Case Report

Non-traumatic esophageal perforation: A case report on Boerhaave's syndrome

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ABSTRACT

Boerhaave's syndrome is a potentially fatal condition characterized by spontaneous perforation of a previously healthy esophagus, due to severe vomiting or straining. It often presents with non-specific symptoms such as fever, pain, and vomiting and hence may go undiagnosed. The Makler's triad, consisting of vomiting, chest pain, and subcutaneous emphysema, may be seen in only 50% of cases. Delayed diagnosis may result in complications such as sepsis, mediastinitis, pneumothorax, and multi-organ dysfunction. In general, patients presenting later than 48 h are conservatively managed with esophageal stenting. Surgical repair is usually reserved for those patients who present within 24 h, or are managed conservatively and develop complications. Mortality rises from 0% if treated within 24 h to about 29% if delayed more than 48 h. We present a case of Boerhaave's syndrome in a 35-year-old male who presented with spontaneous respiratory distress and hemodynamic instability, about 36 h after the onset of vigorous vomiting. The case was managed initially with endoscopic insertion of a self-expanding metallic stent, followed later by surgical closure of the esophageal perforation. The patient, however, developed post-operative septic complications and died after a week.

Key words: Boerhaave, Esophageal perforation, Mediastinitis, Pleural fluid amylase, Self-expanding stent

Boerhaave's syndrome, first described by Herman Boerhaave in 1729, is a rare but potentially fatal condition characterized by a spontaneous transmural tear occurring in a previously healthy esophagus [1,2]. The underlying pathophysiology is a sudden rise in esophageal intraluminal pressure secondary to forceful vomiting. The most common cause of esophageal perforations, however, is iatrogenic, in particular, endoscopic procedures, accounting for 70% of all cases, followed by trauma to the chest or neck [3]. Boerhaave syndrome is relatively rare and seen in only about 15% of cases, but is the most fatal, with a mortality of 30%. Mortality also varies according to the time of presentation, condition of the esophagus, and surgical modality.

CASE REPORT

A 35-year-old male patient, with no comorbidities, presented from another institution with a 36-h history of sudden onset of respiratory distress following 3–4 episodes of vigorous vomiting.

He was tachycardic and tachypneic on coming, with a blood pressure of 80/50 mm Hg and in severe respiratory distress with oxygen saturation of 80% with non-rebreather mask oxygen of 16 L/min. He was intubated and put on mechanical ventilation and started on intravenous fluid boluses, piperacillin-tazobactam 4.5 g 6th hourly, and noradrenaline infusion.

Chest radiograph showed a large right-sided pleural effusion, which was tapped using an underwater seal drain (UWSD), revealing a brownish, thick aspirate. Palpation revealed extensive subcutaneous crepitus around the neck. Computed tomography (CT) scan of the chest, abdomen, and pelvis showed bilateral pneumothorax, more on the left side, extensive subcutaneous and intramuscular emphysema in the neck spaces, and trace pneumomediastinum surrounding the esophagus (Fig. 1). There was no evidence of any focal rent, although a possible sealed-off perforation could not be excluded. Following this, a left-sided UWSD was inserted. Laboratory parameters were normal except for a total leukocyte count of 22,000 cells/mm^3 with 80% neutrophils, and pleural fluid amylase of 38206 U/L (normal up to 100U/L).

Esophagogastroduodenoscopy was done on suspicion of Boerhaave's syndrome which revealed a full-thickness, 4 cm length, and oval-shaped perforation of the esophageal wall about 1 cm above the gastric cardia (Fig. 2a). An 18×100 mm size self-expandable metallic stent (SEMS) was placed across the rent and secured with hemoclips (Fig. 2b). He was kept nil-per-oral postoperatively. His condition did not improve, with persisting respiratory distress and the addition of adrenaline and vasopressin support; hence, he was taken up for laparotomy. The SEMS was removed to facilitate the repair and the tear was closed with a monofilament 3.0 absorbable interrupted suture, reinforced with

