Published in final edited form as:

Dig Dis Sci. 2018 May ; 63(5): 1139–1142. doi:10.1007/s10620-017-4756-y.

(Re)building the Wall: Recurrent Boerhaave Syndrome Managed by Over-the-Scope Clip and Covered Metallic Stent Placement

Monique Barakat, MD, PhD, Mohit Girotra, MD, and Subhas Banerjee, MD

Division of Gastroenterology and Hepatology, Stanford University, CA, USA

Keywords

Boerhaave syndrome; esophageal perforation; esophageal stent; over-the-scope clip

Case presentation and evolution

A 62-year-old man was initially evaluated at a local hospital complaining of shortness of breath and severe, persistent pain in the chest and epigastric regions with radiation to the left shoulder and neck. The pain was exacerbated by hiccupping, coughing and deep inspiration. He reported heavy alcohol consumption and recurrent migraine headaches the night before and that the pain had followed a protracted episode of retching and vomiting. His past medical history was notable for a 4 cm ascending aortic aneurysm and two previous episodes of esophageal rupture occurring under similar circumstances. The first episode of perforation had occurred 12 years previously and was managed with chest tube placement and antibiotics. The second, which had occurred 4 years previously had resulted in a tracheo-esophageal fistula that was managed by endoscopic stent placement and right-sided empyema requiring thoracotomy with decortication and debridement. The esophageal stent was removed three months after placement and repeat upper endoscopy was notable only for Barrett's esophagus.

Based on his history suggestive of recurrent Boerhaave syndrome and evidence of cardio-pulmonary instability, the patient was urgently transferred to our tertiary care medical center for management. Vital signs were notable for a heart rate of 121 beats/min, temperature of 38 °C, blood pressure of 89/62 mmHg and respiratory rate of 21/min with an oxygen saturation of 84% on room air. Physical examination revealed increased respiratory effort with use of the accessory muscles, presence of subcutaneous emphysema and dullness to percussion at the lung bases with diminished breath sounds bilaterally.

Computed tomography (CT) scan with oral contrast administration revealed a defect in the anterior wall of the distal esophagus with contrast extravasation into the mediastinum and bilateral pleural spaces, right greater than left (Figure 1). The patient was admitted to the cardiovascular intensive care unit (ICU). A left-sided chest tube was easily placed, but right-

sided chest tube placement was initially unsuccessful due to regional scarring and adhesions. Image-guided right-sided chest tube placement was subsequently successfully performed by interventional radiology, resulting in drainage of a significant volume of fluid. The patient was then referred for endoscopic management of his Boerhaave syndrome.

Esophagogastroduodenoscopy performed under general anesthesia revealed a 10-mm transmural defect in the wall of the esophagus 32 cm from the incisors (Figure 2A, B) through which the mediastinum was visible and brownish fluid was draining. The edges of the defect appeared indurated; mucosal scarring was evident in the adjacent esophagus. An over-the-scope clip (OTS clip, Ovesco Inc, Tubingen, Germany) was then mounted on the endoscope which was re-advanced into the esophagus. Grasping and approximation of the edges of the defect with a 'twin grasper' accessory device (Ovesco Inc, Tubingen, Germany) was challenging due to the indurated, fibrotic tissue surrounding the defect, presumably resulting from the patient's previous esophageal ruptures. The OTS clip was eventually deployed using continuous suction only (Figure 2C). No further drainage of brown fluid from the mediastinum was noted after clip placement. Repeat esophagram the following morning demonstrated a small amount of right-sided contrast extravasation from just inferior to the OTS clip, suggesting a residual small unclosed defect.

Repeat upper endoscopy demonstrated the previously-placed OTS clip located 32 cm from the incisors and a residual 1 mm microdefect just distal to the inferior aspect of the OTS clip. A 23 × 103 mm fully covered metallic stent (Wallflex, Boston Scientific, Marlborough, Massachusetts, United States) was then deployed across the OTS clip and residual perforation site under endoscopic and fluoroscopic guidance. Three endoscopic clips (Resolution 360™ Clip, Boston Scientific, Marlborough, Massachusetts, United States) were deployed at the proximal and distal aspect of the stent to minimize the risk of stent migration (Figure 2D). Repeat esophagram the following morning demonstrated the stent in place across the esophageal defect and OTS clip, with no evidence of overt extravasation from the residual small esophageal defect.

