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Boerhaave Syndrome

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ABSTRACT

Boerhaave syndrome (BS) is an uncommon and life-threatening disorder and is defined as spontaneous transmural esophageal perforation, which typically occurs after a projectile vomiting. The prognosis is dependent on early detection of the disease and appropriate management within 12 h of perforation. The typical presentations of BS are vomiting, subcutaneous emphysema, and pain in the lower chest. However, severe symptoms rarely happen, and about one-third of all patients present with atypical clinical features. Therefore, BS should be suspected in any patient presenting with sudden thoracoabdominal pain with a history of vomiting. Chest X-ray is the most helpful diagnostic tool, and computed tomography scan is applied for further evaluations. Although surgical treatment is an important option for many patients, a less invasive procedure with or without endoscopic stent or internal or external drains, with respect to patient's clinical condition, is preferable.

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Introduction

Boerhaave syndrome (BS) was first described by a Dutch physician and anatomist, Hermann Boerhaave, in 1724. His patient was a 50-year-old Dutch grand admiral, Baron Janvan Wassenaer, who died in 1723 after one hour of self-induced vomiting, which led to esophageal perforation. BS is one of the most life threatening diseases of the gastrointestinal tract with high mortality and morbidity rates (1-3).

The pathophysiology of BS involves a transmural esophageal perforation ensuing a sudden rise in intraluminal esophageal pressure due to vomiting against a closed glottis and failure of relaxation of the cricopharyngeal. The most common anatomical location of the rupture is at the left posterolateral wall of the distal esophagus, 2-3 cm proximal to the gastroesophageal junction. Moreover, the second common location of rupture is at the subdiaphragmatic or upper thoracic esophageal segments (4-8).

This condition usually occurs following excessive alcohol consumption or overeating, both of which can lead to vomiting. Because BS is an uncommon condition, misdiagnosis can result in mediastinitis, sepsis and even shock. Timely diagnosis and effective management within 24 h reduce mortality rate by approximately 30%.

Without adequate treatment, BS survival is in days and the mortality rate approximates to 100% (9-13).

There are multiple factors deteriorating BS mortality rate, such as difficulty in accessing the esophagus, lack of a strong serosal layer, abnormality in blood supply, and proximity to the vital structures. Generally, BS mortality rate is approximately 30%, which is mainly due to subsequent infections including mediastinitis, pneumonitis, pericarditis, or empyema (14-16).

Epidemiology

According to the literature, this disorder is reported in all races. The annual incidence rate of esophageal perforations is low, approximately 3.1 per 1,000,000 individuals, and 15% of all cases have BS. Globally, BS is significantly prevalent in males, with male to female ratio of 2:1 to 5:1. BS is more common in the patients

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aged between 50 and 70 years. Although several studies demonstrated that 80% of all patients are middle-aged males, this syndrome is reported in neonates and in people older than 90 years, as well (17-21).

Clinical Manifestations

BS typically presents with a history of nausea and vomiting followed by a sudden onset of severe lower chest and epigastric pain. The pain may radiate to the back or left shoulder and often worsens after eating. Another common manifestation is shortness of breath due to pleurisy or a pleural effusion. Mackler's triad denotes the classic presentations of BS including vomiting, lower chest pain, and subcutaneous emphysema. Additionally, atypical findings of BS include hoarseness due to the involvement of recurrent laryngeal nerve and neck or upper chest pain in patients with cervical esophagus perforation (23-27).

Imaging and work-up

The diagnosis of BS relies on radiographic evidence.

Plain radiograph

Chest X-ray (CXR) findings are often non-specific and can be observed on a posteroanterior and lateral views. CXR findings of BS include pneumomediastinum, subcutaneous emphysema, left pleural effusion, and left pneumothorax. Left plural infusion is one of the most common findings, and about 10% of CXRs are normal.

In 20% of patients Naclerio's V sign, which is a focal and sharply marginated region of paraspinal radiolucency on the left side immediately above the diaphragm (Figure 1).

