Acute Respiratory Failure in a Patient With Spontaneous Esophageal Rupture (Boerhaave Syndrome)

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Introduction

Spontaneous rupture of the esophagus (Boerhaave syndrome) is a rare but life-threatening condition that was first described by a famous Dutch physician, Herman Boerhaave, in 1724.[1] Boerhaave syndrome nearly always results from an abrupt rise in the intraluminal esophageal pressure. Vomiting, chest pain, dyspnea, and subcutaneous emphysema are the most commonly reported symptoms. The barogenic nature of the rupture probably explains the wide mediastinal contamination, which can rapidly cause severe respiratory distress, sepsis, and shock. Thus, a prompt diagnosis plays a crucial prognostic role. Because Boerhaave syndrome is rare and shows a heterogeneous clinical course, no accepted standard treatment has been established. Most authors agree that surgery, supported by appropriate medical treatment, is the first-line therapy.

We report a case of 79-year-old man who presented with severe chest pain and respiratory distress and who was diagnosed and successfully treated with primary surgical repair.

Case Summary

A 79-year-old man presented to the emergency department of the Medical University Hospital in Warsaw, Poland, with a 3-hour history of severe retrosternal pain and progressive dyspnea, preceded by an episode of vomiting, following a large meal. His medical history was unremarkable except for intermittent asthma, which had never been regularly treated. He specifically denied any history of gastrointestinal disease, alcohol abuse, or cigarette smoking.

On admission the patient was severely ill and presented severe respiratory distress (respiratory rate 40 breaths/min), peripheral cyanosis, blood pressure 200/100 mm Hg, temperature 36.6°C, heart rate 100 beats/min, and S_{pO_2} 69% while breathing room air. Physical examination revealed diminished breath sounds and dullness to percussion at the base of the left lung. Heart sounds were normal. Abdominal examination showed slight epigastric tenderness and normal bowel sounds.

Since arterial blood analysis confirmed hypoxemia (P_{aO_2} 37 mm Hg) and hypercapnia (P_{aCO_2} 53 mm Hg), we administered oxygen via face mask to maintain $S_{pO_2} > 90\%$. Electrocardiogram revealed sinus tachycardia with left-axis deviation and right bundle branch block. Chest radiograph showed a left-sided apical pneumothorax and a homogeneous opacity at the base of the left lung (Fig. 1). He was transferred to the respiratory intensive care unit.

Intravenous fluids and analgesics were given to control the chest pain. Complete blood count and serum chemistry were normal. Computed tomography (CT) confirmed left-sided pneumothorax, revealed the presence of pleural effusion (Fig. 2), and allowed us to exclude other thoracic causes of pain, dyspnea, and respiratory distress (eg, aortic dissection, pulmonary embolism). A normal abdominal radiograph and CT made an abdominal source of symptoms unlikely. We decided to drain the left pleural space, and immediately after chest-tube insertion we retrieved air bubbles and pleural fluid containing food particles. We applied suction to the chest tube and started intravenous antibiotics. Contrast esophagogram showed an intrathoracic extra-esophageal leak of the contrast from the left lower third of the esophagus (Fig. 3).

Spontaneous rupture of the esophagus (Boerhaave syndrome) was the diagnosis. Since the time to diagnosis was

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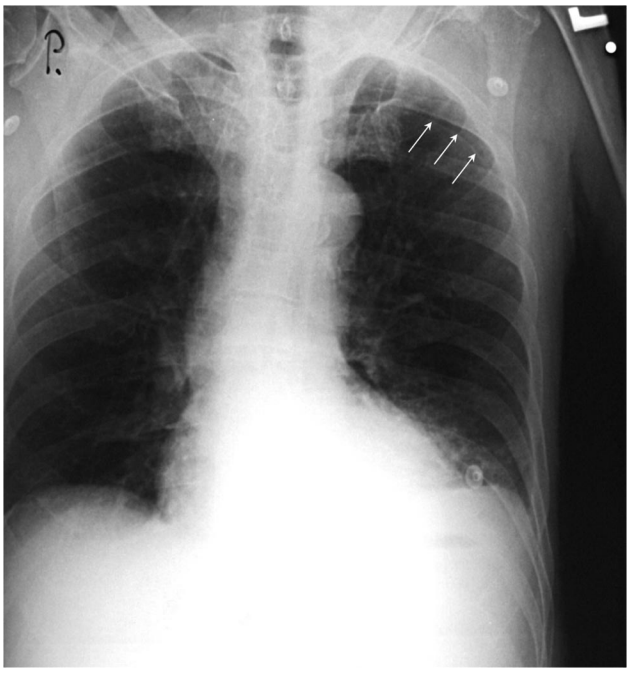