Following this second endoscopic intervention, the patient's hemodynamic status improved, his fever resolved, his pain gradually improved, and his chest tube output gradually decreased, eventuating in discharge several days later. Repeat upper endoscopy with stent removal was performed 4 weeks following discharge. The previously-placed OTS clip was visualized, with no evidence of a residual esophageal defect.

Discussion

Esophageal perforation is the most morbid disruption of the gastrointestinal (GI) tract, with a 35% overall mortality rate.¹ Iatrogenic injury is the most common cause of esophageal perforation, accounting for up to 90% of cases. Traumatic esophageal injury accounts for the remainder of esophageal perforation cases. Boerhaave syndrome accounts for a small subset of traumatic esophageal perforations. Transmural esophageal perforation developing after forceful emesis, first described by Boerhaave in 1724,² is thought to result from a sudden Valsava induced increase in intraluminal esophageal pressure during vomiting, when the cricopharyngeal muscle fails to relax during the emesis.³ The left posterolateral wall of the

lower third of the esophagus, 2–3 cm proximal to the gastroesophageal junction is the most common site of rupture in Boerhaave syndrome. Boerhaave syndrome, most commonly occurring in men aged 50 – 70 years is distinct from Mallory-Weiss syndrome in that the latter is a more superficial and less morbid esophageal tear associated with vomiting.²

While iatrogenic perforations are typically recognized during or shortly after a procedural intervention, the diagnosis of Boerhaave syndrome can be more challenging, as more than one-third of patients initially display atypical signs and symptoms.⁴ Mackler's classic triad of vomiting, chest pain and subcutaneous emphysema is present in approximately 14% of patients. When esophageal perforation is suspected, chest x-ray, water soluble contrast upper GI series, and CT scan may all be considered. These studies may demonstrate free air or fluid collections in the mediastinum or retroperitoneum.³ A discrete site of contrast extravasation from the esophagus may be visualized.

Common initial misdiagnoses in patients with Boerhaave syndrome include myocardial infarction, primary empyema, pericarditis, or pneumothorax.³ Diagnosis and intervention within 24 hours of esophageal rupture is associated with a favorable survival rate of 75%.⁴ Mortality rates of >50% have been reported with management delays of >24 hours, whereas delays of >48 hours are associated with mortality rates of >90%.¹ Mortality is typically related to an infectious process associated with the esophageal rupture, including mediastinitis, empyema, pneumonitis, and pericarditis.

The optimal therapeutic approach to Boerhaave syndrome remains undefined and may depend on perforation size and sequelae, as well as on the patient's overall clinical status.^{5, 6} The therapeutic approach includes administration of antibiotics, closure of the esophageal defect, drainage of any fluid collections and provision of nutritional support while the defect heals.

In past decades, surgery was the principal option available for the repair of the esophageal defect.⁷ Surgical approaches include open, laparoscopic, or video-assisted thorascopic surgery to accomplish mediastinal washout, direct repair of the esophageal defect, or esophageal resection and diversion.^{6–8} The choice of surgical approach typically depends on the timing of presentation and hemodynamic stability of the patient, with the surgical 'gold standard' of management being open laparotomy or thoracotomy.⁸

The evolution of endoscopic techniques and device technology now provides a variety of minimally invasive and less morbid therapeutic interventions. Endoscopic approaches for management of esophageal perforations include placement of fully covered esophageal stents, through-the-scope (TTS) clips, OTS clips, and endoscopic suturing (OverStitch, Apollo Endosurgery Inc, Austin, TX, United States).⁵ TTS or OTS clips alone may suffice for closure of small perforations, whereas stenting is often undertaken for the management of larger esophageal tears. Anchoring the esophageal stent is important, as fully covered stents have a high migration rate, particularly in the absence of retaining strictures.⁵ Esophageal stents have been anchored in place using TTS clips, OTS clips or endoscopic suturing.⁵

