

## CLINICAL VIGNETTE

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# An Unexpected Cause of Dysphagia and the Role of Supplemental Imaging in Achalasia

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Sai S. Kambampati, Andrew Su, MD, Adarsh M. Thaker, MD and Alireza Sedarat, MD

### Case

A 23-year-old male presented with acute on chronic dysphagia. Five years endoscopy and biopsy showed eosinophilic esophagitis (EoE). He had mild improvement with oral viscous budesonide for six months and was subsequently managed with diet modifications alone. One month prior to presentation he developed worsening dysphagia, inability to tolerate solids, and accelerated weight loss. Endoscopy showed a dilated, food-filled esophagus and a tight lower esophageal sphincter (LES) with endoscopic appearance consistent with achalasia. No mucosal biopsies were taken. Subsequent high-resolution esophageal manometry showed lower esophageal resting pressure of 24.6 mmHg (nl 4.8-32mmHg), median integrated LES relaxation pressure of 20.0 mmHg (nl <15 mmHg) with panesophageal pressurization in >90% of swallows (nl <20%). This pattern was interpreted as possibly consistent with type II achalasia.

He was referred to our center for treatment of achalasia and considered for peroral endoscopic myotomy (POEM). Given the atypical history, repeat endoscopy with endoscopic ultrasound (EUS) was performed. The esophagus was severely dilated with copious amounts of retained liquid and solid food. There were distinct thick concentric rings (Figure A) and punctate white adherent spots. The LES was severely spastic (Figure B). Endoscopic ultrasound (EUS) revealed unremarkable esophageal wall thickness with normal mucosal, submucosal, and muscular layers of the esophagus. Surprisingly, bulky abdominal and mediastinal lymphadenopathy (Figures C and D) were seen without clear compression of the LES. Proximal and distal biopsies of the esophagus and fine needle aspiration (FNA) of the lymph nodes were performed. Subsequent computed tomography (CT) of the chest, abdomen, and pelvis confirmed diffuse bulky lymphadenopathy (Figure E).

Esophageal biopsies showed >40-50 eosinophils per high power field in the esophageal body, consistent with a diagnosis of ongoing EoE. Lymph node aspiration revealed Hodgkin's Lymphoma. While EoE has been reported to produce manometric features similar to achalasia, was felt coincidental given his long-standing EoE was relatively well controlled until recently.<sup>1</sup> His symptoms were attributed to a new diagnosis of secondary achalasia related to a paraneoplastic process in the setting of Hodgkin's Lymphoma.

### Discussion

Achalasia was first mentioned in 1674 when treatment was esophageal dilation with a dilator made of whalebone. The term was not officially coined until the early 1900s when it was recognized as an esophageal motor disorder involving impaired relaxation of the LES. While the pathophysiology is now understood to be related to a loss of inhibitory neurons in the myenteric plexus, particularly nitric oxide, the exact etiology remains unclear. The majority of cases are primary, or idiopathic, achalasia.

In evaluating cases of achalasia, it is important to rule out causes other than idiopathic, primary achalasia. Pseudoachalasia, also referred to as secondary achalasia, is rare but can account for up to 5% of patients presenting with manometric features of achalasia.<sup>2</sup> Over 60% of non-primary achalasia is secondary to malignancy, while the remainder can be secondary to other causes such as Chagas disease, benign tumors (e.g. leiomyomatosis), postoperative complications, and diseases of the central nervous system.<sup>3</sup> The most common malignancies associated with achalasia are carcinomas of the esophagus and proximal stomach, often from direct tumor involvement of the LES or gastric cardia. Other reported malignancies include non-small cell lung cancer, breast cancer, prostate cancer, and pancreatic cancer.<sup>4</sup> When there is no direct invasion or compression of the gastroesophageal junction by a tumor, a paraneoplastic syndrome is suspected and may be antibody mediated (anti-Hu or anti-Nu Ab).

Cases of achalasia secondary to lymphoma, and specifically Hodgkin's lymphoma, are exceedingly rare and mostly limited to a few case reports making the true incidence hard to estimate.<sup>5,6</sup> Both direct tumor infiltration and a paraneoplastic syndrome have been reported. The latter is suspected in our patient due to the lack of physical compression or infiltration on EUS. Although the terminology is not standardized, may be more meaningful to reserve the term pseudoachalasia for direct tumor involvement of the gastroesophageal junction and paraneoplastic-mediated achalasia when a humoral factor is suspected or confirmed.

The evaluation of patients with suspected achalasia generally consists of an esophagram, upper endoscopy, and manometry. Manometric findings in primary versus secondary achalasia can be identical. The diagnosis of primary achalasia is made when secondary causes have been sufficiently ruled out. In one study,

evaluation with EUS and subsequent findings changed management in up to 15% of patients with esophageal motor disorders on manometry, including achalasia.<sup>7</sup> An especially high suspicion of malignancy-associated pseudoachalasia is required in patients with advanced age, short duration of symptoms (less than one year), and/or severe weight loss (out of proportion to the presentation).<sup>8</sup> These features should prompt additional evaluation such as cross-sectional imaging and/or EUS.

