

Spontaneous Intramural Esophageal Rupture: An Uncommon Presentation of Eosinophilic Esophagitis Requiring Endoscopic Clipping

Tooba Tariq, MD¹, Asad Rizvi, MD², Karun Badwal, DO¹, and Ransome Eke, MD, PhD³

¹Department of Internal Medicine, Western Michigan University, MD Homer Stryker School of Medicine, Kalamazoo, MI

²College of Osteopathic Medicine, Michigan State University, East Lansing, MI

³Division of Epidemiology and Biostatistics, Western Michigan University, MD Homer Stryker School of Medicine, Kalamazoo, MI

ABSTRACT

Spontaneous intramural esophageal rupture (SIER) is a form of acute esophageal trauma defined as an injury deeper than a Mallory-Weiss tear but not extending completely through the muscular propria as in Boerhaave syndrome. SIER is a rare complication of eosinophilic esophagitis (EoE); after extensive literature review, we found 7 case reports of SIER complicating EoE. We present a case of SIER complicating EoE in a 46-year-old man with an atypical presentation requiring endoscopic clipping to achieve successful hemostasis.

INTRODUCTION

Eosinophilic esophagitis (EoE) is a chronic, immune-mediated disease characterized clinically by esophageal dysfunction manifesting as dysphagia and food impaction. The diagnosis is confirmed histologically by evidence of eosinophilic infiltration of the esophageal wall. Complications of EoE, including esophageal strictures, rupture, and tears, can occur due to increased friability of the esophageal mucosa as a consequence of chronic inflammation.^{1,2}

Spontaneous intramural esophageal rupture (SIER) is a rare form of acute esophageal trauma, defined as an injury deeper than Mallory-Weiss syndrome but not extending completely through the muscularis propria as in Boerhaave syndrome.^{3,4} SIER usually presents as chest/epigastric pain and hematemesis in elderly females, especially those who are coagulopathic⁵. Iatrogenic rupture is relatively common, but to our knowledge there are only 7 reported cases of SIER in the setting of EoE.^{2,3,6-10}

CASE REPORT

A 46-year-old man presented with a 1-day history of massive hematemesis (several washbasins full of bright red blood in 24 hours) and lightheadedness, followed by a syncopal episode prompting him to come to the emergency department. He reported intermittent symptoms of gastroesophageal reflux in the past, which were neither evaluated nor appropriately treated (he used cimetidine as needed). He had no history of food allergies, asthma, allergic rhinitis, or family history of atopic diseases. He reported taking ibuprofen 800 mg and aspirin 75 mg twice weekly.

The sequence of events started after he experienced an episode of pill-induced dysphagia followed by vomiting. During the hospitalization, the patient remained tachycardic with normal blood pressure. He was started on proton-pump inhibitor (PPI) infusion, and serial hemoglobin levels were obtained. He never complained of chest/epigastric pain.

ACG Case Rep J 2018;5:e66. doi:10.14309/crj.2018.66. Published online: September 26, 2018.

Correspondence: Asad Rizvi, MD, College of Osteopathic Medicine, Michigan State University, East Lansing, MI, United States 48825 (sasadrizvi@gmail.com).



Copyright: © 2018 Tariq et al. This work is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view a copy of this license, visit <http://creativecommons.org/licenses/by-nc-nd/4.0>.

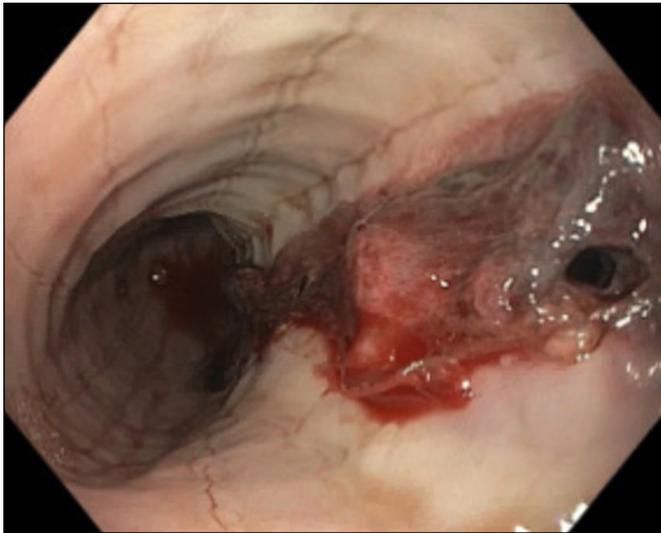


Figure 1. Esophagogastroduodenoscopy (EGD) showing a 15-cm non-circumferential, deep submucosal tear from mid- to distal esophagus with 3 spots of active arterial bleeding.

