

Boerhaave' s syndrome: a case with an atypical right-sided oesophageal perforation

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Background. Boerhaave's syndrome is a complete transmural perforation of the thoracic oesophagus, with the oesophageal tear occurring on the left posterolateral side in 90 % of the patients.

Case report. We present a case of Boerhaave's syndrome with an atypical feature of right-sided oesophageal rupture. Chest CT has the advantage of imaging all thoracic structures and complications and excluding differentially diagnostic conditions in the case of a high clinical suspicion of oesophageal rupture.

Conclusions. CT is especially important in patients whose severe clinical condition does not permit esophagography.

Key words: esophageal diseases, rupture, spontaneous; tomography, X-ray computed

Introduction

Boerhaave's syndrome is a spontaneous complete transmural perforation of the thoracic oesophagus secondary to food bolus impaction. Perforation in most cases results from violent retching or vomiting after excessive food and alcohol intake. The syndrome

owes its' name to famous Dutch 17th century physician Hermann Boerhaave who described a case of Great Admiral of Dutch fleet who overate and after vomiting acquired this condition.

The syndrome is life threatening, with overall mortality of about 35%.¹⁻³ It is a consequence of neuromuscular incoordination, with a barogenic injury caused by a sudden increase in intraluminal pressure against a constricted cricopharyngeus muscle which fails to relax. Since the perforation is caused by vomiting and oesophageal barotrauma, it is, literally spoken, not spontaneous, but this term distinguishes it from much more often iatrogenic rupture. The oesophageal tear is usually linear, 1-5 cm in length, vertically ori-

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ented. It occurs in a majority of cases (90%) on the left posterolateral side, 2-3 cm above the gastroesophageal junction.^{1,3} Gastric content extraluminates into mediastinum and pleural space, ensuing mediastinitis, sepsis and shock. No haematemesis occurs because blood escapes outside the oesophageal lumen after rupture. Haematemesis is a feature of more common Mallory-Weiss tear, which is a gastric (rarely oesophageal) mucosal and submucosal tear.

The clinical features of Boerhaave's syndrome are often misleading. The so-called Mackler triad is virtually pathognomonic, consisting of forceful vomiting, sudden onset of pain (substernal, neck, abdominal-epigastric, or radiating to the shoulder) and subcutaneous emphysema. Complete triad is seldom present. Other symptoms include dyspnea, tachypnea, abdominal rigidity and signs of haemodynamic shock. The mainstay of therapy is prompt surgical intervention with closure of the tear, intravenous volume resuscitation and mediastinal drainage, except for some cases of small-contained iatrogenic injuries and cervical perforations. Boerhaave's syndrome occurs 2-5 times more often in males than in females, typically in middle-aged men.^{1,3}

We present a case of Boerhaave's syndrome with extensive bilateral intrathoracic pathology and an atypical feature of right-sided oesophageal rupture, reported in a minority of cases.^{4,5}

Case report

A 57-year old male patient was admitted to a county hospital after suffering a vomiting series. He presented with epigastric pain, forceful vomiting and signs of gastro-intestinal bleeding, without subcutaneous emphysema. His cardiac condition was stable. Plain abdominal film was normal. After insertion of nasogastric tube, about 700 ml of haematinic

content was removed. Endoscopy revealed extensive blood coagula in the oesophageal and stomach lumen.

Thirty-six hours after the presentation the patient was transferred to Clinical Hospital Center in haemodynamically stable condition, with intermittent vomiting. Initial oesophagography and endoscopy both failed due to dyspnea and psychomotoric excitation of the patient. During ultrasound examination he developed cardiorespiratory arrest, and was transferred to the intensive care unit after cardio-pulmonary resuscitation. Ultrasound examination showed signs of intestinal paresis and extensive pleural effusion. Thoracocentesis acquired gastric-like content. Since the patient was not able to undergo oesophagography, urgent endoscopy was performed and demonstrated 1 cm long oesophagus tear, on the right lateral side, just above the gastro-oesophageal junction with irregular margins, covered with fibrin. The adjacent mucosa was normal and without varices. There were no signs of bleeding artery or fresh blood flow, while stomach was filled with blood clots.



Figure 1. Chest CT scout view. Non-homogenous infiltration of the left lung, widened upper and middle mediastinum and signs of paralytic ileus.

CT was performed to demonstrate the extent of the disease. A scout view displayed a non-homogenous infiltration of the left lung, widened upper and middle mediastinum and signs of paralytic ileus (Figure 1).