**Figures**

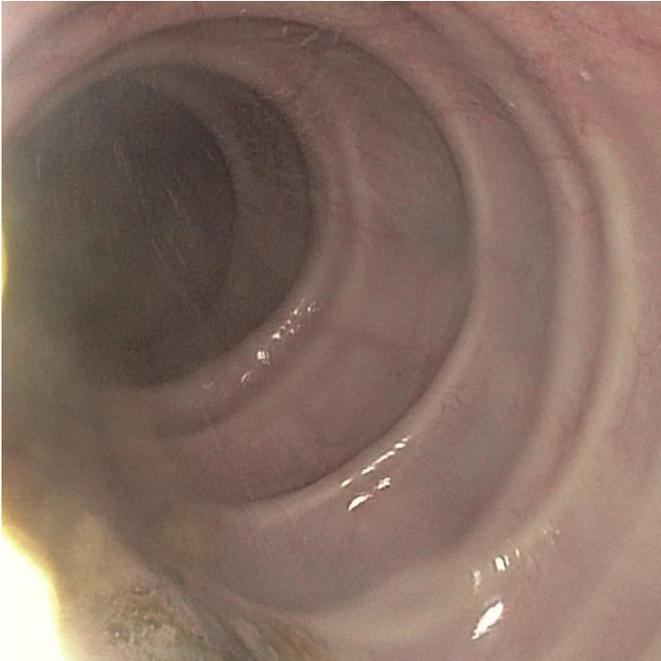


Figure A



Figure B



Figure C

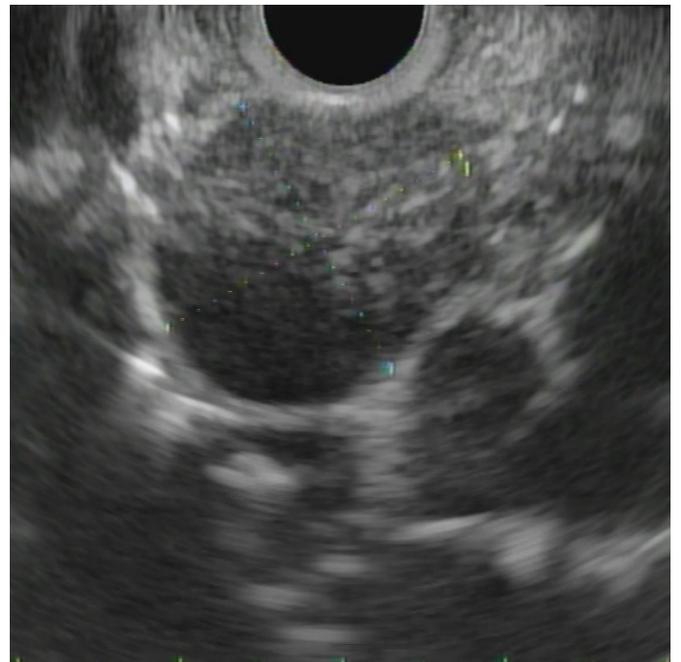


Figure D

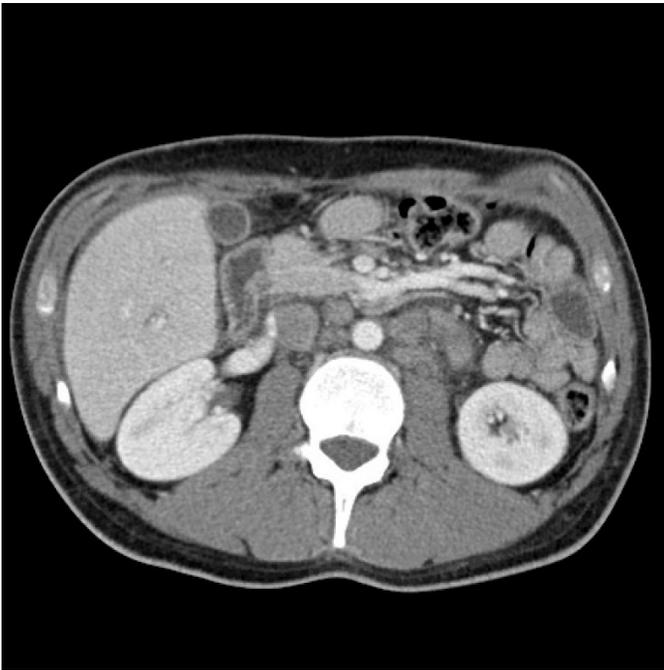


Figure E

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Submitted April 24, 2019