A chest radiograph on presentation revealed no evidence of perforation or presence of air in the mediastinum. An urgent esophagogastroduodenoscopy (EGD) demonstrated a 15-cm long, non-circumferential, deep submucosal tear (based on subjective assessment by the gastroenterologist) from mid- to distal esophagus with at least 3 spots of active arterial bleeding (Figure 1). Eleven endoscopic clips were placed, including 2 over-the-scope clips (Ovesco Endoscopy, Tübingen, Germany) (Figure 2).

A computed tomography scan of the chest with water-soluble contrast was suspicious for a small pneumomediastinum and extraluminal contrast. Cardiothoracic surgery was consulted,

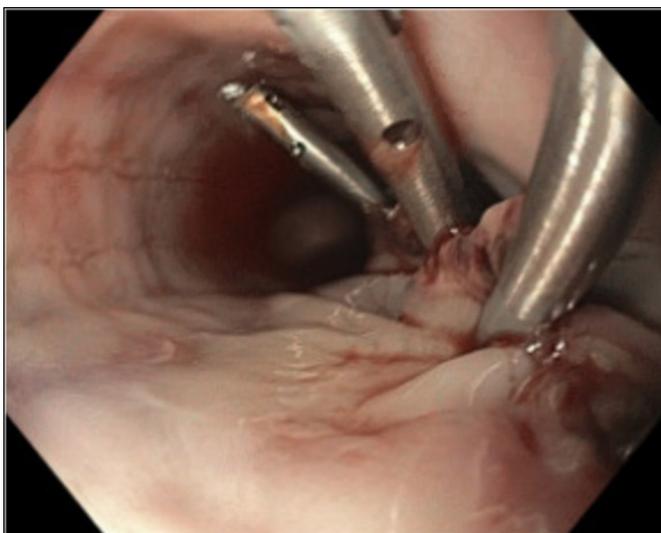


Figure 2. Endoscopic clips were placed to control bleeding.

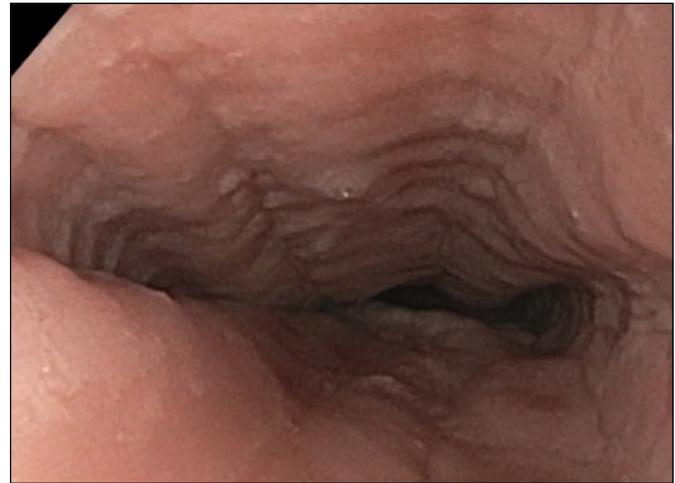


Figure 3. Repeat EGD 8 weeks later showing an almost completely epithelialized laceration with esophageal linear furrowing.

and a barium esophagram was negative for perforation. Treatment with intravenous antibiotics was initiated and PPI was continued, while the patient was maintained on bowel rest. The outcome was satisfactory, and the patient did not develop any symptoms or signs of perforation. A repeat esophagram on day 5 again showed no evidence of perforation. His diet was advanced as tolerated, and he was advised to take PPI 40 mg daily for 8 weeks.

