



Boerhaave syndrome complicated by undiagnosed gastroesophageal junction outlet obstruction: a case report

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Background: Boerhaave syndrome is a condition carrying high morbidity and mortality, necessitating emergency intervention. Along with appropriate resuscitation and drainage, accepted definitive therapy aims for either open transpleural primary closure, placement of an endoluminal covered stent, or a combination of both. Only in very rare cases does a rupture occur exclusively within the abdomen, without violation of either pleural space. We present a case in which the patient's vague history and current imaging suggest undiagnosed gastroesophageal junction outlet obstruction (GEJOO), possibly achalasia, which in large part contributed to his rupture. Also unusual is the isolation of the spontaneous tear to the abdomen. In this case, we detail the integration of strategies to repair his injury, protect the repair, and help prevent recurrence of dysphagia or rupture.

Case Description: A 64-year-old male presented with 6 hours of chest pressure after having choked on meat and then retched extensively. During the admission, he was tachycardic, in distress with pallor and diaphoresis, a distended and tender abdomen, without peritonitis. Computed tomography (CT) esophagram revealed a tortuous and dilated esophagus with tight narrowing at the gastroesophageal (GE) junction, contrast spillage in the lower right epigastrium, and extensive free air in the retroperitoneum and abdominal space. The patient was ultimately diagnosed with a distal esophageal perforation with isolation to the intra-abdominal space. He was taken urgently to the operating room where the decision was made to proceed with a laparotomy. The perforation was repaired primarily with suture, followed by myotomy and reinforcement of the repair with a 360-degree fundoplication. A gastric feeding tube was placed for initial decompression in the early postoperative period.

Conclusions: Herein we have presented a patient with a life-threatening, spontaneous esophageal tear, with the complicating feature of a previously underestimated GEJOO. Prompt recognition of his acute injury, along with identification of his esophageal anatomy, allowed for a technically feasible repair, with very smooth, early and midterm recovery. GEJOO should be considered when a patient presents with a rupture of the esophagus and abnormal esophageal anatomy on imaging. This will allow for potential incorporation of myotomy and fundoplication in the surgical repair.

Keywords: Achalasia; case report; Boerhaave syndrome; gastroesophageal junction outlet obstruction (GEJOO); esophageal rupture

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Introduction

Boerhaave syndrome is a condition carrying high morbidity and mortality, necessitating emergency intervention. Along with appropriate resuscitation and drainage, accepted definitive therapy aims for either open transpleural primary closure, placement of an endoluminal covered stent, or sometimes a combination of both strategies. Only in very rare cases does a rupture occur exclusively within the abdomen, without violation of either pleural space. Occasionally, a patient's premorbid esophageal anatomy presents special challenges—such as stricture, malignancy, or other structural complexity—that requires alternative approaches to addressing both immediate and long-term recovery.

We present a case in which the patient's vague history and current imaging suggest undiagnosed gastroesophageal junction outlet obstruction (GEJOO), possibly achalasia, which in large part contributed to his rupture. Also unusual is the isolation of the spontaneous tear to the abdomen. In this case we detail the integration of strategies to repair his injury, protect the repair, and help prevent recurrence of dysphagia or rupture. We present the following case in accordance with the CARE reporting checklist (available at <https://dmr.amegroups.com/article/view/10.21037/dmr-22-61/rc>).

Case presentation

A 64-year-old, relatively healthy nonsmoker male presented with 6 hours of chest pressure after having choked on chicken meat and then retched extensively. He reported two prior episodes of dysphagia in his lifetime, including an episode of food bolus 5 months earlier, at which time he underwent an esophagogastroduodenoscopy (EGD) that revealed an extensive mucosal tear at 30–35 cm from the incisors. There was no stricture identified at that time; the patient was discharged on oral Protonix and had no follow-up.

During the present admission, he was tachycardic, in visible distress with pallor and diaphoresis, with a distended and tender abdomen, without peritonitis. Computed tomography (CT) esophagram revealed a very tortuous and dilated esophagus with tight narrowing at the gastroesophageal (GE) junction, contrast spillage in the lower right epigastrium, and extensive free air in the retroperitoneum and abdominal space. There were also bilateral small pleural effusions without evidence of intrapleural contrast extravasation (see *Figures 1-3*).

