CASE REPORT

A rare case of Boerhaave's Syndrome

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ABSTRACT

Boerhaave's syndrome is a rare condition but potentially fatal condition characterized by a transmural tear of the distal esophagus induced by a sudden increase in intra luminal pressure. Diagnosis is challenging and usually delayed as the classical triad of vomiting, abdominal or chest pain, and subcutaneous emphysema is absent in many patients. Treatment has not been standardized and may be conservative, endoscopic or surgical.

We present a typical case which illustrates possible diagnostic pitfalls and the therapeutic conundrum surrounding management of the syndrome. Based on time of presentation and eventual presence of sepsis, a therapeutic algorithm is proposed

Keywords : Oesophagus; Tear; Boerhaave's Syndrome; Diagnosis Treatment

Introduction:

Originally described in 1724 - Dutch physician Hermann Boerhaave refers to an esophageal tear caused by an abrupt rise of intraluminal pressure [1]. It is relatively rare but life-threatening condition that requires urgent diagnosis and treatment. Delay in treatment results in severe and potentially lethal complications such as mediastinitis, empyema, septic shock, and multiple organ failure. A high index of suspicion is imperative for timely diagnosis and to utilize well- selected radiological and endoscopic investigations. Prompt adequate therapy reduces morbidity and mortality. Some centers advocate early and extensive surgery as the cornerstone of treatment, yet others prefer a more conservative, endoscopic, or less invasive surgical approach. Such experience-based difference in attitude likely explains the lack of well-established treatment guidelines.

Case Report

A 40 yr old female presented to our emergency department with an inter costal drainage tube (ICD) in her left hemi thorax draining

seropurulent discharge. She gave history of having suffered three episodes of projectile vomiting on full stomach 24 hours ago. The vomiting was followed by chest pain and shortness of breath. She sought medical help at a peripheral hospital where she was diagnosed to be suffering from hydropneumothorax left and surgical emphysema, and was treated with left sided tube thoracostomy and was referred to our hospital

On arrival, she had tachypnoea, tachycardia requiring oxygen support to maintain saturation. She also had subcutaneous emphysema extending from neck to upper abdomen. Abdomen examination was unremarkable. Laboratory investigations suggested neutrophilic leukocytosis and arterial blood gas revealed respiratory acidosis.

Computed Topography of neck, chest & upper abdomen with oral contrast (Figure 1) showed extensive subcutaneous emphysema, Pneumo mediastinum, Pneumo thorax, bilateral pleural effusion & left ICD in situ. There was oral contrast leak in distal 1/3 rd esophagus proximal to Gastro esophageal junction (Fig. 1).

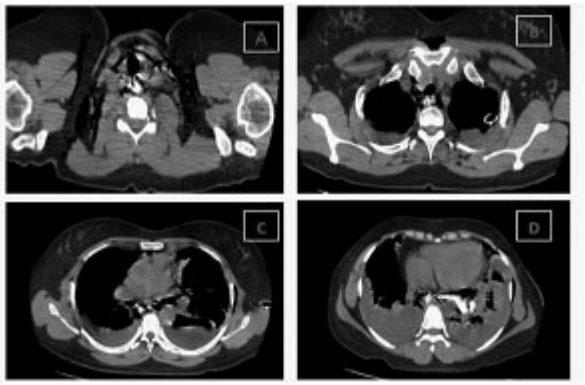

Figure 1 A: Subcutaneous emphysema B: Pneumomediastinum C: Pneumothorax, Bilateral pleural effusion, Left ICD In situ D: Oral contrast leak (distal 1/3 rd esophagus)

We arrived at a diagnosis of Boerhaave's syndrome. Blood cultures were taken; after ample fluid resuscitation and appropriate intravenous antibiotic administration, she was taken up for explorative laparotomy with option to convert to right lateral thoracostomy.

Laparotomy revealed presence of purulent fluid in the region of diaphragmatic hiatus. Following the suctioning of the pus, perforation of the size 2X1 CM was found at oesophago -gastric Junction (EGJ) on left side. The margins of the perforation were healthy (Fig. 2).

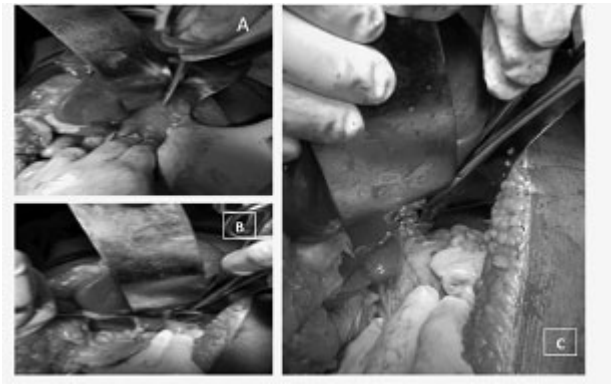

Figure 2 A: Sero-purulent material on opening the hiatus B: Perforation site showing Ryles tube C: Closure with interrupted sutures (3-0 vicryl)

After thorough lavage, margins of perforation was freshened and closed primarily and buttressed with fundus of stomach. A feeding jejunostomy (FJ) was done. Right intercostals drainage tube was also kept to drain right pleural cavity. In the immediate postoperative period the patient was on ventilatory support and was extubated on the 3rd post operative day. She was started on FJ feeds on POD 2. And she tolerated the FG feeds well. On POD 10, the patient developed episodes of intermittent high fever. Imaging with contrast swallow, revealed leak at the site of primary repair and contained collection nearby. About 100 ml

of purulent collection was drained by repositioning the existing drain. Patient recovered well following this and was discharged on POD 14 with drain and FJ in situ. In the follow up after a week, drain was removed and oral feeds were started after confirming no leak from the previous site.

