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Case report

# Transhiatal esophagectomy in Boerhaave syndrome – Case report and literature review

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ABSTRACT

*Introduction:* Boerhaave syndrome or spontaneous rupture of the esophagus wall is a rare life-threatening condition. It is more common in male gender and is due to a very swift rise in intraluminal pressure during vomiting. The patient usually presents with chest pain after vomiting. In some cases, there is subcutaneous emphysema in the neck or upper chest. Due to its rarity, the diagnosis is often not straightforward. Chest radiography can reveal pneumothorax, pleural effusion or pneumomediastinum, but diagnosis is more likely possible with an oral contrast X-ray study.

*Case presentation:* This paper reports a clinical case with surgical approach, in a 68-years old patient with a 48 h period between onset of symptoms and diagnosis of a Boerhaave syndrome. Firstly, the patient was admitted with a presumptive diagnosis of pneumonia. The patient was with chest pain, fever and vomiting. An emergent transhiatal esophagectomy was performed with primary anastomosis with no significant post-operative morbidity and allowing for the patient to return to previous daily routine with a good quality of life.

*Discussion and conclusion:* Boerhaave syndrome is a rare life-threatening surgical condition. Surgery is the most effective treatment. It is necessary to have a high index of suspicion. Treatment should promptly start because prognosis is related with time from diagnosis, with increasing mortality rate if no treatment is performed.

## 1. Introduction

Boerhaave syndrome (BS) is a condition characterized by a spontaneous barogenic rupture of the esophagus wall. It was described for the first time in 1724 by Dr. Herman Boerhaave, a Dutch anatomist and physician, and represents 15% of all esophageal perforations [1,2]. It has an estimated incidence of 1/6000, but has very high morbidity and mortality rates, the last reaching 40% [3,4,5]. The mortality is near 100% if no treatment is performed [6]. BS is more frequent in male gender, between 50 and 70 years old [1,3,7]. There are two common risk factors to BS: heavy meal ingestion and abundant alcohol consumption [3,5,6]. BS presents typically with chest pain and subcutaneous emphysema after vomiting [4]. It could present also with hematemesis (rare), dysphagia or respiratory distress [7,8]. Esophageal perforation could be easily missed during diagnosis, and more than a half of cases are misdiagnosis [4,9,10]. An oral contrast X-ray study is mandatory if there is suspicion of BS [5,8,11]. The chest radiograph is a simple exam and could show pneumothorax, hydropneumothorax, pneumomediastinum or subcutaneous emphysema in the neck or upper chest [5,8,10]. A water-soluble contrast, like gastrografin, could be administrated to confirm the diagnosis [8]. Prompt diagnosis and high index of suspicion, with an early therapeutic intervention are important factors for a good prognosis [4,6,11]. Delayed diagnosis results in severe mediastinal infection, sepsis and death. BS management needs to be adapted to patient presentation, lesion extension, time from diagnosis and viability of esophageal wall [5,9].It can imply a conservative, endoscopic or operative management [11–15]. Conditions needed for conservative management are: early diagnosis (<24 h), absence of symptoms or sepsis, contained cervical or thoracic perforation, no pre-existent esophageal disease and a close surveillance by expert team [11].

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All other situations should prompt a surgical approach. Esophagectomy is needed if there is an important esophageal wall destruction [16]. This patient was treated in an academic hospital.

This work is in line with the SCARE 2020 criteria [20].

