Reminder of important clinical lesson

A lesson in clinical findings, diagnosis, reassessment and outcome: Boerhaave's syndrome

Melanie Keane, T Gowripalann, A Brodbeck, P Bothma

Department of Anaesthetics/Intensive Care, James Paget University Hospital, Great Yarmouth, UK

Correspondence to Dr Melanie Keane, Melanie_Keane@hotmail.com

Summary

A man in his 30s presented with a brief episode of vomiting, acute abdominal pain and subsequent development of shortness of breath. On initial examination and investigation, the clinical impression was of a right-sided pneumothorax, pneumonia and pleural effusion. Early antibiotic treatment and management showed a clinical improvement, with the patient reporting resolution of his symptoms. This episode was short lived, with a further deterioration in his condition and worsening of symptoms. Ensuing examination, imaging and investigations demonstrated an oesophageal leak into the right pleural cavity. Following urgent stabilisation measures and insertion of a chest drain, he underwent successful surgical repair. Boerhaave's syndrome is an emergency situation, requiring quick recognition, diagnosis, aggressive treatment and management to optimise a good outcome.

BACKGROUND

This case demonstrates the difficulties in prompt recognition and diagnosis of Boerhaave's syndrome. It is a rare, well-known emergency situation, requiring prompt diagnosis and management to achieve a good outcome. Difficulties with collaboration between patient and physician, as to investigations and management strategies, can lead to a delay in diagnosis, as shown in this case.

CASE PRESENTATION

A man in his 30s presented to an accident and emergency department after waking with acute vomiting and abdominal pain. His pain was localised to the epigastrium, described as sharp and intense (8 out of 10) and localised on palpation. He denied any previous episodes, altered bowel habit, haematemesis or recent changes to diet or oral intake. No other significant medical history was noted. Examination revealed a tender epigastrium but no acute abdomen, and reduced air entry at the right base of his chest. He had a mild tachycardia and was afebrile. Initial imaging confirmed suspected right-sided pneumonia, a pleural effusion and small pneumothorax. At this time the patient declined any invasive management (chest drain or pleural tap), thus was commenced on intravenous antibiotics and reported marked symptom improvement, tolerating an oral diet.

At 48 h after admission he was clinically tachypnoeic (RR 26, oxygen saturations—SpO₂ 95% on air), tachycardic (110 beats per min) and vocalised marked breathlessness. On examination he had right upper quadrant discomfort on palpation, but epigastric tenderness had resolved. Imaging demonstrated a further worsening in right-sided pathology and despite discussion; he continued to decline a chest drain but agreed to a pleural tap, with brown thick fluid aspirate and rapid improvement in symptoms.

At 72 h he again reported a worsening in symptoms—tachycardic, tachypnoeic and agreed to supplemental oxygen therapy (FiO₂ 0.35). Following further discussion,

he agreed to have a seldinger chest drain inserted for management, with some initial success until drainage stopped abruptly. At this point he underwent an urgent CT scan of his thorax and was transferred to the high dependent unit for further management.

INVESTIGATIONS

Initial blood chemistry demonstrated a small rise in inflammatory markers—white cell count 25×10^9 /litre (normal upper limit 11×10^9 /litre) and a C-reactive protein 7 mg/l (normal upper limit 12 mg/l), all other markers were within normal parameters. At 24 h, arterial blood gas, on air, demonstrated a slight elevation in lactate 2.29 mmol/l (normal upper limit 2 mmol/l) and arterial partial pressure of oxygen (PaO₂) of 13.98 kPa (normal lower limit >10.6 kPa). At 48 h he showed a continued rise in C-reactive protein to 75 mg/l and at 72 h a further significant rise to 337 mg/l, consistent with working diagnosis of pneumonia.

Initial chest film confirmed a small right-sided pneumothorax, pleural effusion with probable consolidation to lower and mid-zones (figure 1). A repeat chest film at 48 h established a sustained right-sided pneumothorax, with almost complete opacification of the lung. At this point the working diagnosis was deemed to be a likely progression of pneumonia and a parapneumonic effusion/empyema.

The diagnostic pleural tap aspirated thick brown fluid, which was sent for culture and sensitivity, urine legionella was negative and a sputum culture was subsequently found to be positive for cocci, candida and enterococcus.

On recurrence of breathlessness and abrupt cessation of drainage from chest drain, a CT scan revealed a hydro pneumothorax with mediastinal shift, signs of a highly aggressive lung infection and the seldinger chest drain deeply embedded within the lung parenchyma.

A repeat CT scan with contrast proved an obvious leak into the right pleural cavity, the communication in the lower distal oesophagus and pleura. It also determined

Figure 1 Admission chest film showing right-sided effusion, small pneumothorax and lower zone consolidation.

a moderate right-sided pneumothorax, empyema and decompression of the mediastinal shift (figure 2).

DIFFERENTIAL DIAGNOSIS

Boerhaave's syndrome, first described in 1724, is a rare, life-threatening, consequence of spontaneous rupture of the oesophagus, due to acute barotrauma. Diagnostic errors are most commonly attributed to common differentials, a perforated gastric ulcer, followed by myocardial infarction, pulmonary embolism, pneumonia, dissecting aneurysm and pancreatitis.¹ Although radiological investigations such as plain chest x-ray (widened mediastinum, $\pm$ commonly, left-sided effusion) can assist narrowing the differentials, signs are often subtle and the definitive imaging is contrast CT scan.

