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Permalink

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Journal

Chest, 148(4)

ISSN

0012-3692

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Publication Date

2015-10-01

DOI

10.1378/chest.2279880

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Peer reviewed

Achalasia Masking as Mild Intermittent Asthma: Acute Respiratory Failure, Aspiration Pneumonia, and Pneumothorax in a Teenage Girl

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Chest. 2015;148(4_MeetingAbstracts):776A. doi:10.1378/chest.2279880

Abstract

SESSION TITLE: Pediatrics and Congenital Cases

SESSION TYPE: Affiliate Case Report Slide

PRESENTED ON: Tuesday, October 27, 2015 at 07:30 AM - 08:30 AM

INTRODUCTION: Primary achalasia is a rare esophageal motility disorder caused by failure of the lower esophageal sphincter to relax. Childhood achalasia is often misdiagnosed as gastroesophageal reflux, failure to thrive, eating disorder or asthma.

CASE PRESENTATION: A 16-year-old girl presented with three days of vomiting, abdominal pain, and fever. She was diagnosed 3 years earlier with asthma and pneumonia, and had similar abdominal pain 6 months ago. For the past year, she sometimes felt short of breath swallowing solids. Initial vital signs were T 105.9°F, HR 176 bpm, RR 40/min, BP 74/54 mmHg, and O₂ saturation 59%. After intubation for severe respiratory distress, copious thick yellow tracheal secretions grew *E. coli*. Labs showed leukocytosis, bandemia and elevated inflammatory markers. CXR showed bilateral infiltrates and widened mediastinum (Image 1). A follow-up CXR revealed a medial right air bubble, and a chest tube was placed. As the air bubble did not resolve, CT chest obtained showed severe esophageal distension, tracheal compression, basilar consolidations, and bronchiectasis (Image 2). She was diagnosed with achalasia with aspiration pneumonitis when a barium swallow demonstrated complete esophageal dilatation and absent peristalsis. After laparoscopic Heller's myotomy with partial fundoplication, she improved dramatically and went home with anti-reflux medicines and inhaled corticosteroids.

DISCUSSION: With an incidence of 1.6/100,000, childhood achalasia presents with progressive dysphagia first to solids and then liquids. Diagnosis is delayed on average by five years. Older children may complain of reflux, sore throat, hoarseness, choking, or dyspnea, and develop food avoidance behaviors. Younger children and infants may have feeding problems and respiratory symptoms such as chronic or nocturnal cough, recurrent respiratory infections, wheezing, stridor, or acute upper airway obstruction. Preferred treatment is laparoscopic Heller's myotomy. Lung complications from chronic aspiration include obliterative bronchiolitis, abscess, empyema and fibrosis.

CONCLUSIONS: Childhood achalasia should be on the differential when suspecting asthma or gastroesophageal reflux. Careful history taking is paramount. Mediastinal widening is present in achalasia but not asthma. Achalasia often results in lung injury due to chronic aspiration.

Reference #1: Franklin AL, et al. Childhood achalasia: A comprehensive review of disease, diagnosis and therapeutic management. *World J Gastrointest Endosc.* 2014 Apr 16;6(4):105-11.

DISCLOSURE: The following authors have nothing to disclose: Khanh Lai, Mini Mehra, Eliezer Nussbaum

No Product/Research Disclosure Information