weight loss
   d. by MRI examination
4. Aspiration is common and may be difficult to treat due to
   a. acid damaged to main stem bronchus
   b. aspiration pneumonitis
   c. changes in bacterial flora in secretions
   d. viral nature of most aspiration pneumonias

5. Treatment of achalasia is important to prevent respiratory damage and includes all of the following except:
   a. endoscopic surgery
   b. Botulism toxin injections
   c. repair of the myenteric plexus
   d. esophagectomy

References