Discussion

Boerhaave's syndrome is a of barogenic tear of the distal esophagus caused by sudden rise in the intraluminal pressure. It accounts for approximately 15% of all cases of esophageal rupture and has a mortality rate up to 40%. In majority of cases, the rupture is located in the left posterolateral wall of the distal third of the esophagus due to anatomic weakness at the junction of clasp and sling fibers at EGJ [2, 3]

This syndrome should be readily suspected in a patient with a history of severe repeated bouts of vomiting, experiencing excruciating chest pain and developing subcutaneous emphysema [4]. This Mackler's triad is seen only in 14% of patients. Hamman's sign (i.e., a mediastinal "crackling" accompanying every heart beat) may be heard in left lateral decubitus position in the presence of mediastinal emphysema [2]

However, up to one-third of patients have atypical symptoms or are admitted with severe respiratory distress and/or shock. The sequence of events following rupture includes esophageal and gastric contents getting sucked into mediastinum by negative intrathoracic pressure, which causes chemical burn by gastric juice. Further super-imposed necrotizing infection due to digestive enzymes and oral bacteria with rapid tissue destruction results in severe sepsis.

Chest X ray reveals classic signs like mediastinal emphysema and mediastinal widening, which take several hours to develop and is seen in only 12- 15% of patients [5]. CT scan of thorax and mediastinum has largely replaced the former Chest X rays and gastrografin swallow tests due to its sensitivity in detecting even small amounts of extra luminal air, ?uid collections, paraspinal or pleural air-fluid collections [7]. Endoscopy aids in direct visualization of tear but its role is controversial as it may potentially aggravate the esophageal tear.

Management of Boerhaave's syndrome remains a controversial issue. Basically, three methods of treatment are distinguished: a conservative, an endoscopic, and a surgical approach [8, 9]. Conservative management involves a small highly select set of patients as coined by Cameron (1970) which include, minimal clinical sepsis, disruption contained in the mediastinum, drainage of the cavity back into esophagus [10]. It essentially includes restriction of oral intake, nasogastric tube insertion, parenteral antibiotics, proton pump inhibitors, and percutaneous drainage of mediastinal or pleural abscess in indicated cases.

Endoscopic therapy is increasingly used

nowadays in patients who are diagnosed early without widespread contamination and sepsis. Endoluminal placement of a self-expandable metallic stent to bridge an esophageal tear has shown encouraging results. But endoscopic stenting in Boerhaave's syndrome has a rather undetermined and center-dependent success rate. It is not without any side effects such as enhanced mediastinal or pleural contamination, reflux and accidental stent migration [11].

Surgical treatment is considered the gold standard and outcomes are best if identified and intervened within 24 hours of onset. Primary surgical intervention is required when patients present with sepsis, large uncontained leaks, and extensive contamination and in those who deteriorate during conservative trial of management [12, 13]. Factors determining surgical intervention include extent of the perforation, eventual concomitant pathologies that require synchronous management, degree of mediastinal or pleural contamination, the extent of sepsis and patient's co morbidities.

It ranges from Primary closure with or without reinforcement, drainage, exclusion and diversion or oesophagectomy. [8,9,14]. Primary repair by tension-free apposition of healthy mucosal and sub mucosal tissue along with reinforcement using autologous tissue like

intercostal muscle, pleural or omental flap is most commonly employed. The approach can be via thoracotomy or laparotomy depending upon the level and extent of injury.

Simple lavage and drainage is done when direct repair is thought to have high chance of leakage and contamination in extremely sick patient without extensive esophageal injury. In some patients who presented late with sepsis, heavy mediastinal contamination having devitalized esophageal tissue, repair may be impossible. In these patients bipolar exclusion was done isolating thoracic esophagus with cervical oesophagostomy and ligation of the cardia, preventing reflux of gastric content.

oesophagectomy is not done routinely and has high morbidity in emergency conditions. However, in the presence of necrotized esophageal tissue beyond salvage or when underlying esophageal pathology is suspected, oesophagectomy should be done. Minimally invasive surgeries either by thoracoscopy or laparoscopy are evolving. The results of both laparoscopic and open approach are comparable with less postoperative pain and early recovery in the former. Haveman et al. compared open thoracotomy with video-assisted thoracoscopic surgery (VATS) and found similar success rates [15]. However, more prospective studies are warranted to establish the potential advantage of VATS in treating patients with Boerhaave's syndrome.

Mortality rate varies from 25-89%. In Post emetic perforation, it is found that mortality rates are less than 25% if intervened within 24 hours, whereas it steeply increases to around 85% if operated more than 48 hours later. [16,17]. This

underscores the importance of early diagnosis and prompt management.

Conclusion

Boerhaave's syndrome still represents a diagnostic and therapeutic challenge. It is due to rarity of disease, with only retrospective case series, case reports, and expert opinions available. Multiple treatment options and operative strategies are available and have to be individualized according to patient. Early recognition and prompt treatment are needed to maximize survival

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