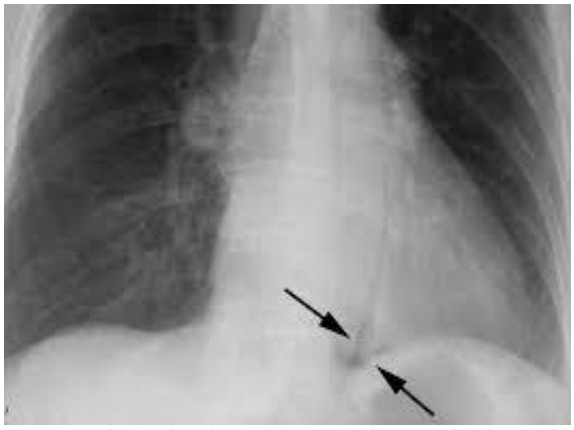

Figure 1. The Naclerio's V sign seen on the frontal radiographs of the chest as a V-shaped air lucency in the left lower mediastinal segment

Esophagography

Esophagography is an imaging technique that uses water-soluble contrast agents and often demonstrates a contrast leakage at supradiaphragmatic level in most esophageal perforation cases (Figure 2). Water-soluble contrast is utilized to prevent barium-related severe mediastinitis in patients with esophageal perforation, hence reduced morbidity and mortality. More than 20% of patients have reported false negative results; therefore, if clinical suspicion remains, Esophagography should be repeated after 4-6 h (27-30).

CT scan

An unenhanced CT scan reveals the presence of typical localization and periesophageal air collections indicating esophageal perforation. Post-contrast CT scan may show direct contrast extravasation and thickened esophageal walls. Other reported findings include pneumothorax,

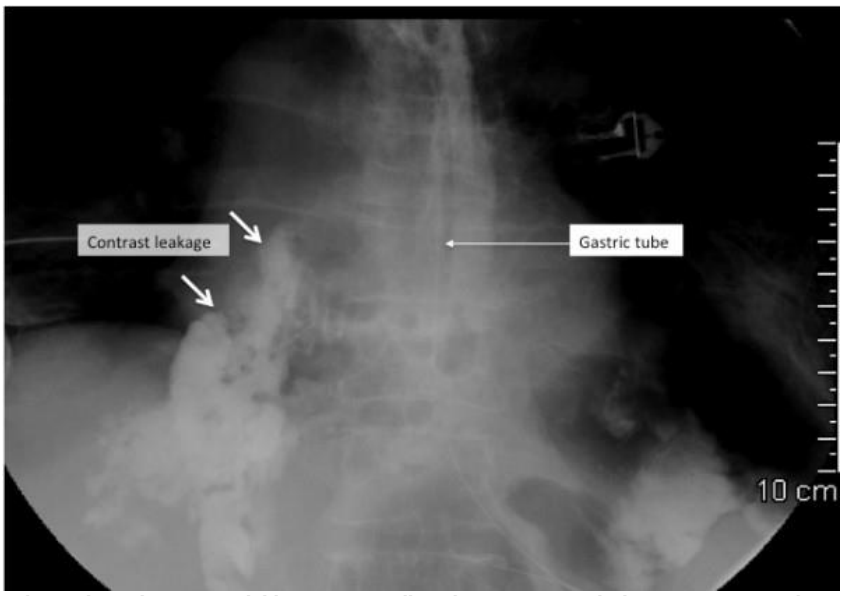

Figure 2. Plain chest radiography with a water-soluble contrast swallow showing contrast leakage in a patient with Boerhaave syndrome

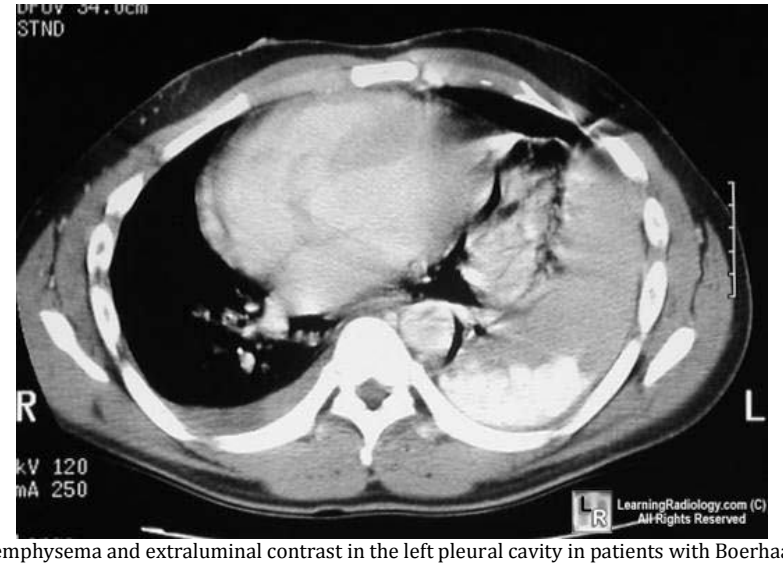

Figure 3. Mediastinal emphysema and extraluminal contrast in the left pleural cavity in patients with Boerhaave syndrome