Two months later, a repeat endoscopy showed an almost completely epithelialized laceration, although linear furrows were noted (Figure 3). Biopsies revealed extensive mucosal eosinophilic infiltration (focally >50 /high-powered field) in the proximal and distal esophagus. A diagnosis of EoE was made, and treatment with topical fluticasone propionate was initiated. The patient was scheduled for a follow-up EGD in a year.

DISCUSSION

EoE typically affects young to middle-aged Caucasian men with history of an allergic disorder.¹¹ Complications related to EoE include strictures, food impaction, and secondary gastroesophageal reflux disease, but tears, lacerations, and perforations are rarely reported.¹² Mucosal tears or lacerations during endoscopic dilation of narrowed segments are suggestive of increased mucosal fragility from chronic inflammation and eosinophilic infiltration, with subsequent remodeling described as “crepe paper sign.”^{1,2,8,10} Our patient had no history of atopy or episodes of dysphagia/food impaction, which commonly occur in EoE. However, he had chronic, intermittent symptoms of heart burn, which is the predominant symptom (75%) in EoE.¹³

First described in 1960s, SIER is the least common form of esophageal injury with roughly 50 reported cases^{3,6} It can be

spontaneous, traumatic, emetogenic, iatrogenic, or related to aortic disease and bleeding disorder.¹⁴ Patients frequently describe retrosternal pain (most common presenting symptom in 80–85% of cases), dysphagia/odynophagia, and hematemesis. However, this triad is observed in approximately 35% of the patients.

SIER does not lead to complete perforation of the esophageal wall as in Boerhaave syndrome, but it is more extensive than in Mallory-Weiss syndrome. Nevertheless, the pathogenesis involving a sudden increase in intra-abdominal pressure due to coughing/vomiting remains common to all three patterns of esophageal injury.

The sequence of events in our patient were sudden onset of dysphagia, which led to an episode of vomiting and ultimately resulted in frank hematemesis. Hence, the deep long tears in the esophageal wall were due to a combination of factors including a rapid rise in intraluminal pressure as well as increased mucosal friability from ongoing inflammation secondary to EoE.

In cases of SIER, chest radiographs are usually normal with no significant abnormalities. Although the definite diagnosis of SIER is made with EGD, it is not usually recommended as an initial test due to the risk of perforation. Instead, the preferred early investigative modality is a computed tomography scan with water-soluble contrast, which not only excludes perforation, but can also lead to the diagnosis of rupture by demonstration of the characteristic “double-barreled esophagus” sign.^{2,4} Boerhaave syndrome is considered a surgical emergency, whereas conservative treatment is usually sufficient for SIER.^{3,14,15} In patients presenting with frank bleeding, endoscopic exploration should be implemented because it is diagnostic and therapeutic.³

This case highlights a very rare and scarcely documented complication of EoE. Furthermore, this report also describes an atypical presentation of SIER due to absence of chest/epigastric pain (reported in >80% patients). While nonoperative management is sufficient in most cases, our patient required endoscopic intervention to achieve hemostasis. This intervention is reported in only 3 previous cases of SIER complicating EoE.^{2,3} Other successful treatment options include metal stent placement for circumferential tears.¹⁶ Surgical management including esophagectomy should be considered if there is a perforation resulting from circumferential mucosal dissection and Boerhaave's syndrome associated with EoE.^{10,17} Furthermore, in case of non-resolution of an intramural esophageal hematoma, endoscopic incision of the mucosal bridge has been described.¹⁸ SIER should be considered in EoE patients presenting with dysphagia, chest pain, and hematemesis, and endoscopic clipping may be utilized in those with significant bleeding.³

DISCLOSURES

Author contributions: T. Tariq wrote the manuscript. A. Rizvi and K. Badwal edited the manuscript. R. Eke edited the manuscript and is the article guarantor.

Financial disclosure: None to report.

Informed consent was not obtained for this case report as the patient was unreachable after multiple attempts. All identifying information has been removed.