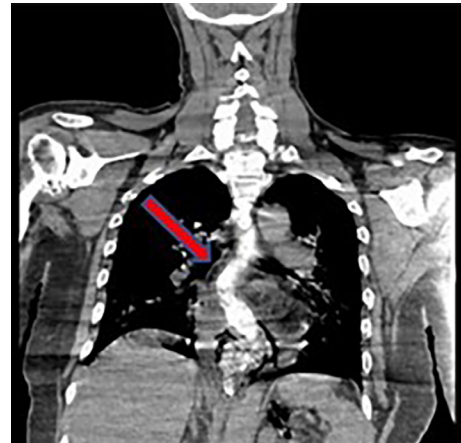


Figure 1 Coronal CT indicating a tortuous esophagus (red arrow). CT, computed tomography.

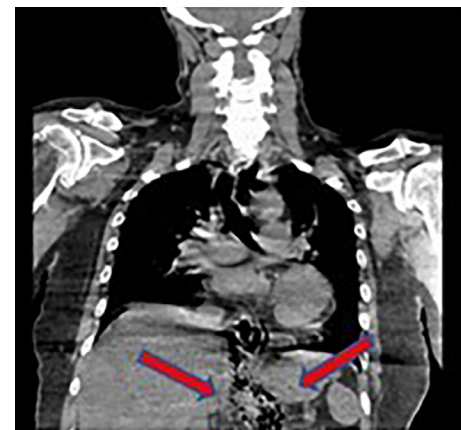


Figure 2 Coronal CT indication free intra-abdominal air surrounding the distal esophagus (red arrows). CT, computed tomography.

Abnormal lab values demonstrated leukocytosis to 19.4 K/ μ L and creatinine elevated to 1.56 mg/dL.

After fluid resuscitation and antibiotics, the surgical team made plans for immediate, definitive surgical intervention.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

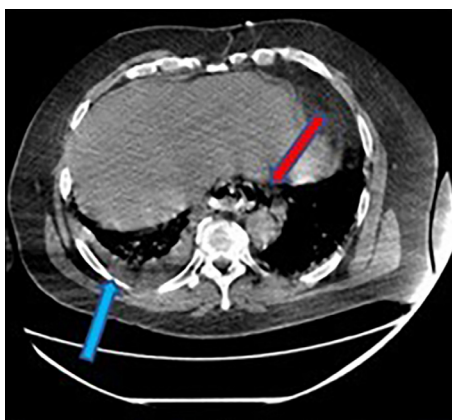


Figure 3 Axial CT indicating free intra-abdominal air (red arrow) surrounding the esophagus and a small right pleural effusion (blue arrow). CT, computed tomography.

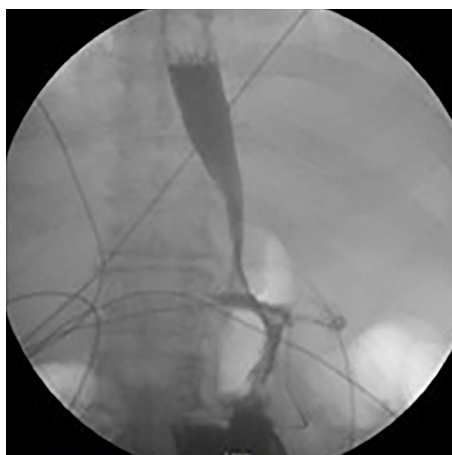


Figure 4 Water-soluble contrast study indicating no extravasation at repair site and normal esophageal morphology.

Operation

In the operating room, the decision was made to approach the distal esophageal leak via laparotomy. After safe entry, a nasogastric tube was guided into the stomach. The hiatus was then gently exposed, revealing a 4-cm longitudinal transmural defect at the right posterolateral aspect just 2 cm proximal to the GE junction. No other abnormalities were seen at the hiatus, therefore approximately 8 to 10 cm of mobilization was undertaken into the mediastinum. The tissue at the esophageal injury was friable, and mucosa unable to be fully exposed based on body habitus and difficult exposure, so a classical two-layer closure was not possible. We did, however, place full-thickness interrupted

silk sutures followed by another running silk layer, with the nasogastric tube as a guard against stricture.