pneumomediastinum, pleural effusion (usually left-sided), mediastinal fluid collections, and emphysema in the chest wall and neck (Figure 3). In comparison to CXR, CT scan better demonstrates the anatomical location and severity of perforation (31-34).

Endoscopy

There is controversy regarding the role of endoscopy in the diagnosis of BS. This procedure may raise the risk of extending the original perforation and forcing additional air through the perforation into the mediastinum. Endoscopy may be useful when a perforation is suspected, but radiographic findings are negative or it is impossible to use swallowing contrast agent (Figure 4) (35).

Treatment

Patients with suspected BS should be identified at an early stage and immediately treated. Appropriate management consists of both conservative and surgical interventions. Choosing between conservative and aggressive treatment modalities is dependent on the extent of perforation, patient's medical condition, and delay in evaluation and presentation.

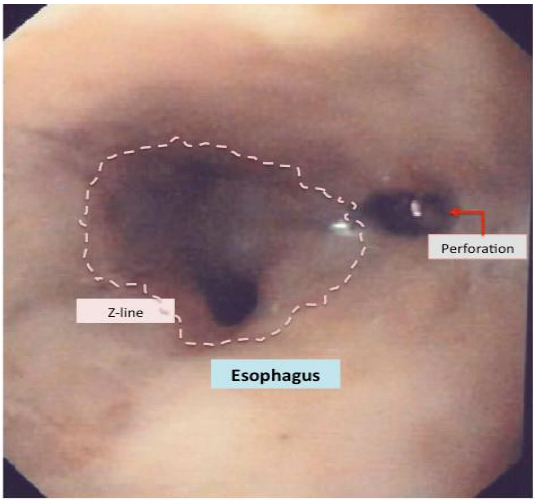

Figure 4. Endoscopic view of a distal spontaneous perforation 24 h after the presentation of clinical symptoms. However, endoscopic appearance may suggest a period exceeding at least 36 to 48 h from the presentation of symptoms

Medical care

In most patients, surgical management is the gold standard; nevertheless, non-invasive treatment might be convenient for the patients meeting the following criteria: A) The esophageal perforation is located in the mediastinum; B) There is a well drainage from the cavity into the esophagus; C) Patient presents with minimal symptoms; and D) There is minimal evidence of sepsis.

Conservative therapy includes intravenous fluids and broad-spectrum antibiotics (imipenem or cilastatin), nasogastric suction, proton pump inhibitors (PPIs), early use of nutritional support, and keeping the patient nil per os (NPO).

Surgical Care

If the diagnosis is made within the first 24 hours after perforation, primary repair is possible. Direct repair of the tear and drainage of the mediastinum and pleural cavity minimize the risk of further contamination and infection.

Surgical approach is determined by the location of esophageal perforation and surgeon's preference, and left thoracoabdominal incision is preferred. Omental flap is used for covering the primary closure. In addition, all the patients undergo laparotomy for gastrostomy and jejunostomy to help the drainage and nutrition.

Based on the study by de Schipper et al. (38), endoscopic procedure can be considered in certain patients. Endoscopy should be employed

for those patients diagnosed within 48 hours of the esophageal rupture without any signs of sepsis. However, if the patient presents signs of sepsis, thoracotomy with hemifundoplication and pleural/mediastinal drainage should be performed.

If diagnosis is made after 48 h of perforation, conservative therapy is recommended, and if septic profile is present, surgical treatment should be considered. According to the results of de Schipper et al., the survival rates of conservative, surgical, and endoscopic methods were 75%, 81%, and 100%, respectively (36-42).

Conclusion

The best outcome in BS is associated with early diagnosis and appropriate treatment. Due to the diverse range of presentations, the diagnosis is difficult and effective treatments are often delayed. Therefore, high index of suspicion for BS must be considered in any clinical condition in which the patient presents with a suspicious history with or without characteristic complaints.

Conflict of Interest

The authors declare no conflict of interest.

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