To our knowledge, the present case is the first report of dual endotherapy with OTS clip closure followed by endoluminal stent placement for the management of recurrent Boerhaave syndrome. Although placement of an OTS clip alone usually suffices for closure of smaller esophageal defects,⁹ OTS clip placement was challenging in this patient due to the recurrent nature of his transmural esophageal rupture with resultant fibrosis and rigidity in the region of the transmural defect. Indurated/fibrotic tissue is challenging to grasp and close with OTS clips with studies reporting a 42.9% closure rate for chronic fistulae compared to a 90% closure rate for fresh defects.¹⁰ Prior retrospective studies have indicated a need for salvage therapy, typically esophageal stent placement, in up to 40% of patients with Boerhaave's syndrome due to failure of primary endoscopic or surgical management.⁵ Based on this high failure rate with monotherapy, one might consider dual endoscopic therapy with clip closure and endoluminal stent placement as the initial approach for patients with Boerhaave syndrome and particularly for those with recurrent Boerhaave syndrome. This case report highlights the continued transition in management of gastrointestinal conditions from invasive and morbid surgical intervention to rapid, safe and effective endotherapy, brought about by the progressive evolution of endoscopic techniques and technology.

References

1. de Schipper JP, Pull ter Gunne AF, Oostvogel HJ, et al. Spontaneous rupture of the oesophagus: Boerhaave's syndrome in 2008. Literature review and treatment algorithm. *Dig Surg*. 2009; 26:1–6. [PubMed: 19145081]
2. Thorbjarnarson B. The Mallory-Weiss and Boerhaave syndromes. *N Y State J Med*. 1986; 86:285–6. [PubMed: 3461328]
3. Tonolini M, Bianco R. Spontaneous esophageal perforation (Boerhaave syndrome): Diagnosis with CT-esophagography. *J Emerg Trauma Shock*. 2013; 6:58–60. [PubMed: 23493470]
4. Wise MP, Salmon JB, Maynard ND. Boerhaave syndrome: a diagnostic conundrum. *BMJ Case Rep*. 2009; 2009
5. Dickinson KJ, Buttar N, Wong Kee Song LM, et al. Utility of endoscopic therapy in the management of Boerhaave syndrome. *Endosc Int Open*. 2016; 4:E1146–E1150. [PubMed: 27853740]
6. Schweigert M, Beattie R, Solymosi N, et al. Endoscopic stent insertion versus primary operative management for spontaneous rupture of the esophagus (Boerhaave syndrome): an international study comparing the outcome. *Am Surg*. 2013; 79:634–40. [PubMed: 23711276]
7. Vyslouzil K, Kral V, Neoral C, et al. Surgical treatment of spontaneous esophageal perforation--Boerhaave syndrome. *Rozhl Chir*. 2000; 79:521–3. [PubMed: 11210602]
8. Kimberley KL, Ganesh R, Anton CK. Laparoscopic repair of esophageal perforation due to Boerhaave syndrome. *Surg Laparosc Endosc Percutan Tech*. 2011; 21:e203–5. [PubMed: 21857462]
9. Kobara H, Mori H, Rafiq K, et al. Successful endoscopic treatment of Boerhaave syndrome using an over-the-scope clip. *Endoscopy*. 2014; 46(Suppl 1 UCTN):E82–3. [PubMed: 24676827]
10. Haito-Chavez Y, Law JK, Kratt T, et al. International multicenter experience with an over-the-scope clipping device for endoscopic management of GI defects (with video). *Gastrointest Endosc*. 2014; 80:610–22. [PubMed: 24908191]

Key Points

- The esophagus is the site of perforation within the gastrointestinal tract associated with the most morbidity, with an overall 35% mortality rate.
- Transmural esophageal perforation developing as a consequence of forceful retching or coughing is known as Boerhaave syndrome
- Delays in diagnosis are associated with a significant increase in mortality in patients with Boerhaave syndrome
- Whereas surgery was previously the mainstay of management, several effective endoscopic management options are now available.
- Dual endoscopic therapy with clip closure followed by covered stent placement may offer more robust defect closure.

Figure 1.

(A) CT scan demonstrating extensive intrathoracic pneumomediastinum with a defect in the distal esophagus (red arrow) approximately 3 cm above the gastroesophageal junction and (B) a thick-walled gas, fluid and contrast-containing collection in the posterior right pleural space (red arrows point toward collection).

Figure 2.
Endoscopic image of the transmural esophageal defect before (yellow box in A, yellow arrows in B) and after placement of an over-the-scope clip (C).

Figure 3.

(A) Endoscopic image of the proximal end of the covered metallic esophageal stent prior to anchoring clip placement. (B) Fluoroscopic image demonstrating the esophageal stent secured in place by three through-the-scope clips proximally and distally.