ACG CASE REPORTS JOURNAL



CASE REPORT | ESOPHAGUS

A Treatment Option for Esophageal Intramural **Pseudodiverticulosis**

Amy Tyberg, MD, and Daniela Jodorkovsky, MD

Division of Gastrointestinal and Hepatobiliary Diseases, New York Medical College, Westchester Medical Center, Valhalla, NY

Abstract

Esophageal intramural pseudodiverticulosis (EIPD) is a rare condition often presenting with esophageal strictures. Treatment is often limited to endoscopic dilatation and treatment of the underlying esophageal pathology. We present a case of a patient with longstanding GERD on famotidine (she experienced anaphylaxis with proton pump inhibitors [PPIs]) who presented with dysphagia and weight loss. Work-up revealed a diagnosis of EIPD with a 5-mm mid-esophageal stricture. Therapy with dilatation was unsuccessful until the addition of sucralfate, after which dilatation was successful and symptoms resolved. In patients who are unable to take PPIs, the addition of sucralfate may enhance the success of dilatations of esophageal strictures and EIPD.

Introduction

Esophageal intramural pseudodiverticulosis (EIPD) is a rare condition of unclear pathogenesis that was first described in 1960.1 Since then and through 2011, only about 200 cases have been reported worldwide.2 The true incidence of EIPD is unknown, though in a retrospective review of esophageal radiograms from 1986, EIPD was found to have a prevalence of approximately 0.15%.3 The primary symptom of this condition is dysphagia, usually due to the presence of an esophageal stricture that often accompanies diagnosis.^{4,5} Associated conditions reported in the literature include diabetes, alcoholism, gastroesophageal reflux disease (GERD), fungal infections, and esophageal neoplasms.^{4,5} Dysmotility disorders are occasionally associated with EIPD, including 2 reported cases of achalasia and 1 case of nutcracker esophagus, 6,7 though no causal relationship has been elucidated.8 Treatment is often limited to endoscopic dilatation and treatment of the underlying esophageal condition, such as acid suppression therapy or treatment of fungal infections.^{4,5,9-11} In rare cases, esophagectomy has been required.¹² We present a case of a patient with EIPD managed successfully with dilatation and sucralfate.

Case Report

A 58-year-old female was admitted to the hospital with several months of progressive dysphagia and a 20-lb weight loss. Her medical history was significant for GERD, for which she was only prescribed famotidine due to a prior anaphylactic allergy to proton pump inhibitors (PPIs). A barium esophagram showed a smooth midesophageal stricture and several intramural diverticula and intramural tracts distal to the stricture (Figure 1).

An esophagogastroduodenoscopy (EGD) was done to examine the stricture. The luminal diameter was approximately 5 mm, and length was 6 mm. There was no desquamation, ulceration, or furrowing of the mucosa. The stricture could only be traversed with an XP 180 endoscope. Distal to the stricture, the esophagus was found to have innumerable shallow depressions (Figure 2). Multiple biopsies were taken from within and adjacent to the stricture. Pathology showed histologic evidence of esophagitis with neutrophils and lymphocytes, and rare

ACG Case Rep J 2014;1(3):134-136. doi:10.14309/crj.2014.28. Published online: April 4, 2014.

Correspondence: Amy Tyberg, New York Medical College, Gastrointestinal and Hepatobiliary Diseases, Munger Pavilion Suite 206, Valhalla, NY, 10595 (amy.tyberg@gmail.com).

Copyright: © 2014 Tyberg et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

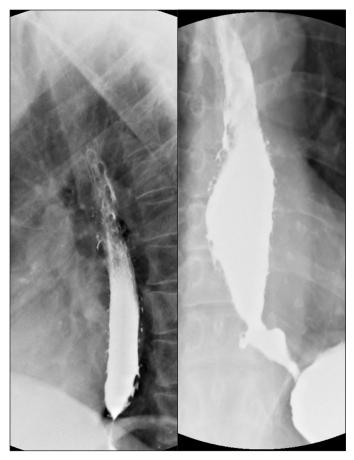


Figure 1. Barium swallow showing intramural diverticula and intramural

eosinophils without evidence of malignancy or an alternative diagnosis. The patient was diagnosed with EIPD.

A series of near-weekly balloon dilatations were performed with minimal success. The first dilatation session used a 6-mm, followed by a 7.5-mm, through-the-scope (TTS) balloon. Ten days later, bougie dilatation was performed using a 27 French dilator, and moderate force was required. A TTS balloon was then used to dilate from 9 mm to 10 mm. Two weeks later, the stricture was still unable to be traversed with a GIF-160, and the diameter was estimated to have returned back to 8 mm. A TTS balloon was used to dilate from 8 mm to 12 mm. The fourth dilatation session showed that the stricture still could not be traversed: the diameter had narrowed again. A TTS balloon was used to dilate from 12 mm to 14 mm.