The syndrome is more common in men, with a mean age between 40 and 60 years, in the majority of cases.² Many published case reports comment on Mackler's triad

Figure 2 Demonstrating an obvious leak from the distal oesophagus into the right pleural cavity with contrast.

of vomiting, chest pain and surgical emphysema, as key clinical signs and symptoms.^{3–4} Others suggest acute symptoms and signs to include haemodynamic instability and a Hammer sign on auscultation (crackling on chest auscultation—pneumomediastinum).^{5–7}

Our patient gave a brief history of vomiting and epigastric pain, which settled. There were no clinical findings consistent with the described Mackler's triad or a Hammer sign. Chest film suggested right-sided pathology and was more likely to be consistent with pneumonia. He was able to tolerate oral intake, had no further vomiting and was haemodynamically stable. On commencing antibiotic therapy and subsequent review he reported marked symptom improvement and it seemed reasonable to continue with the working diagnosis of pneumonia.

Although he later developed intermittent episodes of breathlessness and a continued rise in inflammatory markers, these continued to be explained by pneumonia with parapneumonic effusion, as interpreted by a repeat chest film. Declining interventions such as a chest drain or pleural tap also hindered definitive confirmation of the diagnosis at this point.

Following a further deterioration in his condition a CT scan and urgent decompression of the chest with a large bore drain resulted in a rapid improvement in symptoms and vital signs returned to normal limits. However, a correlation between oral intake and high output from the drain was made, raising the suspicion of an oesophageal tear/rupture. This was confirmed on a repeat CT scan with contrast.

TREATMENT

Initial treatment was broad-spectrum antibiotics with a later addition of flucloxacillin to cover suspected staphylococci as the causative organism. The patient's treatment and management were hindered by declining any intervention other than the above drug therapy at this time.

He reported symptom improvement on commencement of antibiotics and subsequently following a pleural tap. This led to a delay of 72 h before his symptoms worsened and agreed to have a seldinger chest drain inserted. Following his CT scan and arrival to the high-dependency unit, a large bore chest drain was inserted with immediate decompression of the chest, >1 litre of purulent fluid drained and a rapid improvement in symptoms was reported. Subsequently, a correlation between oral intake and high output from the drain was made, raising the suspicion of an oesophageal tear/fistula.

He remained nil by mouth and had a repeat CT scan with contrast demonstrating a tear in the right posterolateral side of the distal oesophagus. He was transferred to a thoracic unit, where he underwent a successful right thoracotomy, drainage and surgical repair.

OUTCOME AND FOLLOW-UP

Surgical repair was successful and he made a full recovery, discharged home within 3 months of initial presentation.

DISCUSSION

Descriptions of Boerhaave's syndrome depict a rupture, usually occurring on the left postero-lateral wall of the lower third of the oesophagus.^{8–9} Literature describes the

syndrome as rare, yet well recognised by the medical community and associated with significant morbidity and mortality.^{10 11} The combination of gastric juices and digestive enzymes leaking into the mediastinum promotes mediastinitis and sepsis, which are fatal in most untreated cases.⁸

Prior to the onset, there is often an episode(s) of excessive alcohol/food intake and/or hyperemesis. These are accompanied by the aforementioned Mackler's triad and Hammer sign on clinical examination. Our case did not support these criteria and on reviewing published case reports, few describe these features at presentation.

Younger adult cases are increasingly being reported within the literature.^{1–15} This has significance as the younger adult may be able to compensate relatively well until overwhelming sepsis ensues and clinical signs become evident, delaying consideration of the diagnosis.

It is commonly reported as a left-sided pathology, with radiological features demonstrating a widening of the mediastinum, surgical emphysema and left-sided effusion. Our case described right-sided features, with no mediastinal widening or evidence of surgical emphysema. Shaker *et al*,¹⁴ following a retrospective clinical review, also concluded that a chest x-ray was more likely to show a pneumothorax and effusion, rather than mediastinal pathology. Additionally, Teh *et al*¹⁵ reported only 5 out of 34 patients reviewed postdiagnosis of the syndrome, had a pleural effusion on chest x-ray and a median delay to diagnosis of 4 days.

The importance of having an awareness of the differing signs and symptoms of this syndrome enables the physician to consider this as a differential diagnosis. It is particularly important where initial management strategies are met with mixed success in symptom management.

Mortality and outcome are reported to be improved by prompt diagnosis and management. Shaker *et al*¹⁴ describe a decline in mortality rate with early diagnosis and surgical treatment, within 24 h of presentation from 40% to 6.7%. Surgical repair is seen as the gold standard, with significant reduced mortality from early intervention <24 h from presentation. However, some emerging reports suggest that this timeframe can be extended to 48 h and beyond.^{8 11 12} A delay of >56 h was seen as unlikely to benefit from a surgical repair, but our case demonstrated the benefit of surgical repair even if presentation is >72 h. Current and previous health status should be considered, as any underlying co-morbidities may obviously influence success and decisions to undertake surgical repair. However, as described in our case, a young otherwise relatively healthy adult should be considered for late surgical repair.

Learning points

- ▶ Boerhaave's syndrome is an emergency situation, requiring quick recognition, diagnosis and aggressive treatment and management to optimise a good outcome.
- ▶ A high index of suspicion should be considered where a young adult presents with clinical signs suggesting aggressive chest infection and mixed success in the treatment of symptoms.
- ▶ A rapid progression of chest pathology regardless of minimal haemodynamic instability should raise the suspicion of consideration of Boerhaave's syndrome.
- ▶ We suggest considering referral for surgical repair in patients diagnosed and/or presenting late with this condition, beyond the 'golden 24 h'.

Competing interests None.

Patient consent Obtained.

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