## 2. Case report

A 68-year-old Caucasian male was admitted to the Emergency Department with a 2 h onset of chest pain, on the left anterior and posterior hemithorax. The pain was severe and started after an episode of vomiting. Previous medical history included hypertension, hyperuricemia and a Mallory-Weiss syndrome some years before, whose treatment only required upper gastrointestinal endoscopy. On physical examination, arterial blood pressure was 144/78 mmHg, cardiac frequency 108/min, peripheral oxygen saturation 90% and temperature 37,7 °C. His cardiopulmonary auscultation revealed decreased respiratory sounds in both inferior pulmonary areas. There were no changes on abdominal examination. The arterial blood gases evaluation revealed pH 7.45, pO2 57.2 mmHg, pCO2 36.2 mmHg, HCO3–36.4 mEq/L and O2 saturation 90.4%. The chest radiography showed a pulmonary opacity on the left (Figs. 1 and 2). Blood samples revealed mild increase in white blood cell count 10.200/$mm^3$. Electrocardiography and high sensitivity troponin I were normal and ruled out myocardial infarction. The patient was admitted to the Internal Medicine Department with a presumptive diagnosis of pneumonia, started intravenous antibiotic and oxygen. After 48 h, the patient's condition deteriorated with severe chest pain and respiratory distress. The arterial blood gas sample showed a type 1 respiratory failure and blood tests showed an increase of white blood cell count, c-reactive protein and d-dimer. A chest computed tomographic angiography was performed to rule out pulmonary embolism, but instead it showed an important pneumomediastinum that was close to the left-posterior wall of the esophagus and an hydropneumothorax on the left chest, and a destruction of the lower esophagus, suggesting a spontaneous rupture of the esophagus (Figs. 3 and 4). The patient was evaluated by a surgical team that decided to perform an emergent transhiatal esophagectomy with primary anastomosis and left thoracic drainage. Intraoperative findings revealed esophageal perforation with no peritonitis but with pleural effusion. The surgery was performed by an experient surgeon in esophageal surgery The specimen revealed an esophageal perforation (Fig. 5). The anastomosis was done in the neck in a latero-lateral fashion, using a linear stapler. A nasojejunal tube was left in place to allow post-operative early enteral nutrition. There were no complications during surgery. The patient was then transfered to the intensive care unit, where he stayed for 8 days. After an oral swallow X-ray revealed no anastomotic leak, a clear liquid diet was started on the 6th day after surgery. He was discharged home on the 21st day after surgery due to the need for post-operative rehabilition. The patient was evaluated in an appointment 1 month after discharge, where he underwent another oral contrast esophagogram that did not reveal any anastomotic leak or stenosis (Fig. 6). On follow-up appointments the patient is doing well, with no major upsets in his daily life. One year later, the patient was asymptomatic.

## 3. Discussion

Boerhaave syndrome or spontaneous rupture of the esophagus is a rare life-threatening surgical condition with high morbidity and mortality rates [2,3,8].

It commonly happens on the left posterior aspect of the distal esophagus (in 90% of cases), 2–3 cm above gastroesophageal junction [2,3,9].This area has a parietal sheet weakness due angulation with left diaphragmatic crus, wall penetration by vessels and nerves and lack of supporting neighboring structures [7,8,17].

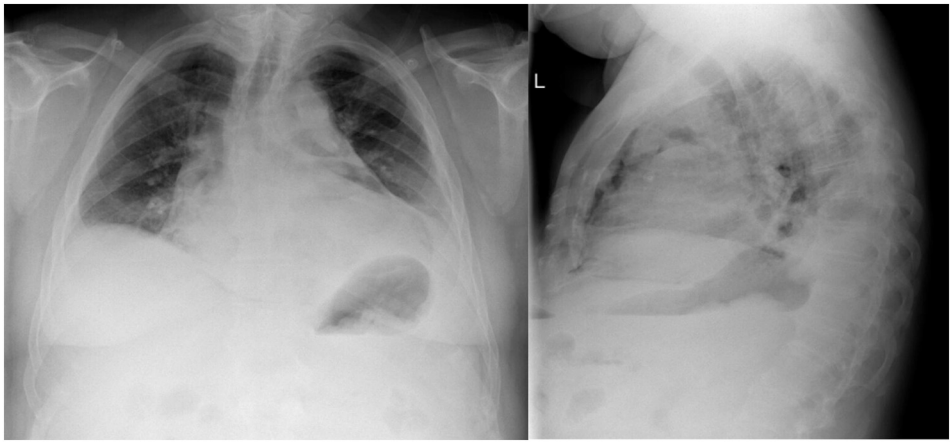
BS pathogenesis happens after a violent vomiting against a superior sphincter that is not totally relaxed, due to discoordinated esophageal motility during vomiting, resulting in increased intraluminal pressure and full-thickness rupture of esophagus wall [2,3,7,17]. There are cases that describe BS after epileptic seizure, asthma attack or the Heimlich maneuver [17]. In other few cases there is no apparent predisposing factor [8]. The complete wall rupture leads to exteriorization of oral secretions and gastric fluid to the mediastinum and pleural space, that spreads due to thoracic inspiratory negative pressure [4,18].