Received February 13, 2018; Accepted June 25, 2018

REFERENCES

1. Jackson WE, Mehendiratta V, Palazzo J, Dimarino AJ, Quirk DM, Cohen S. Boerhaave's syndrome as an initial presentation of eosinophilic esophagitis: A case series. *Ann Gastroenterol*. 2013;26(2):166–9.
2. Quiroga J, Prim JMG, Moldes M, Ledo R. Spontaneous circumferential esophageal dissection in a young man with eosinophilic esophagitis. *Interact Cardiovasc Thorac Surg*. 2009;9(6):1040–2.
3. Predina JD, Anolik RB, Judy B, Akers S, Freiman D, Ahmad N, Singhal S. Intramural esophageal dissection in a young man with eosinophilic esophagitis. *Ann Thorac Cardiovasc Surg*. 2012;18(1):31–5.
4. Ortiz Moyano C, Gómez Rodríguez BJ, Pellicer Bautista F, Herrerías Gutiérrez JM. Intramural rupture of the esophagus: Clinical presentation and outcome. *Rev Esp Enferm Dig*. 2006;98(4):303–5.
5. Gluck M, Jiraneck GC, Low DE, Kozarek RA. Spontaneous intramural rupture of the esophagus: Clinical presentation and endoscopic findings. *Gastrointest Endosc*. 2002;56(1):134–6.
6. Stephens NA, Shah SK, Walker PA, Jayanty V, Rajjman I, Khalil K. Recurrent spontaneous esophageal dissection. *JLS*. 2014;18(2):342–5.
7. Sgrò A, Betalli P, Battaglia G, Bardini R, Crivellaro MA, Svaluto G, Gamba PG. An unusual complication of eosinophilic esophagitis in an adolescent: Intramural esophageal dissection. *Endoscopy*. 2012;44(Suppl 2 UCTN):E419–20.
8. Vernon N, Mohanany D, Ghetmiri E, Ghaffari G. Esophageal rupture as a primary manifestation in eosinophilic esophagitis. *Case Rep Med*. 2014;2014:673189.
9. Fischer A, Höppner J, Richter-Schrag H-J. First successful treatment of a circumferential intramural esophageal dissection with perforation in a patient with eosinophilic esophagitis using a partially covered self-expandable metal stent. *J Laparoendosc Adv Surg Tech A*. 2015;25(2):147–50.
10. Liguori G, Cortale M, Cimino F, Sozzi M. Circumferential mucosal dissection and esophageal perforation in a patient with eosinophilic esophagitis. *World J Gastroenterol*. 2008;14(5):803–4.
11. Jacobs JW, Spechler SJ. A systematic review of the risk of perforation during esophageal dilation for patients with eosinophilic esophagitis. *Dig Dis Sci*. 2010;55(6):1512–5.
12. Kedia S, Baruah BJ, Makharia G, Ahuja V. Eosinophilic esophagitis. *Trop Gastroenterol*. 2015;36(3):156–67.
13. Altun R, Akbas E, Yildirim AE, Öcal S, Korkmaz M, Selcuk H. Frequency of eosinophilic esophagitis in patients with esophageal symptoms: A single-center Turkish experience. *Dis Esophagus*. 2013;26(8):776–81.
14. Chiu Y-H, Chen J-D, Hsu C-Y, How C-K, Yen DH-T, Huang C-I. Spontaneous esophageal injury: Esophageal intramural hematoma. *J Chin Med Assoc*. 2009;72(9):498–500.
15. Lee M-H, Wu I-C, Lee J-Y. Spontaneous intramural esophageal dissection mimicking esophageal rupture. *Kaohsiung J Med Sci*. 2017;33(2):102–3.
16. Kim S, Lee SO. Circumferential intramural esophageal dissection successfully treated by endoscopic procedure and metal stent insertion. *J Gastroenterol*. 2005;40:1065.
17. Lucendo AJ, Frigal-Ruiz AB, Rodríguez B. Boerhaave's syndrome as the primary manifestation of adult eosinophilic esophagitis. Two case reports and a review of the literature. *Dis Esophagus*. 2011;24(2):E11–5.
18. Cao DT, Reny JL, Lanthier N, Frossard JL. Intramural hematoma of the esophagus. *Case Rep Gastroenterol*. 2012;6(2):510–7.