introduction

Esophageal perforations are rare with an incidence of 3.1 per 1,000,000 per year [1]. Boerhaave’s syndrome is found in approximately 15% of perforations. It is an extremely rare condition that is associated with a high rate of morbidity and mortality, depending on the location of the rupture, the degree of leakage and the time elapsed since the injury [2,3]. The key to successful management of Boerhaave’s syndrome is early diagnosis, aggressive medical treatment and definitive surgical management. We describe a patient who presented with rupture of both thoracic and abdominal portions of the esophagus due to severe pyloric stenosis caused by a longstanding duodenal ulcer. The patient was managed with a single surgical procedure.

Case Presentation

In our manuscript, we report the case of a 48-year-old gentleman with a history of peptic ulcer disease of 4 years duration who was admitted with epigastric pain of sudden onset radiating to the back, following episodes of vomiting three days prior to presentation. Patient was not compliant on his medications which included proton pump inhibitors.

The patient’s vital signs revealed a HR of 90 bpm, BP of 130/85 mmHg, RR of 20 and a temperature of 36.5°C. Physical examination revealed abdominal distension and epigastric tenderness. Lab test results showed a WBC of 18500 cells/mcL and Hb of 15 g/dL. His ECG revealed no abnormalities. Initial chest X-ray showed a normal study, however the patient refused any further investigations at that time.

Eight hours after hospitalization, the patient developed severe back pain associated with tachycardia and dyspnea. His vital signs at this time were HR 120 bpm, BP 120/80 mmHg, RR 28 and temperature of 37°C. Arterial blood gas with three liters of oxygen showed a pH of 7.7, PCO2 4.8 kps, PO2 18 kps, HCO3 16.5 and SaO2 95%.

An urgent enhanced computed tomography (CT) scan of the chest and abdomen was done and showed a huge right-sided hydro pneumothorax, left-sided pleural effusion and pneumomediastinum. An esophageal tear was suspected (Figure 1). The stomach was markedly distended with absence of distal opacification of the oral contrast and the presence of a perigastric collection (Figure 2).

Patient initially refused to proceed for surgery so bilateral 28 F chest tubes were inserted, with 1.5 L and 1.2 L of turbid fluid drained from the right and the left pleural cavity respectively. The next day patient’s general condition deteriorated with increasing abdominal distension and guarding, his HR increased to 140 bpm and his oxygen saturation dropped to 92% on 3 L nasal oxygen. At this point the patient agreed to undergo emergency surgery, approximately 24 hours after his initial presentation.
As the underlying etiology was abdominal in origin (pyloric stenosis due to duodenal ulcer), surgery was performed using both thoracic and abdominal approaches using two separate incisions.

1. Left thoracotomy
   About 300 ml of turbid fluid with food residue was evacuated. The mediastinal pleura was identified and opened. The esophageal tear was visualized in the lower third of the thoracic esophagus extending 5 to 6 cm down to the abdominal esophagus. The ruptured edges of esophagus were inspected and devitalized tissue was debrided. The esophageal mucosa was then identified and repaired primarily in two layers. We also reinforced the repair using a pleural flap to buttress the suture line. The surgery was completed with copious irrigation and bilateral drainage of the pleural cavities with two chest tubes size 28 F.

2. Midline laparotomy
   Once intraperitoneal access was obtained, l liter of contaminated turbid fluid was drained. Irrigation and peritoneal toileting was performed. The pyloric stenosis was visualized, with no extrinsic compression, so the stenosis was bypassed with a loop gastrojejunostomy.

   The patient was kept on mechanical ventilation for 4 days postoperatively. An esophagogram was done on day 7 post-op, which revealed no leak at the gastroesophageal junction (Figure 3) and confirmed a patent gastrojejunostomy anastomosis (Figure 4).

   The patient’s feeding regimen was gradually progressed from fluid to regular diet and he was eventually discharged on day 19 in a good medical condition.

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Figure 1. Showing diffuse esophageal wall thickening with periesophageal air and bilateral pleural effusions.

Figure 2. Distended stomach with perigastric fluid collection.

Figure 3. Follow-up barium swallow showing no extravasation on the distal esophagus and at the gastroesophageal junction.

