Index of Suspicion

2 Aspiration Pneumonia in a 16-year-old Girl

Khanh Van Lai, MD,* Mini Mehra, MD,† Eliezer Nussbaum, MD*

Departments of Pediatric Pulmonology and Gastroenterology,† Miller Children's Hospital, University of California, Irvine School of Medicine, Long Beach, CA.*

AUTHOR DISCLOSURE Drs Lai, Mehra, and Nussbaum have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

PRESENTATION

A 16-year-old girl with a history of asthma and anemia presents to the emergency department complaining of left-sided abdominal pain and multiple episodes of emesis for 3 days as well as fever for 1 day. She is unable to keep down both liquids and solids. This morning, she has been breathing very fast and appears pale. Past medical history includes pneumonia 3 years ago and mild intermittent asthma for 3 years treated with inhaled albuterol twice weekly. A similar episode of abdominal pain resolved without treatment 6 months ago. She reports that for the past year, she sometimes feels like she "can't breathe" when swallowing and sometimes has had difficulty swallowing solids.

On physical examination, her temperature is 41.1°C (105.9°F), heart rate is 176 beats per minute, respiratory rate is 40 breaths per minute, blood pressure is 74/54 mm Hg, and oxygen saturation is 59% on room air. She appears pale, restless, and in severe respiratory distress. Arterial blood gas shows:

- pH 7.33
- P_{CO_2} 33 mm Hg (4.4 kPa)
- P_{O_2} 55 mm Hg (7.3 kPa)
- Bicarbonate 17 mEq/L (17 mmol/L)

Following endotracheal intubation, suctioning yields copious thick yellow secretions.

Additional laboratory results include:

- White blood cell count 32,200/μL (32.2 × 10^9/L) (7% neutrophils, 77% bands)
- Hemoglobin 10.4 g/dL (104 g/L)
- Platelet count, 544 × 10^3/μL (544 × 10^9/L)
- C-reactive protein 309 mg/L (2,942.9 nmol/L)

A chest radiograph (CXR) shows bilateral pulmonary infiltrates and a widened mediastinum (Fig 1). The girl is admitted to the ICU for acute respiratory failure suspected to have resulted from aspiration pneumonia.

Respiratory tracheal culture grows *Escherichia coli*. A follow-up CXR reveals an air bubble in the medial right chest (Fig 2) and clinicians place a chest tube for a pneumothorax. When the pneumothorax does not improve, clinicians order computed tomography (CT) scan of the chest (Fig 3), which reveals the diagnosis.

DISCUSSION

Chest CT scan showed moderately severe esophageal distension, bronchiectasis, severe tracheal compression, and bilateral basilar consolidations. Barium swallow

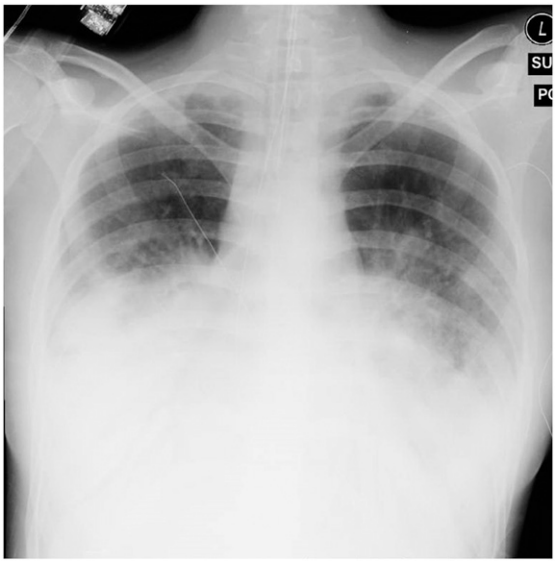

Figure 1. Chest radiograph shows bilateral pulmonary infiltrates and a widened mediastinum.

demonstrated dilation of the entire esophagus, no esophageal peristalsis, and one episode of aspiration, consistent with esophageal achalasia causing acute or chronic aspiration pneumonitis.

