

A 55-Year-Old Man with Chest Pain

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CASE PRESENTATION

A 55-year-old man presented to the emergency department complaining of epigastric pain, retrosternal chest pain, and dyspnea. His symptoms started suddenly early in the morning after he experienced 2 episodes of retching and forceful vomiting of food material followed by a small amount of bright red blood. The patient had consumed excessive amounts of alcohol the night before. He reported that he usually drinks on social occasions and smokes 1 pack of cigarettes a day. He had no other past medical history.

On physical examination, the patient appeared slightly anxious and was in mild respiratory distress. He was afebrile, his blood pressure was 138/83 mm Hg in the right arm and 136/85 in the left, and his pulse was 116 bpm. Oxygen saturation was 97% on room air, with a respiratory rate of 16 breaths/min. His head was normocephalic, but he had subcutaneous emphysema in the neck and upper chest. Cardiovascular evaluation revealed tachycardia without audible murmurs. Lung examination showed diminished breath sounds to auscultation, dullness to percussion, and decreased tactile fremitus over both lung bases. His abdomen was soft with mild epigastric tenderness, but there was no guarding, rebound tenderness, or hepatosplenomegaly. Normal bowel sounds were present. No stigmata of liver disease were detected. Digital rectal examination was normal and negative for fecal occult blood.

Laboratory studies revealed a leukocyte count of $11.7 \times 10^3/\mu\text{L}$, with 79% neutrophils and 9% bands. Other notable laboratory study results were as follows: alanine aminotransferase, 117 U/L (normal, 0–55); aspartate aminotransferase, 260 U/L (normal, 12–45); total bilirubin, 1.7 mg/dL (normal, 0.1–1.2); direct bilirubin, 0.6 mg/dL (normal, 0.0–0.4); alkaline phosphatase, 31 U/L (normal, 37–107); and albumin, 2.0 g/dL (normal, 3.5–5.0). Amylase was 250 U/L (normal, 34–122) and lipase was 2 U/L (normal, 22–51).

Chest radiography revealed extensive pneumomediastinum and bilateral pleural effusions but no free air under the diaphragm (Figure 1).



Figure 1. Posteroanterior chest radiogram demonstrating pneumomediastinum and bilateral pleural effusions.

WHAT IS YOUR DIAGNOSIS?

- (A) Mallory-Weiss tear
- (B) Spontaneous rupture of the esophagus (Boerhaave's syndrome)
- (C) Acute pancreatitis
- (D) Perforated peptic ulcer
- (E) Aortic dissection

WHAT IS THE NEXT BEST STEP IN EVALUATING THIS PATIENT?

- (A) Computed tomography (CT) of the chest
- (B) Emergent exploratory thoracotomy

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- (C) Barium esophagography
- (D) Water-soluble contrast esophagography (Gastrografin [diatrizoate meglumine])
- (E) Esophagogastroduodenoscopy (EGD)

ANSWERS

The correct answers are (B) spontaneous rupture of the esophagus (Boerhaave's syndrome) and (D) water-soluble contrast esophagography (Gastrografin).

This patient presented with the typical clinical features, physical signs, and radiographic findings of Boerhaave's syndrome. He had the classic triad of vomiting, chest pain, and subcutaneous emphysema. Boerhaave's syndrome often follows repeated bouts of retching and vomiting after a heavy meal or binge alcohol drinking.

Mallory-Weiss tears usually present with hematemesis. Patients often give a history of nonbloody vomiting or retching before the onset of hematemesis, but this is not universal and in some cases Mallory-Weiss tears can be spontaneous. A Mallory-Weiss tear would be very unlikely to cause the development of subcutaneous emphysema.

Acute pancreatitis should be considered given the presence of acute abdominal pain, vomiting, elevated serum amylase, and left-sided pleuritic chest pain. In acute pancreatitis, vomiting, if manifest, is more likely to follow the onset of pain,¹ and serum lipase is typically elevated. This patient had a normal lipase concentration, and amylase can often be elevated in patients with recurrent vomiting, regardless of the cause (usually of salivary source).

