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Boerhaave Syndrome: A Case Report and Literature Review

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Abstract **Case Report**

Boerhaave's syndrome is a rare and serious condition associated with high mortality and morbidity. Diagnosis of this syndrome is usually done essentially with the aid of imaging. Treatment for this syndrome has been mainly surgical since its discovery by Herman Boerhaave; however, multiple endoscopic approaches have been successfully used recently withthe advancement of this field. Here, we describe a case of Boerhaave's syndrome; a 18 y woman with histoy of pancreatitis 2 months ago; presented to emergency for complaining of sudden, right-sided, non-radiating, pleuritic chest pain for an hour, associated with cough, shortness of breath, and palpitations; a CT scan found a tear in the distal esophagus and anterior mediastinum; The patient underwent an emergency left thoracotomy, repair of oesophageal junction perforation The patient was discharged to a long-term acute care facility.

Keywords: Pneumothorax (ptx), CT, endoscopic management, spontaneous esophageal perforation, boerhaave's syndrome.

INTRODUCTION

Esophageal perforation is a rare clinical entity with an estimated incidence of 3.1 per 1,000,000 per year [1]. It is most commonly caused by iatrogenic mechanisms, like endoscopy or surgery-related phenomena, or non-iatrogenic trauma. An extremely rare cause is the effort rupture of the esophagus, aka Boerhaave's syndrome, which makes up 15% of esophageal perforation cases [2]. This syndrome usually presents with chest pain [3], which can result in a delayed diagnosis as cardiac etiologies are typically pursued initially, particularly in elderly populations.

Boerhaave's syndrome presenting with tension pneumothorax is a rare presentation with few documented cases in the literature [4]. We present a case followed by a discussion on the factors leading to surgical versus endoscopic management of the patients presenting with Boerhaave syndrome and a review of the literature.

CLINICAL CASE

18-year-old woman presented to the emergency unit complaining of sudden, right-sided, non-radiating, pleuritic chest pain for an hour, associated with cough, shortness of breath, and palpitations. The patient had been feeling nauseous and reported a few episodes of non-bloody emesis. Patient present a medical history of pancreatitis a 2 months ago, no intake of medications and no relevant past chirurgical could be elucidated.

On physical exam, the patient was afebrile, normotensive (blood pressure (BP) 139/79 mmHg), tachycardiac (heart rate (HR) 108 bpm), tachypneic (respiratory rate (RR) 28 br/min), saturating 90% on ambient air.

Labs showed leukocytosis, mild electrolyte derangements, and lactic acidosis, CRP=120; and lipase serum level =700.

Computed tomography (CT) chest with contrast (Figure 1) showed esophageal thickening with a massive pneumomediastinum and pneumopericardium adjacent to the mid and distal third of the esophagus concerning for esophageal perforation. A bilateral Basi thoracic infectious pneumonia.

Shortly thereafter, the patient developed hypoxic respiratory failure and was transferred to the intensive care unit for mechanical ventilation.

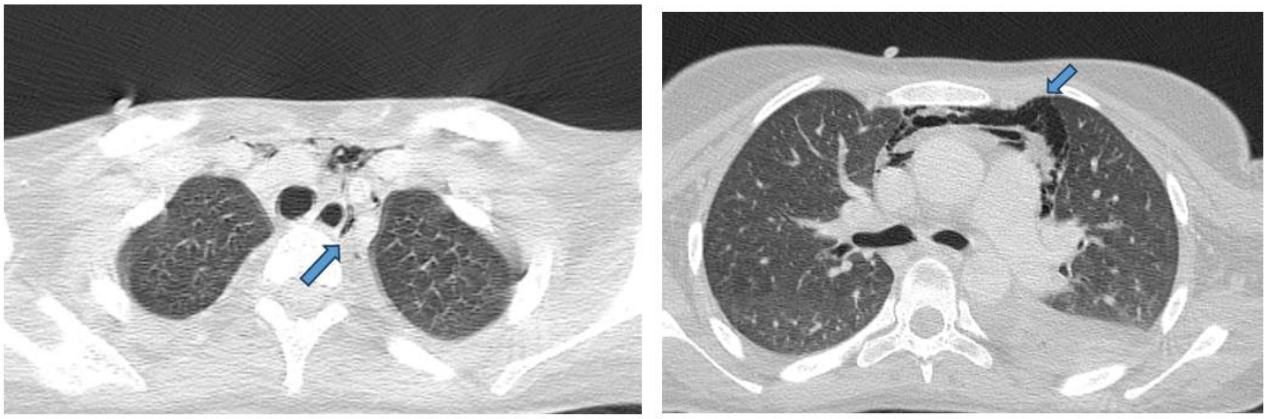

Figure 1: CT chest with contrast showing esophageal thickening with pneuma-mediastinum

DISCUSSION

Boerhaave's syndrome usually occurs due to a sudden increase in the intra-esophageal pressure associated with the negative intrathoracic pressure, causing a traction force that leads to this perforation.

This happens in the setting of forceful vomiting and/or retching [5]. Rupture commonly involves the posterolateral part of the distal intrathoracic esophagus [6]. Nonetheless, it can also occur in other parts of the esophagus such as the intra-abdominal or cervical esophagus. This rupture leads to the leakage of the gastric and esophageal components into the mediastinum causing chemical mediastinitis, bacterial infection, necrosis, and death.

The mortality rate for untreated Boerhaave's syndrome can reach 90% [7]. Although the exact morbidity of this syndrome is unknown, it is considered high [8].

Management of this syndrome starts with supportive measures including resuscitation, administering broad-spectrum antibiotics and proton pump inhibitors, surgical consultation, as well as possible ICU admission. Subsequent management varies depending on whether the perforation is contained or not.

Contained perforations are usually managed medically by avoidance of oral intake, parenteral nutritional support, drainage of fluid collections, and continuing antibiotics. Patients with uncontained perforations or those who fail medical therapy can be managed surgically or endoscopically.

Traditionally, endoscopic therapy has been reserved for poor surgical candidates. Thus far, only one study was found that compares outcomes of surgically versus endoscopically managed Boerhaave's syndrome. This study was in 2013, was performed in Europe, and included only 38 patients. It concluded that endoscopic management has no advantage over surgical treatment in terms of morbidity and ICU stay [9].

However, the advancement in the endoscopy field and expertise in recent years, small sample size, and scarcity of studies on this topic might refute that.

Different endoscopic methods have been used to manage esophageal perforations, including esophageal stents, endoscopic suturing, over-the-scope clips (OTSCs), and through-the-scope clips (TTSCs). The choice of endoscopic intervention depends on the size, location, extent as well as margins of the defect [10].

Endoscopic clipping, using OTSCs or TTSCs, is usually preserved for defects that are no greater than 20 mm.

The success rate of OTSCs was found to be 100% for esophageal perforations in one multicentric retrospective study, which included 188 patients with GI defects that were managed by OTSCs, 10 of which were esophageal perforations [11]. Esophageal stenting has up to 91.4% technical success rate [12]. Due to the risk of stent migration, careful consideration should be given to the location of the defect. Stent clinical failure has been observed in defects located at upper and lower esophageal sphincters, as well as defects larger than 60 mm in size [13].

Endoscopic suturing also has a comparable success rate of 95.7% in one retrospective study; however, this was for different GI defects and not just esophageal perforations [14].

CONCLUSION

Overall, the endoscopic management of GI defects in general and esophageal perforation in specific is a promising field that needs further studying and more evidence to structure it and include it in future guidelines. Increasing awareness among physicians about those successful non-surgical options is also needed to improve outcomes of this syndrome.

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