Patient findings are usually non-specific which leads to a delayed diagnosis, thus increasing mortality [8,17]. The main clinical feature is chest pain after vomiting [2,4]. The Mackler triad (vomiting, lower chest pain and subcutaneous emphysema) is present only in a minority of patients, near 14% [3,4]. Subcutaneous emphysema is the most pathognomonic sign, but is only found in 60% of patients [7,18]. In some cases, the patient could present with sepsis and shock due to mediastinal, pleural or abdominal infection [17]. Hematemesis is rare but could be present [7,8]. Mediastinitis is present in 25–31% of cases and it is suggested by fever, tachycardia, tachypnea and shock could at admission [1,9] A proper clinical examination and history is mandatory [8].

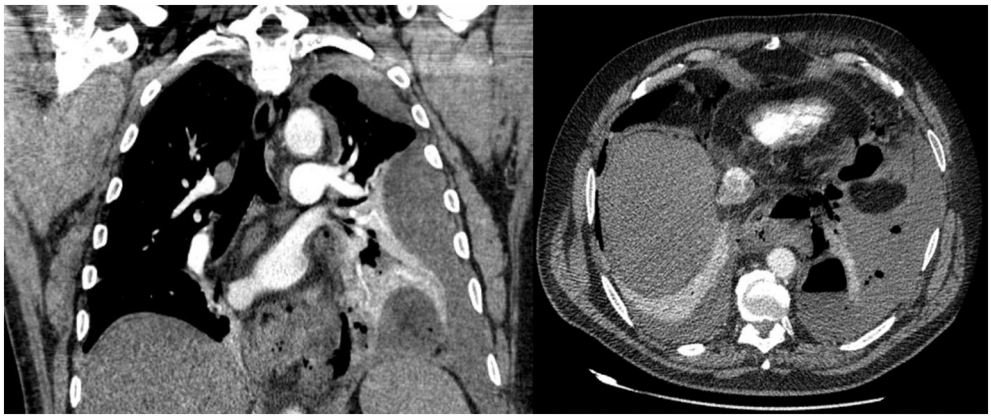
Due to unspecific clinical presentation, BS can mimic other conditions, like acute aortic dissection, myocardial infarction, pericarditis, pneumonia, pneumothorax, pulmonary embolus, perforated peptic ulcer and acute pancreatitis [2,3,18,19].

Blood and fluid samples are non-specific, although diagnostic thoracentesis can reveal food, increased amylase and pH below 6 [14,15,17].

The diagnosis is confirmed with water-soluble contrast, so if there is clinical suspicion of BS this exam is mandated [5,8,14,18–20]. The chest radiography could show left-sided pleural effusion, lung infiltrates and




**Figs. 1 and 2.** Chest radiography at Emergency Department, posteroanterior view and left lateral view. In those images there are signs of pneumomediastinum and consolidation on left chest.



**Figs. 3 and 4.** Chest computer tomography angiography, coronal and sagittal views, suggesting spontaneous rupture of the esophagus – important pneumomediastinum that was close to the left-posterior wall of the esophagus and an hydropneumothorax on the left chest.

