

Boerhaave's Syndrome as a First Presentation of Eosinophilic Esophagitis

Jackson, Whitney E.¹; Mehendiratta, Vaibhav²; Palazzo, Juan P.³; DiMarino, Anthony J.²; Quirk, Daniel M.²; Cohen, Sidney²

¹Department of Internal Medicine, ²Department of Gastroenterology and Hepatology, ³Department of Pathology, Thomas Jefferson University Hospital, Philadelphia, PA, United States

BACKGROUND:

Prior studies have reported esophageal rupture following endoscopy or bolus impaction in eosinophilic esophagitis.

AIM:

The purpose of this study is to examine the frequency and characteristics of spontaneous rupture (Boerhaave's Syndrome) associated with vomiting in eosinophilic esophagitis.

METHODS:

- A retrospective search of inpatient and outpatient records was conducted for the diagnoses "Boerhaave's", "eosinophilic esophagitis", and "esophageal rupture" from January 2001 to January 2011 within the gastroenterology division at an urban tertiary care hospital.
- For each subject identified, medical records, endoscopy reports, biopsy reports and radiographic studies were reviewed.
- A faculty member of the Department of Pathology blindly reviewed all esophageal biopsy specimens.
- Eosinophilic esophagitis was defined as 15 or more eosinophils (EOS) in at least 2 high-power fields (HPFs) or 25 or more eosinophils in any single high power field.

RESULTS:

- Over a period of ten years, 447 patients were identified with a diagnosis of eosinophilic esophagitis.
- Of these, four patients presented with spontaneous esophageal rupture in the setting of eosinophilic esophagitis in the absence of food impaction or endoscopy (4/447, less than 1%).
- None of the patients had an established diagnosis of eosinophilic esophagitis prior to presentation. All four cases presented with a triad of vomiting, chest pain and pneumomediastinum.
- Three of the four patients were male (75%), and ages ranged from 22 to 56 (mean 37) years-old.
- In two of the four patients, water-soluble contrast extravasation was seen on imaging prompting surgical intervention (50%); one of these patients required esophageal resection.
- The other two patients demonstrated no contrast extravasation. These two patients were observed for resolution.
- This represents a unique opportunity to examine full thickness surgical specimen showed invasion of eosinophils into the muscularis propria. Intraepithelial eosinophil infiltration was seen on all mucosal biopsies (>30 EOS/HPF) with significant improvement after steroid (topical or systemic) treatment.