The patient was then prescribed sucralfate suspension 4 times daily. The next dilatation started with a 12-mm balloon, and the stricture was dilated to 15 mm. Dysphagia symptoms largely resolved at this time, though the patient noted on follow-up that she had to be "very careful" eating solid food and chased all oral intake with liquid. One final

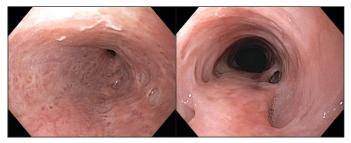


Figure 2. EGD showing esophagus with innumerable shallow depressions.

dilatation session was performed 1 month later. The stricture could be easily traversed with a GIF-160, and the luminal diameter was estimated to be 12 mm. The stricture was dilated with a 15–18-mm controlled radial expansion (CRE) balloon. After these dilatations, the patient's dysphagia completely resolved, and she has since gained 21 lbs. She has not required further dilation at 1 year follow-up.

Discussion

EIPD is a rare disorder with an unclear pathogenesis. It is often associated with esophageal strictures, and therapy consists mainly of endoscopic dilatation. There have been no reports of the use of esophageal stents for the treatment of EIPD in the literature. EIPD can be associated with other conditions, the most common of which are GERD and fungal infections. Treatment of these other underlying conditions, such as acid suppression in the setting of GERD, is often required. In patients who are unable to tolerate standard acid suppression therapy with proton-pump inhibitors (PPIs), the addition of sucralfate may enhance the success of dilatations of esophageal strictures, as our case illustrates. A recent Cochrane review showed sucralfate to be associated with a trend towards esophagitis healing in the setting of GERD, though the effect was modest and not statistically significant.¹³ Sucralfate functions by forming a physical barrier between esophageal mucosa and harmful agents, promoting mucosal healing, and decreasing the inflammatory response.14 All of these mechanisms may explain its beneficial role in this case. Regardless of the mechanism, this case suggests that in patients with EIPD and stricture formation who are unable to tolerate PPI therapy, sucralfate may be a viable alternative.

Disclosures

Author contributions: A. Tyberg collected data, wrote the manuscript, and is the article guarantor. D. Jodorkovsky edited the manuscript.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received: November 20, 2013; Accepted: March 18, 2014

References

- Mendl K, McKay JM, Tanner CH. Intramural diverticulosis of the esophagus and Rokitansky-Aschoff sinuses in the gallbladder. *Br J Radiol*. 1960;33(392):496–501.
- 2. Hahne M, Schilling D, Arnold JC, Riemann JF. Esophageal intramural pseudodiverticulosis: Review of symptoms including upper gastrointestinal bleeding. *J Clin Gastroenterol*. 2001;33(5):378–82.
- Levine MS, Moolten DN, Herlinger H, Lauger I. Esopheal intramural pseudodiverticulosis: A reevaluation. Am J Roentgenol. 1986;147(6):1165–70.
- Brühlmann WF, Zollikofer CL, Maranta E, et al. Intramural pseudodiverticulosis of the esophagus: Report of seven cases and literature review. Gastrointest Radiol. 1981;6(3):199–208.
- Umlas J, Sakhuja R. The pathology of esophageal intramural pseudodiverticulosis. Am J Clin Pathol. 1976;65(3):314–20.
- Turan I, Ozen E, Bor S, Ozutemiz O. Esophageal intramural pseudodiverticulosis associated with achalasia: An unusual endoscopic appearance. *Endoscopy*. 2009;41(suppl 2):E23–E24.
- Walker S, Hippéli R, Goës R. Diffuse esophageal intramural pseudodiverticulosis and nutcracker esophagus in a 54-year-old man. Klin Wochenschr. 1990;68(3):187–90.

- Sabanathan S, Salama FD, Morgan WE. Oesophageal intramural pseudodiverticulosis. *Thorax*. 1985;40(11):849–57.
- Chiba T, Iijima K, Koike T, et al. A case of severe esophageal intramural pseudodiverticulosis whose symptoms were ameliorated by oral administration of anti-fungal medicine. Case Rep in Gastroenterol. 2012;6:103–110.
- 10. Attila T, Marcon NE. Esopahgeal intramural pseudodiverticulosis with food impaction. *Can J Gastroenterol.* 2006;20(1):37–38.
- 11. Chon YE, Hwang S, Jung KS, et al. A case of esophageal intramural pseudodiverticulosis. *Gut Liver.* 2011;5(1):93–5.
- Liu SM, Wu HH, Chang KK, et al. Esophageal intramural pseudodiverticulosis complicated with stricture. *J Formos Med Assoc.* 2010:109(3):241–4.
- Khan M, Santana J, Donnellan C, et al. Medical treatments in the short term management of reflux oesophagitis. *Cochrane Database Syst Rev.* 2007;2:CD003244.
- Gümürdülü Y, Karakoc E, Kara B, et al. The efficiency of sucralfate in corrosive esophagitis: A randomized, prospective study. *Turk J Gastro-enterol*. 2010;21(1):7–11.

Publish your work in ACG Case Reports Journal

ACG Case Reports Journal is a peer-reviewed, open-access publication that provides GI fellows, private practice clinicians, and other members of the health care team an opportunity to share interesting case reports with their peers and with leaders in the field. Visit http://acgcasereports.gi.org for submission guidelines. Submit your manuscript online at http://mc.manuscriptcentral.com/acgcr.