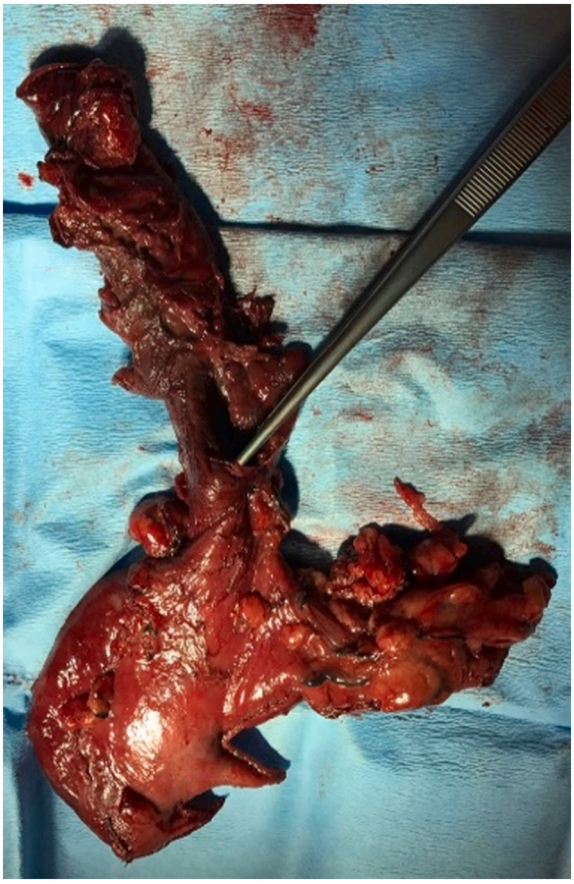


**Fig. 5.** Specimen with esophageal perforation revealed (dissection forcep).

atelectasis, but also pneumomediastinum and hydropneumothorax [2,17]. A radiographic sign that represents retrocardiac streaks of air forming a V (V-sign of Naclerio) could be present in 20% of cases [15,19]. If the perforation is sealed, none of the previous conditions is present in early stage [8]. Computed tomography with oral contrast is the best method for diagnosis, with high sensitivity [17]. It documents the perforation anatomy, peri-esophageal collections and shows signs of mediastinitis [2,3]. Upper gastrointestinal endoscopy has high sensitivity and specificity but may increase injury and mediastinal contamination [9,17]. The presence of pneumomediastinum with a preceding history of vomiting or retching followed by acute chest pain is virtually pathognomonic of BS [8].

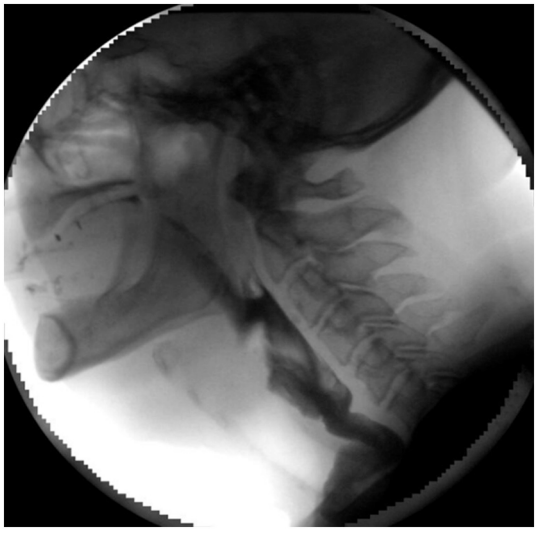


**Fig. 6.** Oral contrast esophagogram 1 month after discharge. There were not anastomotic leak or stenosis.

If not treated promptly it evolves to sepsis, shock, and death. Mortality rate could reach 40%, but mortality rate at 48 h after onset, in absence of a therapeutic approach, reaches 100% [4,11,17,18]. For those reasons, BS should be treated as a life-threatening condition [13].

