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16-year-old female with difficult to control asthma

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Image of the month submission

Title: 16-year-old female with difficult to control asthma

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Question: A 16-year-old female, otherwise healthy, was seen by a respirologist for chronic intermittent wheeze and shortness of breath. She had been previously treated unsuccessfully with puffers for this wheeze. Pulmonary function tests were abnormal, indicative of an obstructive process, and the respirologist was concerned about bronchiectasis and/or difficult to control asthma. A chest x-ray (CXR) was done and the patient was referred to Pediatric Gastroenterology. On further questioning, she described a 9-year history of dysphagia. It had recently worsened, and she was now choking on food and subsequently vomiting three times per week.

On examination, the patient was hemodynamically stable. Her abdomen was soft, non-tender and non-distended. Oropharynx exam and respiratory exams were normal that day. Laboratory tests were unremarkable (normal complete blood count, electrolytes, creatinine, liver panel, and inflammatory markers). Review of the CXR suggested abnormalities of the esophagus. A computed tomography scan of the chest and abdomen was completed (to better assess for bronchiectasis).

What is the diagnosis?

Answer: The CT scan indicated the presence of a severely dilated esophagus, from the cervical segment cranially to the gastroesophageal junction caudally, with maximal measurements of 57 (transverse) x 46 (anteroposterior AP) mm wide. The esophagus was filled with fluid and particles (image 1). There was no evidence of an esophageal mass or external compression on the esophagus. These findings are consistent with achalasia. There was secondary compression of the patient's central airways. Her trachea and right main bronchus were significantly compressed (with complete effacement of the bronchus on expiration) (red arrow, image 2). Further investigations included an upper GI series, which showed the typical 'bird's beak' appearance at the distal esophagus, an upper endoscopy (which was normal), and manometry studies (which were inconclusive). She was referred to adult gastroenterology for a per-oral endoscopic myotomy (POEM) procedure.

Achalasia is usually diagnosed between 25-60 years old, and has an annual incidence rate in children of only 0.11 per 100,000 (1,2). To our knowledge, this is only the second reported case of respiratory distress secondary to severe esophageal dilation in a pediatric patient, as these findings are generally reserved for the adult population (3). Further, achalasia usually presents with dysphagia for solids and liquids, weight loss, regurgitation and heartburn. While this patient did experience dysphagia, it is rare for a patient to have dyspnea as the symptom that brings them to medical attention (3).

This case emphasizes the importance of a broad differential diagnosis, including gastrointestinal conditions, for children presenting with dyspnea, wheeze and stridor. This is particularly appropriate in the setting of difficult to manage asthma. Further, a content-filled esophagus can be confused by radiologists for a consolidation or collapsed lung on CXR, and therefore a thorough history, including gastrointestinal symptoms, helps narrow the differential.

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