Fig. 1. Chest radiograph shows a left-sided apical pneumothorax (arrows) and a homogenous opacity at the base of the left lung. The upper margin of the opacity is well defined and sharp, whereas the lower margin is indistinguishable from the adjacent left hemidiaphragm. As the left costophrenic angle is well preserved and the gastric air bubble is not clearly visible, this might suggest elevation of the left hemidiaphragm. However, the lateral displacement of the dome with a steep lateral slope and gradual medial slope requires differentiation from pleural effusion (subpulmonic effusion).

relatively short (5 hours from admission), he was scheduled for primary esophageal repair. Left thoracotomy was performed 16 hours after the onset of symptoms. A 3-cm linear tear was found in the posterolateral esophageal wall, just above the diaphragm. The rupture was closed in 2 layers and reinforced with the stomach wall (fundoplication). The postoperative period was uneventful. He was discharged 11 days after the operation.

Discussion

Data on the incidence of Boerhaave syndrome are scarce. Hill et al found an approximate incidence of one in 53,000 of all hospital admissions.[2] Spontaneous esophageal ruptures are less common than iatrogenic and traumatic esophageal ruptures, and account for 15–40% of all esophageal perforations.[3-6] Boerhaave syndrome usually occurs in patients 40–60 years old, but has also been reported in neonates and the elderly. There is male predominance, with a sex ratio range of 2:1 to 5:1.[7-9] Conditions predisposing to spontaneous esophageal rupture include alcoholism, gastroesophageal reflux, peptic ulcer, hiatal hernia, and neu-