Figure 4. Barium meal showing a good drainage and absence of leak at the gastrojejunostomy.
DISCUSSION

Spontaneous esophageal perforation occurs as a result of an abrupt rise in the intraesophageal pressure combined with negative intrathoracic pressure due to intense bouts of vomiting [3-5]. The classical presentation of Boerhaave’s syndrome is manifested by Mackler’s triad: 1) forceful vomiting, 2) lower chest pain and 3) subcutaneous emphysema; however mediastinitis, sepsis and septic shock are frequently seen in late presentations [2-5]. It is a serious injury that is associated with a high mortality rate ranging from 20-30% and reaching a rate of up to 100% if not treated [2-4]. The most common site of rupture is in the lower third of esophagus, more so on the left side [2-5]. History of heavy drinking and gastroduodenal ulcers are primary risk factors [2-4]; nevertheless, few cases in the literature have reported an association with pyloric stenosis.

Upright chest radiography is useful in the initial diagnosis as an abnormal finding is found in 90% of patients after perforation [2]. However, 10% of patients may not have any findings on chest X-rays, which is due to the delayed radiographic development of mediastinal and subcutaneous emphysema, as was true in our case.

Contrast esophagogram using water soluble agents such as gastrografin usually shows extravasation of contrast into pleural cavity. The localization of the injury and its length usually aids operative planning when deciding the best surgical approach [2-5]. This investigation has a sensitivity of 80% in detecting esophageal perforations [2,5].

In the case of a negative contrast study, where clinical suspicion remains high, a barium swallow can be used as it can detect up to 90% of the intrathoracic perforations [2]. During this study the patient should lie on the ipsilateral side where the chest pain occurs, to allow the barium material to drain throughout the perforation, otherwise a small leak may be missed [2-5].

CT scan is useful when perforations are difficult to identify or localize or when contrast esophagography cannot be performed. It may show esophageal wall edema and thickening, periesophageal fluid with or without gas bubbles, mediastinal widening and fluid in the pleural space, retroperitoneal and lesser sac [2-5].

Endoscopy is not routinely performed since it carries an additional risk of increasing the size of the original perforation [2].

Boerhaave’s syndrome is rare and there is limited evidence to guide management but according to the literature available it is divided into initial and subsequent management [7].

- Initial management

Its goal is to stabilize and prepare the patient for definitive management.

The following principles are applied to the initial treatment of a patient with an esophageal perforation:

i. Rapid diagnosis
ii. Appropriate hemodynamic monitoring and support
iii. Avoidance of all oral intake, ensuring optimal nutrition, typically parenterally
iv. Adequate broad spectrum antibiotic therapy
v. Intravenous proton pump inhibitor
vi. Control of extraluminal contamination by adequate drainage if present.

- Subsequent management

This approach is presented in an algorithm (Figure 5). It can be medical, surgical or endoscopic and usually depends upon multiple factors:

i. Patient’s condition and associated comorbidities
ii. Response to the initial management
iii. Size and location of the rupture
iv. Type of leak (contained to the mediastinum or spread to the pleura or pericardium)
v. Presence of underlying esophageal pathology.

Medical management: Candidates for medical management include the following [8]:

1. The leak is contained within the neck or mediastinum or between the mediastinum and visceral lung pleura.
2. Contrast is able to flow back into the esophagus from the cavity surrounding the perforation.
3. The injury is not in neoplastic tissue, is not in the abdomen, and is not proximal to an obstruction.
4. The patient has minimal symptoms.
5. Signs and symptoms of sepsis are absent.
6. Access to contrast studies can be obtained at any time of day.
7. An experienced thoracic surgeon is readily available if the patient deteriorates.

Surgical management: Patients who fail conservative attempts at treatment and show signs of clinical deterioration during medical management require surgical intervention [9]. Surgery is indicated in patients who are being managed medically if any of the following develop [8,9]:

a. A perforation that initially had limited extravasation of contrast develops free diffuse extravasation.
b. Extension of the perforation.
c. Clinical deterioration, persistent fevers or sepsis (if a collection that is amenable to percutaneous drainage is present then attempted drainage of the collection prior to proceeding with surgery is reasonable).
d. Progression of pneumomediastinum or pneumothorax.
e. Development of an empyema.

Surgical options can include primary surgical repair of the defect, resection of the defect, diversion, drainage of collections, or in some cases, esophagectomy.