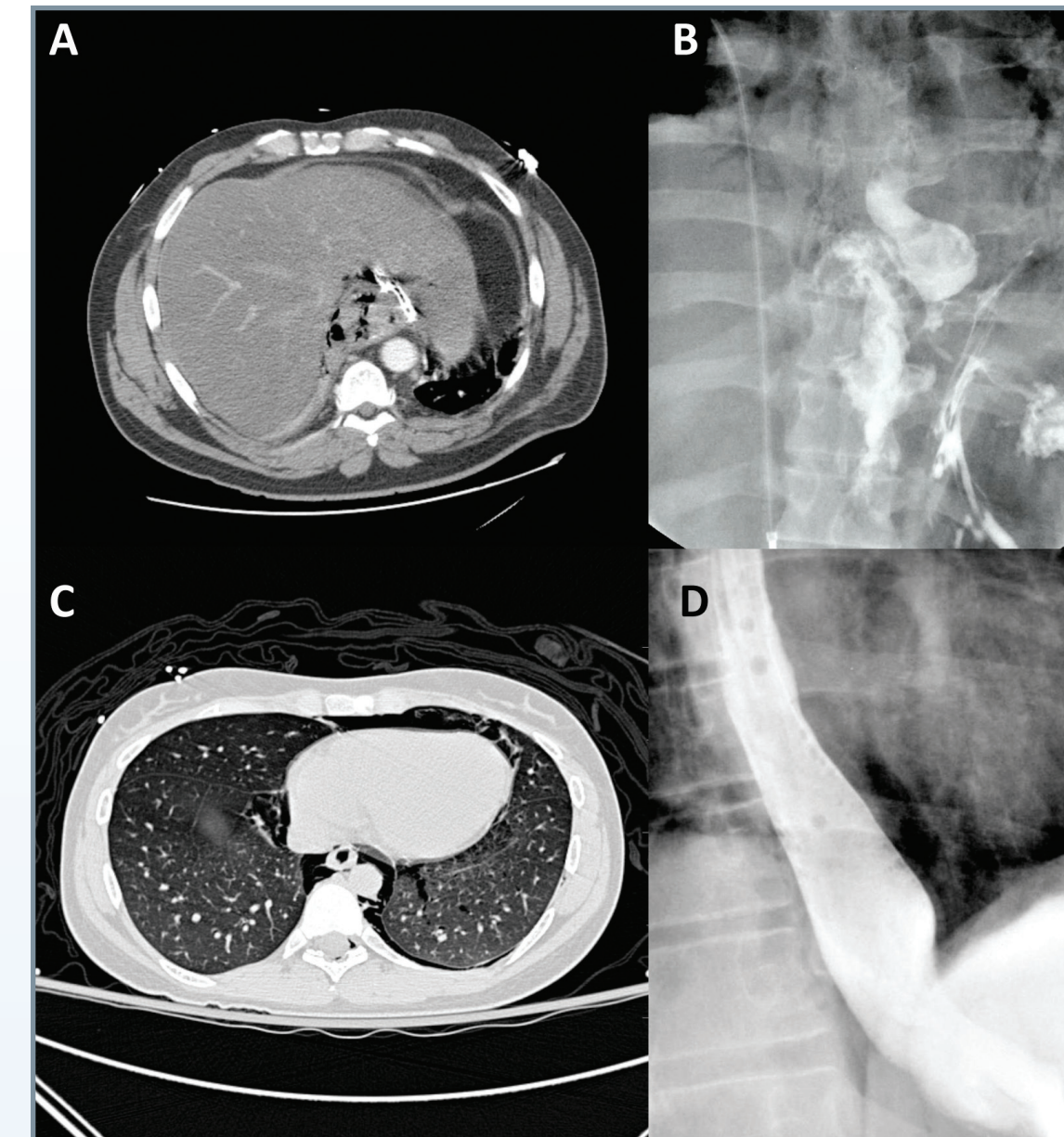


Figure 1: Patient 2: (A.) CT Scan. Mediastinal air and extravasated contrast are present below the level of the carina compatible with rupture of the distal esophagus. Air extends into the gastro-hepatic space. (B.) Water-soluble contrast esophagram. Contrast leaks behind the stomach as well as from the right aspect of the esophagus consistent with perforation. **Patient 3:** (C.) CT Scan. Pneumomediastinum is present as well as air dissecting into the pericardium, but no frank defect is visualized in the trachea or esophagus. (D.) Barium esophagram following initial evaluation with water-soluble contrast esophagram. There is no evidence of esophageal leak and no contrast extravasation.

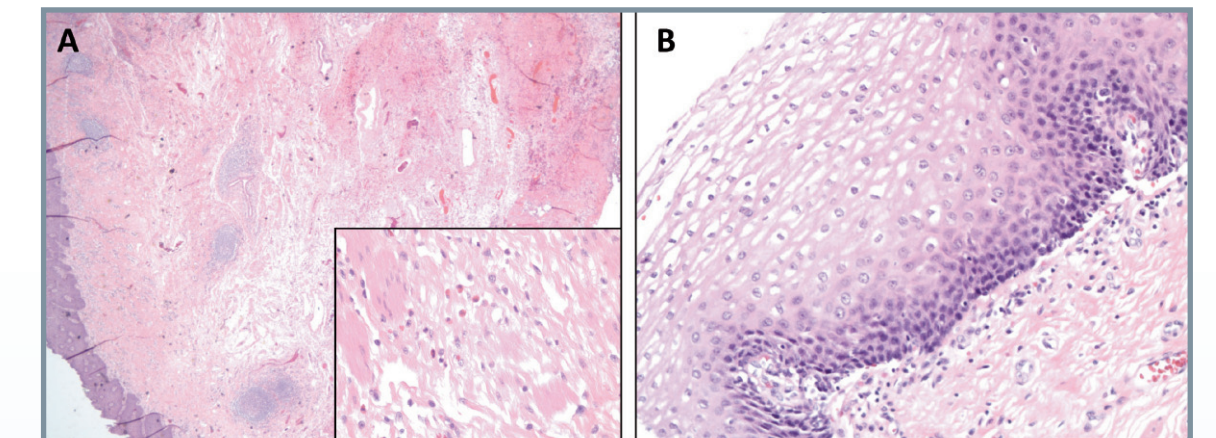


Figure 2: (A.) Transmural esophageal biopsy at time of presentation of Patient 2 demonstrating eosinophilic infiltration into the muscularis propria. Approximately 40 eosinophils per high power field (EOS/HPF). (H and E stain, 10x magnification). **INSERT.** Evidence of eosinophils infiltrating the muscularis propria of the esophagus. (H and E stain, 40x magnification). (B.) Esophageal mucosa after treating Patient 2 with steroids showing no evidence of eosinophils. (H and E stain, 40x magnification).

CONCLUSION:

- Spontaneous esophageal rupture is a rare (less than 1%) but critical presentation of eosinophilic esophagitis manifesting with vomiting, chest pain and pneumomediastinum.
- Surgery is required if extravasation is seen with water-soluble contrast.
- We suggest that eosinophilic esophagitis is a transmural disease rather than a simple mucosal process, thus making the esophageal wall susceptible to rupture with endoscopy, bolus impaction and now spontaneously (Boerhaave's Syndrome).