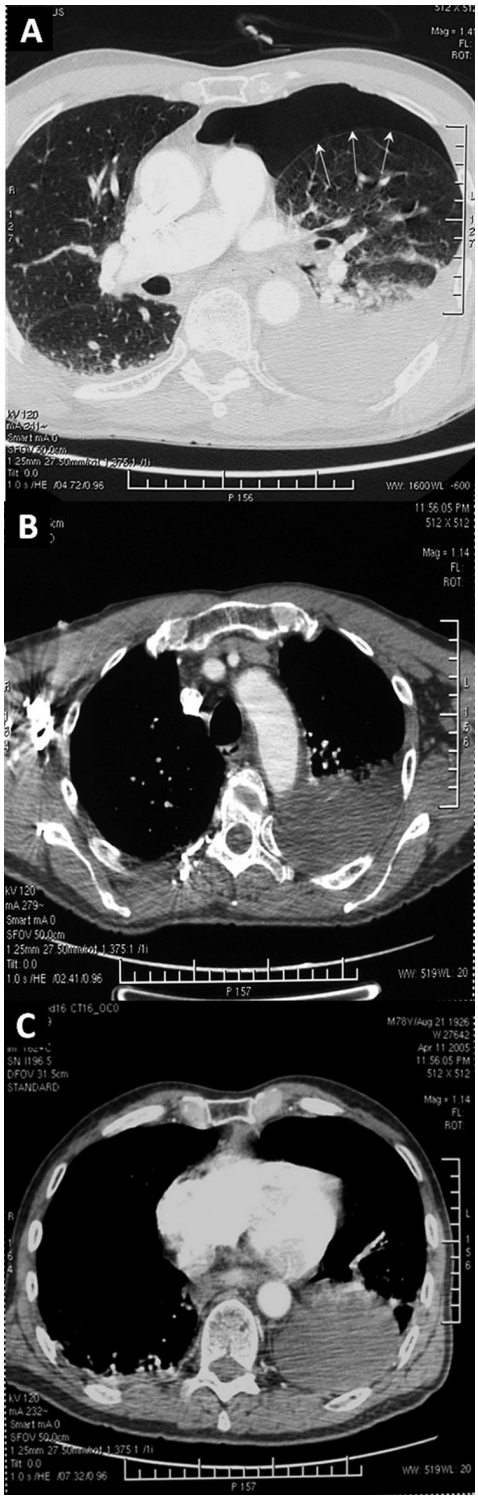

Fig. 2. Computed tomogram shows (A) left-sided medium-size pneumothorax (arrows). The underlying lung parenchyma appears normal and there is no evidence of mediastinal and/or subcutaneous emphysema. The posterior part of the left pleural space is occupied by pleural effusion. B: The soft window setting at the level of the aortic arch reveals free-flowing pleural effusion in the dependent compartment of the left pleural space. C: The scan at the base of the lung shows loculated pleural effusion.

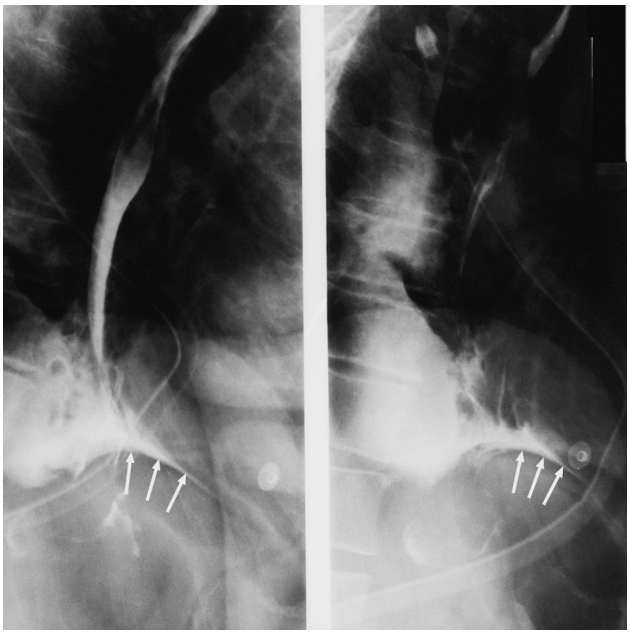

Fig. 3. Contrast esophagogram shows an intrathoracic extra-esophageal leak of the contrast agent from the left lower third of the esophagus. The sharp lower margin of extraluminal contrast medium collection is formed by the left hemidiaphragm (arrows). Only a small volume of the contrast agent passes into the stomach.

rological disorders.[7] In the vast majority of cases the tear is situated in the left posterolateral wall of the lower third of the esophagus and communicates with the left pleural cavity.[10,11] The overall mortality range has been reported as 6–31%, but might be as high as 65–89%.[5-7,9,12] This makes Boerhaave syndrome the most lethal perforation of the gastrointestinal tract.

The "classic" clinical presentation of Boerhaave syndrome includes episodes of retching or/and vomiting (often preceded by excessive food and alcohol intake), followed by a sudden onset of lower chest and upper abdominal pain. Pain, the most striking symptom of Boerhaave syndrome, is experienced by 83% of patients.[7] Vomiting and shortness of breath are also common, reported in 79% and 39% of patients, respectively.[7] Vomiting, lower thoracic pain, and subcutaneous emphysema (found in 28–66% of patients) are known as the Mackler triad, which is very suggestive of Boerhaave syndrome. Up to one third of all patients have an atypical presentation. The differential diagnosis of Boerhaave syndrome includes various thoracic and abdominal conditions (examples presented below).

Blood laboratory tests offer little help in the diagnosis. Normal pancreatic and liver tests make acute pancreatitis and cholecystitis unlikely. Normal troponin activity may help to exclude acute myocardial infarction. In patients with pleural effusion a pleural tap is more informative than other laboratory tests. Macroscopic assessment and/or cytology often reveal undigested food particles, which confirms gastrointestinal-tract perforation.[13] The chemistry of the pleural fluid usually shows low pH and high amylase level.