Pathogenesis

Primary achalasia is an esophageal neurodegenerative motility disorder characterized by failure of the lower esophageal sphincter (LES) to relax. In adults, neural inflammation results in selective destruction of the inhibitory neurons in the myenteric plexus that innervate the LES and distal esophagus. In children, neural inflammation is rarely present, but there is markedly reduced to complete aganglionosis of the myenteric ganglion in the high-pressure zone

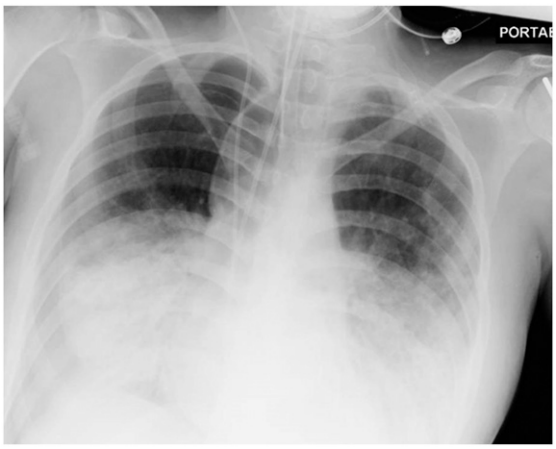

Figure 2. Chest radiograph shows an air bubble in the medial right chest.

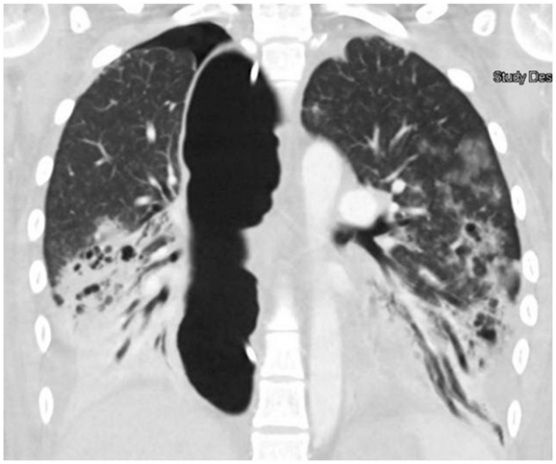

Figure 3. Chest computed tomography scan shows esophageal distension, bronchiectasis, tracheal compression, and bilateral basilar consolidations.

of the LES. Following loss of postganglionic inhibitory neurons, the inhibitory transmitters nitric oxide and vasointestinal peptide are reduced, creating an imbalance between excitatory and inhibitory neurons that prevent LES relaxation.

Epidemiology

The incidence of childhood achalasia is 1.1 in 1,000,000. Although fewer than 5% of patients who have achalasia present before age 15 years, pediatric primary achalasia is more likely to be syndromic than isolated. Two percent of patients with trisomy 21 develop achalasia. Achalasia has been associated with congenital hypoventilation syndrome, glucocorticoid insufficiency, eosinophilic esophagitis, familial dysautonomia, Chagas disease, and autosomal recessive mutations on chromosome 12q13 (achalasia, adrenal insufficiency, and alacrima [AAA] syndrome).

Clinical Presentation

Children with achalasia have characteristic progressive dysphagia, first to solids and then to liquids. Symptoms include vomiting, food regurgitation, heartburn, and retrosternal chest pain after meals or while lying down. With chronic food retention, mechanical and chemical esophagitis ensues. Children develop food avoidance behaviors and weight loss. Younger children and infants may present with feeding difficulties and respiratory symptoms such as chronic or nocturnal cough, recurrent pneumonia, aspiration, and hoarseness. Despite these classic findings, the diagnosis of achalasia is delayed by a mean of 5 years from the onset of symptoms. Common misdiagnoses include gastroesophageal reflux, failure to thrive, eating disorders, eosinophilic esophagitis, and asthma.

Diagnostic Steps

A CXR can distinguish underlying achalasia from asthma by the presence of a widened mediastinum, which reflects esophageal dilation, and an absent gastric air bubble. In contrast, in acute asthma, the cardiac silhouette is notably slender due to air trapping.