We then planned for fundoplication for optimal coverage of the repair, as well as to complement a contralateral myotomy, which would aim to help prevent recurrence of narrowing at the GE junction and adequately treat GEJOO as his presumptive diagnosis. Short gastric vessels were divided with LigaSure device (Medtronic, Minneapolis, MN, USA); the fundus was found to be very generous and estimated to be able to accommodate a loose 360-degree wrap. We then undertook a myotomy on the esophagus contralateral to the injury, extending approximately 6 cm proximal to the GE junction, and 4 cm distal to the GE junction onto the gastric cardia. Nissen fundoplication was then performed, using three double-pledgeted Ethibond sutures (Ethicon, Cincinnati, OH, USA), with the upper two sutures incorporating intact esophageal wall. Per-oral bougie was not used based on tortuosity of the esophagus. When completed, the wrap extended approximately 4 cm, and covered both the injury repair and myotomy sites. Gastric feeding tube was then placed via standard Stamm technique, and a large drain was left within the lesser sac. The patient tolerated the procedure well and was extubated within 6 hours.

During his recovery period, the patient was kept nil per os (NPO). The gastrostomy tube was initially used for decompression, then on postoperative day (POD) 2 he began receiving tube feeds. On POD 5, a water-soluble contrast study was performed (see *Figure 4*), demonstrating nearly normal esophageal morphology and transit, healed leak site, with prompt contrast passage via lower esophageal sphincter (LES) and pylorus. The retroperitoneal drain, from which very little thin drainage was captured after surgery, was discontinued. The patient was then started on a clear liquid diet and quickly advanced to a post-Nissen regimen. He was discharged to home on POD nine and was found in clinic to be recovering very well at POD 22, at which time his gastrostomy tube was removed. The patient experienced no adverse events during the hospital course or post-op follow-up.

Discussion

Esophageal rupture most commonly occurs due to iatrogenic causes, while about 15% of cases involve Boerhaave type injury induced by forceful vomiting. This phenomenon was first described by Dutch physician Herman Boerhaave (circa 1724) (1) and carries a mortality rate of between

10% and 40% in collected series (2). This type of rupture, isolated to the abdomen, is exceedingly uncommon; in one of the largest worldwide meta-analyses of 75 studies, involving 2,971 patients over 12 years, only 77 patients were identified to have had abdominal involvement (3). Meanwhile, we find a case describing achalasia-related injury, in which an elderly female suffered an intrathoracic rupture, was treated with multiple stents, and succumbed to her condition within 1 month (4). Another published case describes Boerhaave syndrome secondary to benign GEJOO. In the case report they describe an elderly female who presented with a gastric volvulus as the cause of the esophageal rupture. The patient underwent surgical drainage and irrigation of the mediastinum and pleural cavities, followed by a gastrostomy to fix and decompress the stomach. The patient was discharged after an uneventful hospital course (5).

Generally accepted principles of successful therapy for esophageal rupture include timely recognition, thorough resuscitation, and earliest treatment according to the individual clinical details of this very heterogeneous condition (3). Primary repair and stent placement show similar survival rates, while patients whose acuity mandates control measure such as t-tube, have a much higher mortality. Patients with esophageal cancer, or who require esophagectomy, fare very poorly (3).

GEJOO, such as achalasia, displays an incidence of approximately 1 per million per year, and is characterized by the Chicago Classification (6). This class of disorders, in general, involves abnormalities of both excitatory and inhibitory motor neurons, interfering with esophageal body muscle contraction as well as relaxation of the LES (6,7). High resolution manometry is currently the standard of care for diagnosis, classification, and determination of recommended treatment and prognosis. Our patient, in extremis, was unable to undergo formal esophageal testing, therefore the strong clinical clues—food bolus episodes in the past, “bird’s beak” type of LES narrowing and esophageal dilation and tortuosity upon presentation—were utilized to plan for immediate surgical remedy that would also accommodate for longer term recovery. Strong data show between 81% and 100% success rate of myotomy and fundoplication for all types of GEJOO (8), and our patient’s postop studies show no GE junction stenosis and resolution of the esophageal dilation and tortuosity, so this appears to have been the right procedure for the patient. In the longer term, our patient will undergo follow-up manometric studies, as well as surveillance imaging and endoscopy.

Conclusions

Herein we have presented a patient with a life-threatening, spontaneous type of esophageal tear, with the complicating feature of a previously underestimated GEJOO. Prompt recognition of his acute injury, along with identification of his esophageal anatomy, allowed for a technically feasible repair, with very smooth, early and midterm recovery.

GEJOO should be considered when a patient presents with a rupture of the esophagus and abnormal esophageal anatomy on imaging. This will allow for potential incorporation of myotomy and fundoplication in the surgical repair.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at <https://dmr.amegroups.com/article/view/10.21037/dmr-22-61/prf>

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at <https://dmr.amegroups.com/article/view/10.21037/dmr-22-61/coif>). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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