Imaging is pivotal in diagnosing Boerhaave syndrome. The upright chest radiograph is usually abnormal and reveals unilateral pleural effusion (present in 90% of patients, usually left-sided) and pneumothorax (80% of patients). Other findings may include pneumomediastinum, subcutaneous emphysema, mediastinal widening, or the so called "V-sign of Naclerio" (a radiolucent V-shaped band of gas bordered by mediastinal and pleural structures).[14] A normal chest radiograph is found in approximately 10% of patients.

Thorax CT allows a more detailed assessment of the mediastinal structures (eg, communication of an air-filled esophagus with a mediastinal or pleural air-fluid collection) and better visualization of the lung and pleural cavities. Thus, CT is important in differentiating Boerhaave syndrome from other pulmonary and extrapulmonary conditions (eg, aortic dissection, myocardial infarction, pulmonary embolism, massive pneumonia). Abdominal radiograph, CT, and ultrasound are very useful in evaluating for potential abdominal causes of symptoms (eg, acute pancreatitis, subdiaphragmatic gastrointestinal perforation, subphrenic abscess, acute cholecystitis).

The diagnosis of esophageal perforation should be confirmed with contrast esophagography (water-soluble contrast is recommended), which provides evidence for the extra-esophageal leak of contrast and may also outline the length and location of the perforation. Since, the sensitivity of this method is 75–90%, a negative result does not exclude esophageal perforation.[12,15-17]

Endoscopy enables direct visualization of the tear site and can yield important information before surgery. However, endoscopy may carry an additional risk of increasing the size and extent of the original perforation.[18]

The 3 major therapeutic approaches are surgical, endoscopic, and conservative. Their effectiveness was recently analyzed by de Schipper et al, who proposed a reasonable treatment algorithm for Boerhaave syndrome.[19] The therapeutic approach should, however, be flexible and highly individualized for each patient.[6] The critical determinants are the interval between injury and potential surgery, the location and extent of the perforation, the patient's underlying physical condition, and the presence of sepsis.[6,19]

Surgery is usually regarded as the first-line therapy, and should be supported with appropriate medical treatment. The most successful procedure involves primary repair of the rupture, with or without local debridement, and adequate drainage of the mediastinum and pleural cavity. This approach is recommended mainly for patients who undergo surgery within 12–24 hours of symptom onset.[3,20]

The longer the delay, the more extensive the tissue necrosis and edema, perhaps precluding a successful repair.

According to the review of the case series by Brinster et al, the mortality rate in patients treated with primary repair was 4% when treatment was initiated within the first 24 hours, and 14% when the intervention was after 24 hours.[3] However, other authors have suggested that a delay of > 24 hours does not preclude primary esophageal repair.[5,9,17] The alternative interventions for low thoracic esophageal ruptures include esophageal resection, drainage alone, T-tube drainage, and exclusion and diversion.

Endoscopic placement of a plastic-covered self-expanding metallic stent to bridge the esophageal tear offers an attractive, noninvasive treatment for esophageal perforation, including spontaneous esophageal rupture.[21,22] Although the results of stent placement in patients with Boerhaave syndrome have been encouraging, some authors still consider self-expanding esophageal stents controversial. Endoscopic stent placement seems to be appropriate for selected patients without wide mediastinal contamination and sepsis.[7,19-21]

Conservative management, which includes intravenous fluids, antibiotics, oxygen therapy or ventilatory support, nasogastric suctioning, drainage via tube thoracotomy, and early use of nutritional supplementation, might be appropriate in selected patients with late diagnosis, a well-contained perforation, and minimal mediastinal and pleural contamination.[3,19]

Teaching Points

Spontaneous esophageal rupture (Boerhaave syndrome) is a rare but life-threatening condition. Because of common chest symptoms, including pain and dyspnea, a large proportion of patients might be initially referred to nonsurgical departments. Since severe respiratory distress, sepsis, and shock may develop rapidly, early diagnosis is crucial. The most successful treatment is primary esophageal repair.

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