Rare disease

Atypical presentation of Boerhaave's syndrome

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Summary

Boerhaave's syndrome is the rare and often fatal condition of spontaneous oesophageal rupture. When present with atypical symptoms, diagnosis is frequently late, which contributes to the high morbidity and mortality. The authors describe a unique case of a 50-year-old man presented with atypical Boerhaave's syndrome, in which the diagnosis was made several days following presentation by observing a large pleural effusion that had evolved rapidly on chest radiographs. He was managed without thoracotomy. Although survival has been reported with non-operative management of contained oesophageal perforations, to our knowledge, this is the rare case report of surviving Boerhaave's syndrome with free pleural rupture, without thoracotomy.

BACKGROUND

Spontaneous intrathoracic oesophageal rupture or Boerhaave's syndrome is a life-threatening condition that requires early diagnosis and effective treatment if death or serious prolonged illness is to be avoided.¹ Without treatment it may be fatal.² Atypical presentation of the Boerhaave's syndrome occurs in approximately one-third of cases which may cause difficulties in making diagnosis.³ We report a case of Boerhaave's syndrome with atypical presentation and late recognition that was managed by simple chest tube drainage and nutritional support through feeding jejunostomy. We also review the literature in which clinical presentation, diagnosis and treatment were analysed.

CASE PRESENTATION

A 50-year-old gentleman, with complaints of sudden onset of pain in the epigastrium and left hypochondriac region, presented to the emergency department of our hospital. Pain was moderate in severity radiating to back and it was associated with two mild episodes of blood-mixed vomiting. The patient used to drink alcohol regularly, maximum 100 g per week, with a maximum of 30 g per day. He had no significant surgical or other medical history. On general physical examination, the patient was conscious, oriented and afebrile. His pulse rate was 80 per minute regular and blood pressure was 110/70 mm Hg in left arm supine position. The chest and cardiovascular system were both within normal limits. Routine laboratory investigations were all unremarkable. The patient was admitted with the provisional diagnosis of acute gastritis and treatment was started. Patient was kept nil per mouth, intravenous fluids and intravenous H₂-blocker was started and patient was sedated. The next day he developed dyspnoea, tachycardia and chest x-ray (CXR) showed left-sided pleural effusion. Pleural fluid aspiration showed total leucocyte count 3840 cells, differential leucocyte count; polymorphic leucocyte (PML) 70%, monomorphic leucocytes (MNL) 30%, red blood cells plenty, glucose 40 mg/dl and protein 3.2 mg/dl. The fluid was also sent for culture and sensitivity.

The patient's dyspnoea and restlessness persisted and worsened over the next 24 h. Repeat CXR showed increasing pleural effusion. A left-sided intercostal drainage tube was then inserted and drained 1.5 litres of dirty brown fluid immediately. The patient became symptomatically better after insertion of a chest tube. He developed fever over the next day for which broad-spectrum antibiotics were started. Upper gastrointestinal endoscopy was undertaken to evaluate the stomach and duodenum, which was reported normal, and ultrasound chest done, at its lower third, at the same time showed oesophageal thickening. Pleural drainage continued draining dirty brown fluid and the patient was kept on similar treatment. After 6 days, food particles were observed in the chest drainage bag. With suspicion of oesophageal perforation, a contrast study of oesophagus was done that showed oesophago-pleural fistula at the lower third of oesophagus (figure 1). The patient was toxic, febrile with purulent drainage in the chest tube. Antibiotic was changed according to culture sensitivity report. Initially thoracotomy, debridement and oesophageal repair was considered but it was deferred in view of late recognition of oesophageal perforation, relatively stable condition of the patient and decreasing fever after initiation of the antibiotics. CXR done at that time showed bilateral expanded lung without any intrathoracic collection. For supplementation of nutrition feeding jejunostomy and to prevent intrathoracic soiling with gastric juice drainage gastrostomy was done. Normal saline irrigation of mediastinum and left pleural cavity through the chest tube was started. A high-protein high-calorie liquid diet was started through the feeding tube and twice-daily normal saline lavage through the chest tube continued. The patient's general condition improved, along with the quantity of pus in the chest drainage tube decreased. A repeat contrast study of the oesophagus after 4 weeks showed persistence of the fistula but of lesser magnitude. This conservative treatment was continued and after 8 weeks drainage through the chest tube became minimal. Barium swallow study then showed no leak of contrast from the oesophagus (figure 2). CXR

Figure 1 Contrast study of the oesophagus showing leak of contrast in the left pleural cavity demonstrating the oesophageal perforation.

revealed expanded bilateral lungs. The chest tube was then removed and patient started orally and was discharged after 2 days of observation. At 10-month follow-up the patient was well and completely asymptomatic.