In two-thirds of patients with achalasia, a barium swallow demonstrates esophageal dilation, narrowed gastroesophageal junction (GEJ) with a "bird-beak" appearance, esophageal aperistalsis, and delayed esophageal emptying.

Upper endoscopy is often performed to rule out secondary achalasia or "pseudoachalasia," which is a narrowing of the GEJ caused by other processes such as carcinomas.

The gold standard of diagnosis is conventional or high-resolution manometry. Typical findings include aperistalsis in the distal two-thirds of the esophagus, incomplete or absent LES relaxation after swallow, and elevated resting LES pressure. High-resolution manometry allows classification of subtypes (I, II, or III) based on the patterns of esophageal pressurization. In type I (classic) achalasia, swallowing results in no substantial change in esophageal pressures. With type II achalasia, swallowing leads to simultaneous pressurization spanning the length of the esophagus. In type III (spastic) achalasia, swallowing results in abnormal esophageal contractions or spasms. The ability to subtype achalasia is prognostically helpful because patients with type II achalasia have the most favorable response to pneumatic dilation, Heller myotomy, or botulinum toxin treatment. However, chest CT scan is essential in elucidating the severity of pulmonary and upper airway complications.

Treatment

For most children, the first-line treatment is laparoscopic or open Heller myotomy, with posterior fundoplication associated with a 90% success rate and low complication rate. For those unable to undergo surgery, endoscopic pneumatic dilation is associated with 40% to 85% success and complications such as esophageal perforation and LES rupture. Nonsurgical therapies such as long-acting nitrates, calcium channel blockers, and botulinum toxin show short-term efficacy but lack long-term benefits.

Prognosis

Complications of achalasia include aspiration pneumonia and megaesophagus, which can cause mechanical airway obstruction and require esophagectomy. Even after corrective treatment, patients have a 16 to 50 times greater risk of developing both squamous cell carcinoma and adenocarcinoma of the esophagus due to chronic bacterial overgrowth, inflammation, and posttreatment reflux. Despite the increased cancer risk, patients with achalasia have the same life expectancy as the general population.

Clinical Course

Following a laparoscopic Heller myotomy with partial fundoplication, this patient experienced dramatic improvements in symptoms. Due to her aspiration history, preoperative high-resolution manometry was considered too risky. Postoperatively, her diet was slowly advanced and she gained weight. Her baseline pulmonary function tests showed very severe obstruction (which did not improve after albuterol treatment), severe restrictive lung disease, and decreased alveolar capillary diffusion deficit. She was discharged from the hospital with lansoprazole, erythromycin ethylsuccinate (for postoperative gastroesophageal reflux disease and delayed gastric emptying), ferrous sulfate, and beclomethasone dipropionate inhaler. One month later, she remained symptom-free, and follow-up spirometry showed modest improvements.

Lessons for the Clinician

- Achalasia should be included in the differential diagnosis of the patient who has a history of dysphagia, difficulty breathing with swallowing, or suspected aspiration.
- Achalasia is differentiated from gastroesophageal reflux by its universal hallmark of dysphagia.
- Clinicians should question the diagnosis of asthma if symptoms do not improve with bronchodilators or corticosteroids and/or baseline spirometry results are normal.
- When chest radiography findings do not correlate with a history of positional dyspnea, clinicians should strongly consider additional evaluation, such as computed tomography scan of the chest.

Suggested Readings for this article are at http://pedsinreview.aappublications.org/content/37/10/442.

Parent Resources from the AAP at HealthyChildren.org

- **Pneumonia:** https://www.healthychildren.org/English/health-issues/conditions/chest-lungs/Pages/Pneumonia.aspx.

Case 2: Aspiration Pneumonia in a 16-year-old Girl

Khanh Van Lai, Mini Mehra and Eliezer Nussbaum

Pediatrics in Review 2016;37;442
DOI: 10.1542/pir.2016-0024

Case 2: Aspiration Pneumonia in a 16-year-old Girl

Khanh Van Lai, Mini Mehra and Eliezer Nussbaum

Pediatrics in Review 2016;37;442
DOI: 10.1542/pir.2016-0024