Perforated peptic ulcer is possible but less likely. Peptic ulcers in the stomach or duodenum are often associated with pain prior to bleeding, but this is not universal. In addition, there was no free air under the diaphragm on chest radiography as might be expected with a perforated abdominal viscus. A perforated peptic ulcer would not be associated with subcutaneous emphysema.

Aortic dissection is unlikely given that the patient has no known history of hypertension, his blood pressure was not elevated on presentation, there was no asymmetric pulse deficit, and his chest pain was not described as severe or excruciating. Although aortic dissection can manifest as widened mediastinum on chest radiography, it should not result in pneumomediastinum. Patients with aortic dissection often describe a painful, tearing sensation as the dissection progresses.

Gastrografin esophagography should be performed whenever esophageal rupture is included in the differential diagnosis. It will reveal the location and extent of the perforation as evidenced by the extravasation of contrast material. Although barium is superior in demon-

strating small perforations, it causes an inflammatory response in mediastinal and pleural cavities and is therefore not routinely used as the primary diagnostic study. If the patient is suspected to have an esophagorespiratory fistula, barium esophagography would be indicated as the lung tolerates barium aspiration well, but Gastrografin aspiration can lead to a chemical pneumonitis.

A CT scan may show extraesophageal air, periesophageal fluid, mediastinal widening, and air and fluid in the pleural spaces, but it would likely not show the exact location of the esophageal rupture. Endoscopy is contraindicated in patients with suspected esophageal perforation as there is a significant risk of extending or worsening the perforation or introducing excessive air into the mediastinum. Emergent thoracotomy, although warranted in this patient, is not appropriate at this juncture because the diagnosis of esophageal rupture has not yet been confirmed.

CLINICAL COURSE OF CASE PATIENT

The patient underwent Gastrografin esophagography, which demonstrated an anterior esophageal rupture at the level of the diaphragm, with free extravasation of contrast material into the mediastinum (**Figure 2**). An immediate surgical consult was requested. Broad-spectrum antibiotics (cefepime and metronidazole) were administered and the patient was urgently taken to the operating room. He underwent left-sided thoracotomy, where a 2-cm perforation in the lower esophagus at the level of the hiatus was seen, with elevation of the parietal pleura covering the distal esophagus. A moderate amount of purulent fluid was present in the left pleural cavity. Primary repair of the esophageal defect was performed. To check esophageal patency, methylene blue was injected via a nasogastric tube; no evidence of leakage into the pleural cavity was observed. A pleural patch was created from the adjacent pleura, wrapping it around the repaired segment of the esophagus. Two chest tubes were inserted into the left pleural cavity. Also, a gastrostomy tube (to decompress the stomach) and a jejunostomy tube (for feeding) were inserted.

The patient tolerated the operation well and had an uneventful postoperative recovery period. Nine days after the surgery, a Gastrografin esophagogram showed no evidence of leakage of contrast material. The patient has been well thereafter with no long-term difficulties.

BOERHAAVE'S SYNDROME

Spontaneous rupture of the esophagus (Boerhaave's syndrome) is a rare but serious condition that requires a prompt diagnosis and early aggressive management. It was first described in 1724 by Hermann Boerhaave, a

Dutch physician.^{1,2} His patient was a 50-year-old admiral (Baron John van Wassenae), who developed a sudden excruciating chest pain while straining to vomit, followed by shock and then death 18 hours later. Autopsy showed a rupture of the distal esophagus into the left chest.

Clinical Presentation

Classically, the patient is a middle-aged, white male with a history of overindulgence in food or drink who develops vomiting, followed by lower chest pain, mediastinal or subcutaneous emphysema (Mackler's triad),³ and cardiovascular collapse. Some patients report that swallowing aggravates the pain.