OUTCOME AND FOLLOW-UP

The patient made an uneventful recovery and was discharged home. He has been fit and healthy and completely symptom-free for last 18 months of follow-up.

Figure 2 Contrast study of the oesophagus showing rapid passage of contrast into the stomach without any leak of dye in the pleural cavity, hence demonstrating the healing of the fistula.

DISCUSSION

Spontaneous oesophageal rupture is a life-threatening condition for which surgical intervention within 24 h after the onset is usually recommended.⁴ Classical presentation with severe chest or epigastric pain, dyspnoea, vomiting, shock and subcutaneous emphysema is not always seen.⁵ Early CXR may not show pleural effusion or mediastinal widening. Symptoms are often non-specific and are easily confused with other disorders such as peptic ulcer, myocardial infarction, pancreatitis, dissecting aneurysm, pneumonia or spontaneous pneumothorax.⁶ For early diagnosis, a careful history, high clinical suspicion and the presence of roentgenological mediastinal and cervical air are the keys.⁷ Irrespective of the duration of the oesophageal rupture, aggressive resuscitation and prompt primary suture closure with reinforcement of the suture line with a well-vascularised pedicled tissue flap is required for optimal results.⁸ Early surgical intervention especially within 24 h of perforation is associated with better results.⁴ When surgical intervention is not offered because of several reasons, conservative management can be tried as discussed in literatures.⁹ Conservative treatment is instituted when patient is either not fit for surgical intervention because of poor general condition or patient's condition is stable and there are no signs of mediastinitis with free drainage of intrathoracic collection. In our case the diagnosis was delayed because of an atypical presentation until almost after 1 week when intrathoracic drainage tube started showing ingested food particles. Oesophagoscopy that was done on next day of chest tube insertion also failed to demonstrate any perforation. This also suggests that oesophagoscopy is not a good investigation for evaluating oesophageal perforation, although it may be useful when trauma (eg, ingested foreign body) is known or suspected to be present. We failed to perform contrast study at the same time, which could have demonstrated the leak. Our decision to manage the patient conservatively was guided by relatively stable general condition of the patient, absence of mediastinitis and effective drainage of the mediastinal and pleural content. We deferred surgery as we thought that the condition of the oesophagus was considered likely to be very friable and not amenable to successful surgical repair. Instead non-surgical management with regular mediastinal 'washout' was undertaken with normal saline lavage through the chest tube. Nutrition was achieved via a feeding jejunostomy. Feeding jejunostomy avoided the cost and complication associated with the parenteral nutrition.¹⁰ After 8 weeks of conservative non-surgical management, the oesophageal fistula completely healed, which was confirmed by the contrast study. Our case thus showed that conservative management is feasible when surgery is contraindicated. Early feeding jejunostomy is a cost-effective and technically easier procedure that can be undertaken safely under local anaesthesia when general anaesthesia is contraindicated to maintain the nutrition.

In conclusion, this case report is further evidence to bring increased awareness to correctly diagnose Boerhaave's syndrome to prevent life-threatening complications. In the case of patient presenting more than 24 h after rupture, for which surgical management with thoracotomy is too hazardous, non-operative management may be an acceptable option.

Learning points

- ▶ Boerhaave's syndrome can be difficult to diagnose and does not always present in the classical way. It may be diagnostic challenge, if presenting late.
- ▶ Careful elucidation of the history and accurate examination allied with appropriate investigations are necessary to make the diagnosis.
- ▶ CT scanning and contrast studies have a role to play in investigating the presence of a perforation and identifying the site.
- ▶ In a patient with Boerhaave's syndrome presenting more than 24 h after rupture, who appears too ill to survive thoracotomy, non-operative management may be an acceptable option.

Competing interests None.

Patient consent Obtained.

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