Pleural effusions were bilateral, with right-sided major pneumothorax, infiltration of the left lung with atelectatic component and minor pneumothorax (Figure 2).

Pneumomediastinum was observed with tiny lucencies especially around distal oesophagus and air-fluid levels within mediastinum (Figures 3,4). Subcutaneous emphysema was also noted.

Shortly after CT examination the patient died. Pathology report confirmed right-sided oesophageal laceration and CT findings.

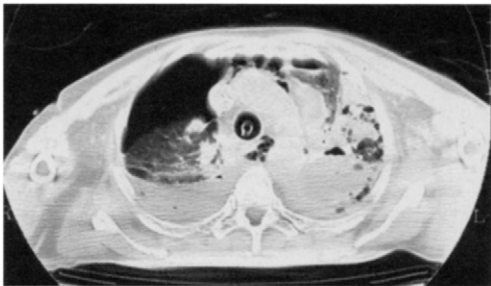


Figure 2. Axial CT. Bilateral pleural effusions, with right-sided major pneumothorax, non-homogenous infiltration of the left lung with atelectatic component and minor pneumothorax.

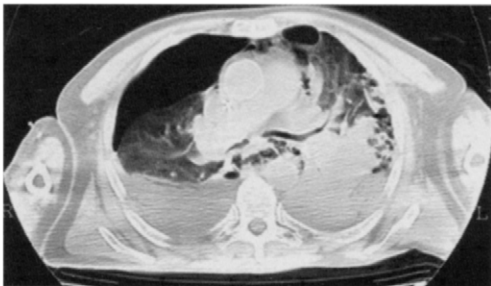


Figure 3. Axial CT. Pneumomediastinum with tiny lucencies especially around distal oesophagus.

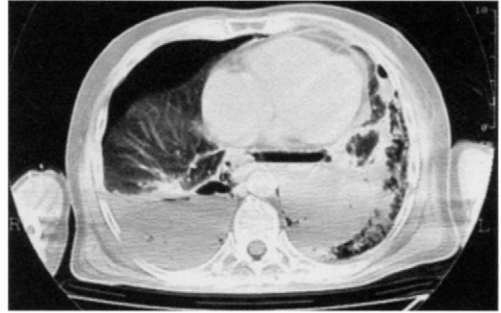


Figure 4. Axial CT. Air-fluid levels within mediastinum. Extensive pleural effusions.

Discussion

Boerhaave's syndrome is a rare, but diagnostically demanding entity. Fast and accurate diagnostics is invaluable. Upright chest radiograph is initially abnormal in 90% of patients with pneumomediastinum as the most important finding.³ The chest radiographs may be normal, because the findings may take an hour after the perforation to appear. The common feature is usually left-sided pleural effusion due to the site of perforation which mostly occurs in the left posterior aspect of the oesophagus; however if perforation occurs into right pleural cavity, it is usually in very young patients. Hydropneumothorax (in 51% of cases), subcutaneous emphysema or mediastinal widening may be present.⁶ A specific, but insensitive radiographic sign is the »V-sign of Naclerio« - an air lucency between lower thoracic aorta, oesophagus and diaphragm in the shape of the letter V, presenting localized mediastinal emphysema.

All described signs were present in this case, seen at CT, with an atypical site of oesophageal tear on the right lateral side and major right-sided pneumothorax. Computerized tomography has the advantage of displaying all thoracic structures and excluding differentially diagnostic conditions, like such as: aortic dissection, myocardial infarction, acute pancreatitis, pneumothorax, oesophagitis, peptic ulcer disease, spontaneous intra-

mural haematoma of the oesophagus (SIHO, oesophageal apoplexy) and Mallory - Weiss tear.⁷ CT scout view may replace chest radiograph in an urgent case. It is required especially in patients whose condition does not allow oesophagography,^{8,9} demonstrating the extralumination of contrast material into mediastinum and displaying the presence, location and the length of the tear.

Presented case emphasises the severity of this syndrome and the importance of awareness of clinical manifestations. Survival rate is 70-75% if surgical repair of the rupture is performed in early injury, within 24 hours of the incident.³ Significant number of patients are late in presentation for medical care.^{10,11} With late intervention, the mortality rate (even with surgical intervention) rises to higher than 50% and to over 80% after 48 hours.^{2,3,12} The delay in raising suspicion of oesophageal perforation results in postponing of appropriate and simple diagnostic method - oesophagography.

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