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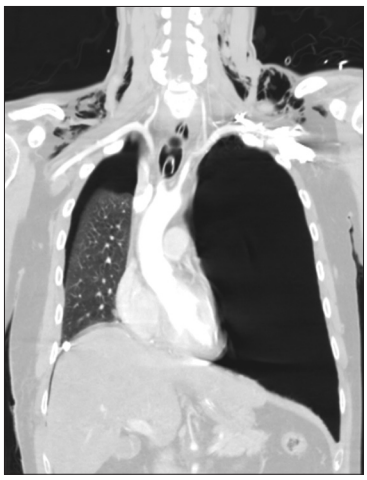

Figure 1: Computed tomography scan showing large left-sided pneumothorax and subcutaneous emphysema

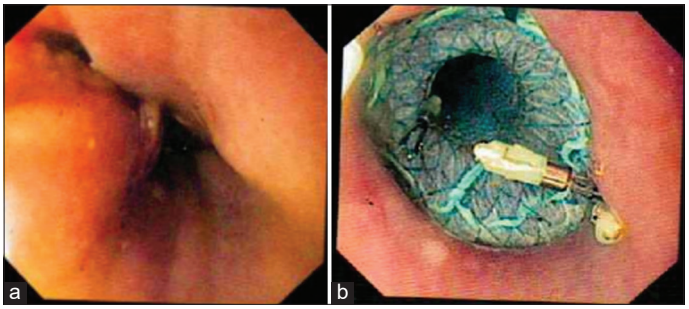

Figure 2: Esophagogastroduodenoscopy (a) full-thickness rent in esophageal wall 4 cm above cardia; (b) self-expandable metallic stent was utilized to seal the perforation

an omental patch, and gastric fundal patch. A feeding jejunostomy was placed, along with a drain near the esophageal hiatus.

During the post-operative period, he remained on vasopressor support including noradrenaline and vasopressin. Pleural fluid culture grew pan-drug resistant *Acinetobacter baumannii* and *Enterococcus faecium*, and deep endotracheal culture grew *Escherichia coli* and *Pseudomonas aeruginosa.* He was started on intravenous polymixin B 500,000 units twice daily, minocycline 100 mg twice daily, and fluconazole 200 mg once daily. The course was further complicated by severe acute respiratory distress syndrome, necessitating a fraction of inspired oxygen of 100% and difficulty in weaning down ventilator supports. On a post-operative day 6, he had multiple episodes of atrial fibrillation, and an episode of asystole on day 7, following which he could not be revived.

DISCUSSION

Boerhaave's syndrome is the spontaneous rupture of a healthy esophagus following an abrupt rise in intra-abdominal pressure, as seen in intensive vomiting or retching, weight lifting, parturition, status epilepticus, defecation, or the use of the Heimlich maneuver [2]. The rupture is usually (in 90% of cases) in the lower third of the esophagus and in the left lateral position, due to an anatomical weakness at that point [4-6]. The average tear is 2.2 cm long and 3–6 cm above the diaphragm [7]. The present case highlights the severity of illness and rapid deterioration associated with delayed diagnosis of esophageal perforation.

Chest pain, vomiting, and subcutaneous emphysema, which form the classical Mackler's triad suggestive of esophageal perforation, are seen in only about 50% of cases [3]. Most patients present with variable and non-specific symptoms such as vomiting, tachypnea, cough, and fever [4]. Esophageal perforation is itself a very rare condition, seen in only about three persons per million population, precluding physicians from obtaining clinical experience, and may lead to a missed diagnosis [2]. This condition is most commonly misdiagnosed as a perforated ulcer, followed by myocardial infarction, pulmonary embolism, dissecting aneurysm, and pancreatitis [6]. Other differential diagnoses of Boerhaave's syndrome include aortic dissection, perforated peptic ulcer, Mallory–Weiss syndrome, pneumonia, and spontaneous pneumothorax [1]. The anatomical location of the esophagus allows the entry of bacteria, digestive enzymes, and food particles through a perforation into the sterile environment of the mediastinum [3]. This leads to an early and rapid onset of complications such as mediastinitis, pneumothorax, empyema, septic shock, and multi-organ failure.

Early recognition of clinical features at presentation is essential [7]. Chest pain characterized by radiation to the back or left shoulder is the cardinal symptom of this condition. Neck pain may occur when the cervical esophagus is involved, in association with dysphonia, hoarseness, and dysphagia. Subcutaneous crepitus suggestive of subcutaneous emphysema may be seen in about 60% of perforations but may take about 1 h to develop following the injury.