Primary repair remains the gold standard and when performed by an experienced surgeon good outcomes can be obtained. General principles for esophageal repair [10]:

1. Debridement of devitalized tissue from the perforation site.
2. Exposure of the entire extent of the mucosal...
injury, even by incising the muscular layer longitudinally if needed.

3. A two-layer repair of the mucosa and muscularis, with meticulous approximation and avoidance of luminal narrowing.

Early diagnosis and treatment can improve the prognosis [3,4]. Delayed surgical intervention beyond 24 hours can result in high mortality [11]. Operative treatment is based on immediate intervention using a thoracic, abdominal or thoraco-abdominal approach with closure of the perforation.

**Endoscopic management:** Endoscopic treatment of an esophageal perforation should be considered in patients with extensive underlying comorbidities who are unlikely to tolerate surgery [12]. Endoscopic therapy should be performed in close conjunction with a thoracic surgeon. Outcomes following endoscopic therapy for Boerhaave’s syndrome have not been compared with surgery in randomized trials, but observational studies suggest that a significant proportion of patients treated with endoscopic therapy require re-intervention and surgery.

A retrospective study compared clinical outcomes in 20 patients who underwent surgery with 13 patients who underwent endoscopic stenting for Boerhaave’s syndrome. In this study, there was no difference in intensive care unit or hospital length of stay between the two groups. However, patients treated endoscopically had a higher rate of mortality and 11 of 13 patients treated with endoscopic stents required operative intervention [12]. Both self-expandable metal stents (SEMS) and self-expandable plastic stents (SEPS) have been used to treat esophageal perforations [13,14]. While stent migration rates are higher with plastic as compared with metal stents, metal stents have a significantly higher incidence of post-procedure strictures [12]. Stent removal is advised within six weeks of placement [15].

Minimally invasive therapy of perforations at the esophagogastric junction using over-the-scope clipping without stenting has also been described. Esophageal defects have been closed with endoclips, fibrin glue or metal stents. A combination approach utilizing over-the-scope clips has been used to approximate the sides of the defect, followed by placement of a fully covered stent positioned over the defect and across the gastroesophageal junction and released under endoscopic guidance [16]. Fibrin sealant has also been used in the treatment of a longstanding esophagobronchial fistula resulting from Boerhaave’s syndrome [17].

Although treatment of delayed presentations remains con-

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**Figure 5. Management of esophageal perforation**

CT: computed tomography  NPO: nothing by mouth  NG: nasogastric  IV: intravenous  TPN: total parenteral nutrition  SEMS: self-expandable metallic stents  *Endoscopy should only be performed in centers of expertise.  **Depending upon the type, location, severity of the esophageal disease, and size of the perforation.
troversial, primary repair of the perforation site is the optimal procedure. The exceptions to performing a primary repair include a cervical perforation that cannot be accessed but can be drained, diffuse mediastinal necrosis, a perforation too large for the esophagus to be re-approximated, an esophageal malignancy, preexisting end-stage benign esophageal disease (e.g. achalasia) or if the patient is clinically unstable [18-20]. When there has been a delay in diagnosis greater than 24 hours, and/or substantial extraluminal contamination from the leakage of fluid and debris has occurred, the integrity of the repair can be enhanced with the use of a flap from the serratus muscle, latissimus dorsi, diaphragm, parietal pleura, omentum, and gastric fundus [21].

We operated on this patient using a lateral thoracotomy and midline laparotomy in two separate incisions. This allowed us to maintain the physiological function of the diaphragm, prevent thoracic contamination and reduce the risk of postoperative pulmonary sequelae and other complications [22].

According to our literature review, this is the first case report of Boerhaave’s syndrome with pyloric stenosis that was managed using a single-stage procedure with primary repair of the esophageal rupture combined with gastrojejunostomy, avoiding late distal gastrectomy and gastrojejunostomy reconstruction Billroth II.

CONCLUSION

Spontaneous esophageal perforation secondary to pyloric stenosis remains a rare finding in a rare disease with enormous diagnostic and therapeutic challenge. Early diagnosis and management can reduce its high mortality and morbidity rate. In patients with Boerhaave’s syndrome and pyloric stenosis due to duodenal ulcer disease, a single-stage procedure can be considered as a surgical option in selected patients.

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