BS management includes several options: conservative, endoscopic or surgical [9,14,17]. It needs to take into account time of diagnosis, extent of damage, local lesions and of patient status [13].

The main principles of treating this condition are limit contamination, adequate drainage of pleura and mediastinum and efficient antibiotic treatment to eliminate the infection [9,18]. Conservative management should include cessation of oral intake, broad-sprectum antibiotic therapy, intravenous proton pump inhibitors to decrease gastroesophageal reflux, restoration of hydro-electrolyte balance, fluids and enteral/parenteral nutrition and nasogastric (decompressive) tube and need to pleural drainage. It should be stated that conservative management should only be applied in selected patients and with very close monitoring [1,5,9,13].

Endoscopic treatment is possible by using esophageal luminal stents or by managing the esophageal wall defect with placement of clips but is reserved to selected cases and the first one is not superior comparing to primary sugical repair [13]. The endoscopic approach is preferred when there are no signs of sepsis and there is minimal contamination of the pleural and mediastinal spaces [14].

Surgical management is considered the most effective treatment for BS and some papers refer to it as a "gold standard" [8,9,12,13]. It is indicated when there is clinical worsening if the initial option for conservative or endoscopic management [14,17]. Surgery should include pleural cavity drainage, debridement and primary repair, drainage of pleural and mediastinal spaces, gastric decompression if indicated and enteral feeding access if necessary [2,5]. As for a primary repair an esophagectomy or esophageal exclusion can be performed [1,7]. Primary repair should be done in cases with early diagnosis (24 h until 72 h in some studies), in clinically (cardiovascular and pulmonary) stable patients, without symptoms of sepsis [13]. It is the treatment of choice along with pleural drainage [1,9,13,18]. There is need to debridement of necrotic tissues before performing a tension-free two-layer suture. However, the longer and more delayed diagnosis, the more aggressive and extensive surgery is needed. Esophagectomy is an option if there is severe mediastinitis after large thoracic perforations, if patient condition allows. If there is a large defect, esophagectomy is also an option. Diversion without esophagectomy is reserved for critical patients or if extensive infection is present and esophagectomy is not recommended [1,13]. In this approach, an end esophagostomy is performed to eliminate oral secretions and terminal esophagus is stapled or connected to a T-tube. It is includes a decompression gastrostomy and a feeding jejunostomy for early enteral nutrition [18].

In this case report, the patient had a delayed diagnosis (near 2 and-a-half days) with a large pleural effusion and pneumomediastinum with a large lower esophageal defect. However, he had no signs of septic shock or relevant laboratory signs of systemic inflammatory response. Despite his age and comorbidities, this situation required a multidisciplinary approach (General Surgery, Anesthesiology, and Intensive Care Departments), which lead to an emergent transhiatal esophagectomy with left chest pleural drainage. This approach allowed for the removal of the destroyed esophageal and access to the lower mediastinum, avoiding the morbidity of a thoracic approach. Esophagectomy with an anastomosis to the neck in stable patients in these suitable conditions allows the surgeon to perform a primary anastomosis, far from infection site.

The patient recovered well without major complication, successfully discharged home on 21st day after surgery and with no significant consequences to his quality of life.

## 4. Conclusion

Boerhaave syndrome is a rare life-threatening surgical condition. A good patient history and high degree of suspicion are required for a swift diagnosis, due to its nonspecific signs. A delay in diagnosis could be fatal to the patient. There are several options in BS management, depending on patient status, time of onset and presence of septic shock. Surgery is the most effective treatment and transhiatal esophagectomy with primary anastomosis could be an option in patients not undergoing primary repair.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

João Simões – data collection and writing the paper.
André Lázaro – writing the paper and review.

## Ethical approval

The patient was informed about publication of his case.

## Guarantor

João Manuel Martins Simões

## Declaration of competing interest

None.

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