In contrast to patients with Mallory-Weiss tears, hematemesis, if present, is seldom a significant feature of the presentation.¹ On physical examination, a critically-ill patient is often observed, usually sitting up in bed in a forward-crouching position.⁴ As mentioned, subcutaneous emphysema is frequently seen. Chest examination may reveal findings suggestive of hydrothorax or pneumothorax. Hamman's sign—a crunching, rasping sound that is synchronous with heart beat, heard over the precordium and sometimes at a distance from the chest in mediastinal emphysema—can be heard in approximately 20% of patients.⁵ The abdomen is usually tender, especially in the epigastric region, with varying degrees of rigidity, sometimes simulating an intra-abdominal catastrophe. When the rupture is confined to the mediastinum, the patient may not look particularly sick and vital signs may be deceptively normal.¹

Boerhaave's syndrome is classically postemetic, but there are reports of similar spontaneous perforations occurring as a complication of lifting a heavy weight, severe asthma, parturition, and prolonged coughing or hiccups or in the newborn, in whom rupture into the right pleural cavity is more likely.^{6,7} The esophagus is more susceptible to rupture at a lower pressure than the rest of the alimentary tract because it lacks a serosal layer. The sudden rise of intraluminal pressure caused by an uncoordinated act of vomiting against an unrelaxed cricopharyngeal sphincter causes the tear, most commonly in the lower esophagus above the left diaphragm.^{6,8}

Diagnostic Evaluation

The diagnosis is often missed initially because this syndrome is rare and the conditions that can mimic it are more common. Among these are perforated or bleeding ulcers, acute pancreatitis, myocardial infarction, pulmonary embolism, dissecting aneurysm, spontaneous pneumothorax, and acute cholecystitis.¹

Plain radiography may detect widened mediastinum,



Figure 2. Water-soluble contrast esophagography image demonstrating a normal proximal and mid-esophagus (black arrow) and an anterior esophageal rupture at the level of the diaphragm, with free extravasation of contrast material into the mediastinum (white arrow).

hydrothorax, hydropneumothorax, or mediastinal emphysema but can be normal in 10% to 12% of patients.^{1,9–11} A contrast study is required to locate the exact site of the perforation, which helps in determining the best surgical approach. Normally, a water-soluble contrast is used, such as Gastrografin. Barium might provide better imaging quality, especially in small perforations, but it can induce an inflammatory reaction in the mediastinal and pleural cavities that leads to granuloma formation.¹²

In perforations of the cervical esophagus, which are most commonly instrumental, cervical emphysema is common but mediastinal emphysema is relatively rare.¹ Identification of mediastinal emphysema may avert a mistaken diagnosis of perforated peptic ulcer;¹³ conversely, the presence of subdiaphragmatic air is extremely rare in esophageal perforation.¹⁴ A mixed picture may occur in rupture of the esophagogastric junction.¹⁵

Management

Early aggressive operative management should be undertaken in cases of spontaneous esophageal rupture to maximize patient survival. Surgery is generally required for thoracic perforations, whereas cervical perforations can often be managed without surgery.¹⁶ Various surgical techniques to repair the perforated esophagus have been described in the literature,

including primary repair, reinforced primary closure with either tissue or mesh, mediastinal drainage alone, T-tube drainage, exclusion and diversion, and intraluminal stenting and resection.¹⁷ Primary repair can be performed thoracoscopically, but this approach has been restricted to the management of iatrogenic perforation recognized early.¹⁷ Intraluminal stenting and resection have largely been restricted to the management of malignant lesions and caustic strictures that have perforated at endoscopy and cannot be recommended in Boerhaave's syndrome.¹⁷ A case of failed primary repair with the development of esophago-bronchial fistula subsequently treated with endoscopic placement of fibrin glue has been reported.¹⁸ A case of failed surgical therapy for Boerhaave's syndrome with resulting esophagopleural fistula has been treated endoscopically with fistula tract coagulation and using an endoscopic suturing device.¹⁹

Patients are usually placed on broad-spectrum antibiotics. A feeding jejunostomy tube placed at the time of surgery gives a reliable and safe enteral route for feeding. In cases where enteral feeding is not feasible, total parenteral nutrition provides a good alternative. Also, gastrostomy should always be considered to provide adequate gastric drainage and to decrease reflux of gastric contents back into the esophagus, which can delay the healing of the esophageal defect.

In general, mortality for cases treated nonoperatively approaches 100% after 1 week.^{6,20–23} Rarely, when diagnosis is delayed for more than 5 days, the patient is stable with no signs of sepsis and no signs or symptoms of empyema, and the perforation is confined to the mediastinal cavity and is well drained internally, nonoperative therapy may be appropriate.²⁴ When patients managed conservatively begin to show signs of sepsis, they should be operated on without hesitation. **HP**

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