Clinical examination should be followed up by a radiological work-up for a definitive diagnosis. Chest radiographs may show a pneumothorax in about 77% of cases and mainly on the left side. Other common signs include pleural effusion, pneumomediastinum, mediastinal air-fluid level, subcutaneous emphysema, hydrothorax, pneumothorax, and lung atelectasis. Any sign of esophageal perforation should be followed up by a contrast esophagogram using a water-soluble oral contrast, which may show a contrast leak. Contrast esophagogram is the diagnostic procedure of choice and is more sensitive, with a definitive diagnosis being reached in 17 out of 22 patients by this method [3]. An alternative is a contrast-enhanced CT scan of the chest and abdomen which may also show a leak or other suggestive findings such as mediastinal fluid, pleural effusions, pneumomediastinum, and dilated esophagus. In the present case, although the CT scan confirmed a pneumomediastinum, there was no obvious esophageal rupture. Upper endoscopy is useful in case of suspected perforations with negative radiography, or when a contrast esophagogram is contraindicated, and also in localizing the site of perforation. Diagnosis may also be confirmed by a pleural fluid analysis showing the presence of food particles, a high amylase content, and a low pH.

Patients vary widely in age as well as in the severity of illness, which precludes them from adopting any single approach to management. Initial treatment involves cessation of oral intake, administration of intravenous fluids, proton pump inhibitors or H2 blockers, appropriate broad-spectrum antibiotics, and parenteral nutrition, in an intensive care unit. Following this, a decision about surgical versus non-operative management has to be reached.

Non-operative treatment involves endoluminal placement of a SEMS, functional isolation of the esophagus and stomach using draining nasogastric and feeding nasojejunal or jejunostomy tubes, and eventual mediastinal, pleural, or abscess drainage [1,3]. This can be offered to selected, non-septic patients with a small or well-contained perforation, in cases of inoperable malignant strictures or when the presentation is delayed 5 days or more. The success of endoscopic stenting in Boerhaave's syndrome is center-dependent and may carry side effects such as worsening mediastinal or pleural contamination or stent dislodgement leading to pyloric obstruction. Stenting is a temporary measure, as the majority of patients will require mediastinal drainage by opening the parietal pleura along the entire length of the esophagus and debridement of necrotic and contaminated tissue [2]. The time for stent removal is unclear, but ideally requires complete clinical and radiographic resolution, which may take 6 weeks–3 months. Another measure is the insertion of a nasogastric tube down to the level of perforation to be used as a suction-rinsing drain with physiologic saline or antibiotic solutions [4]. A major risk following these conservative measures is the development of abscesses which may lead to deterioration.

Surgery is preferred for patients who present within 24 h, or with sepsis, large uncontained leaks, and extensive contamination, or for any patient managed conservatively who develops septic complications [2,3,5,7,8]. Delayed presentation leads to increased tissue necrosis and edema, which may prevent successful repair. Primary esophageal repair involves a single layer of interrupted absorbable sutures and may be opted for when the rupture is <3 cm and presentation is <24 h. Reinforcement with vascularized tissue decreases fistula formation and mortality, compared to repair without reinforcement. If the tissue is friable and cannot be directly closed, tear closure should be done using pleural, omental, or intercostal muscle flaps over the defects. An alternative is to divert the esophageal secretions using a T-tube forming an esophagocutaneous fistula which will allow for healing to take place without contamination. The T-tube can be removed in about 4–6 weeks, following which the fistula will close spontaneously [9]. Esophagectomy and reconstruction are the best choices when there is a phlegmon or intractable obstruction, perforation of a diseased esophagus, or severe injury involving an extensive length of the esophagus.

A contrast study should be obtained on the 5th post-operative day to document the integrity of the repair. Continued esophageal leakage has been seen to occur in 30% of patients, necessitating additional procedures [3,10]. Oral feeds can be started in about 9 days in patients treated conservatively, and in 25 days in surgical patients.

CONCLUSION

Boerhaave's syndrome is a rare condition that may mimic common thoracic or abdominal disorders and requires a high degree of suspicion for diagnosis. Any history of recurrent and forceful vomiting before the onset of chest pain should warrant investigations into an esophageal rupture. Early recognition of symptoms, diagnosis, and treatment is key to avoiding morbidity and mortality.

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