Clinical Reviews

Boerhaave's Syndrome: A Review of the Initial Management with Illustrative Case History

K Ping Ng, S Brauer & J Snape

Key points:
- Boerhaave's syndrome is an uncommon but serious clinical condition requiring a high index of suspicion to prevent delay in diagnosis.
- Sudden onset of pain following persistent and/or vigorous vomiting is the usual presenting symptom.
- Subcutaneous emphysema and identification of mediastinal or pleural effusion on chest x-ray are strongly suggestive of the diagnosis.
- Water-soluble contrast oesophagogram is the preferred choice of investigation as it is less likely to provoke or exacerbate inflammation in Boerhaave's syndrome compared to barium contrast.
- Non-operative treatment may be considered in some cases, depending on the cause and site of rupture, the underlying condition of oesophagus, the interval between rupture and treatment as well as the general condition of the patient.
- Early discussion with senior surgical colleagues is advised in all cases.

Abstract

Boerhaave's syndrome is a rare condition on the acute medical 'take' but has a high mortality and significant morbidity if not diagnosed early. This article reviews the approach to management at the 'front door', summarising the initial investigations and treatment. A recent case presenting to our unit is included at the end of the review to illustrate some of the issues raised.

Introduction

After the swift deterioration and death of the Grand Admiral of the Netherlands, Dr Hermann Boerhaave was intrigued at autopsy by the smell of duck and the sight of roast duck flesh and olive oil in the left pleural cavity, which were sourced to a transverse tear in the distal oesophagus.[1] Looking back at the patient's history, Dr Boerhaave related the findings to the Admiral's self-induced vomiting that took place 18 hours prior to the death. It may seem similar to bulimic behaviour nowadays. However, it was a common custom practiced by the rich and wealthy glutton in the past to self induce vomiting with an ipecac-like substance after a meal in order immediately to begin another.

Dr Boerhaave first described this condition of spontaneous oesophageal rupture in 1724. It has since been reported in the literature on numerous occasions to denote an oesophageal rupture, which is not caused by direct trauma, foreign body, instrumentation or secondary to surrounding infection.[2–3] Alexander the Great was also believed to have died from this condition.[4] This eponym (Boerhaave's syndrome) is currently used interchangeably with the term 'spontaneous rupture of oesophagus'.

Epidemiology

Boerhaave's syndrome is a rare condition, accounting for 15% or less of all cases of oesophageal rupture in the USA while the remainder is mainly iatrogenic in nature.[5] It is most common between the ages of 40 and 60 years[6] although some cases have also been reported in patients at the extremes of age.[7] Males are generally more at risk than females (2–5:1).

Although Boerhaave's syndrome is rare, it has the highest mortality of any form of gastro intestinal perforation[7] – up to 100%[8] if untreated. Since the first successful surgical repair in 1947, things have improved due to greater awareness of the disease, improved imaging and intensive care. The mortality is now about 30%.[8] Nevertheless, according to one study of 127 cases, 10% of patients are only diagnosed at post mortem.[9]

Mediastinitis, pericarditis, empyema, intrathoracic abscess, pneumonia and sepsis leading to shock and multi-organ failure are the main culprits for morbidity and mortality especially if presentation, diagnosis and treatment are delayed.[5] One study indicated an average mortality rate of 10–20% if patients with Boerhaave syndrome received surgical intervention within 18 hours of diagnosis. Delayed repair was liable to result in dehiscence.[10]

The prognosis is also affected by the site of perforation, the underlying condition of the oesophagus and general health of the patient.[11]

Pathophysiology

The principal underlying mechanism which results in oesophageal rupture in Boerhaave

Dr Khai Ping Ng
Ng MB ChB
F2 Doctor, Geriatric / General Internal Medicine
Kings Mill Hospital

Dr Sue Brauer
FRCR
Consultant Radiologist
Kings Mill Hospital

Dr Jeremy Snape
MB FRCP
Consultant Geriatrician
Kings Mill Hospital

Correspondence:
Dr J Snape
Kings Mill Hospital
Mansfield Road
Sutton in Ashfield
Notts. NG17 4JL
Emali: jeremy.snape@sfh-tr.nhs.uk

syndrome is the **rapid rise** in intraluminal pressure. The most common cause is emesis after high alcohol or food intake.[8] Other courses include straining at stool, childbirth, weight-lifting, seizure, forceful swallowing, fits of coughing, laughing, hiccupping, hyperemesis gravidarum, and Heimlich manoeuvre. In rare cases acutely raised external pressure transmitted via an open mouth to the oesophagus at the scene of an explosion has also been reported to lead to the rupture.[1,5,12]

The crucial contributor to the final event of rupture is not the absolute degree of the intraluminal pressure but the speed of the rise in pressure.

In addition, pre-existing abnormalities of oesophagus, for instance, gastro-oesophageal reflux disease, oesophagitis, oesophageal stricture, web, achalasia, ring and neoplasm increase the likelihood of spontaneous oesophageal rupture.[5,1] Similarly, patients with hiatus hernia are also suggested to be at increased risk.

Approximately 90% of the ruptures are left sided posterolateral in position and in the lower oesophagus about 3 to 6 cms above the diaphragm.[13,5] This is the most vulnerable area of the oesophagus due to a reduced thickness of the musculature and supporting surrounding tissues.[1] The average length of tear according to one study was 2.24 cm ranging from 0.6 cm to 8.9 cm.[14]

Clinical Features

Vomiting, chest pain and subcutaneous emphysema are the classical Meckler's triad of Boerhaave syndrome.[7,12,15] One small study of 14 cases suggested this triad is rare and only present in less than 10% (1 out of 14) of the patients.[16]

Chest pain

The majority of patients present with a sudden onset of pain, which is often preceded by vomiting.[17] The pain is generally excruciating, constant, stabbing and pleuritic in nature.[6,18] It may radiate to epigastric area, back or even left shoulder tip due to irritation of the diaphragm.[1] Swallowing often aggravates the pain and precipitates coughing.

If perforation occurs below the diaphragm, patients with Boerhaave syndrome may present with signs of peritonitis.[1] Unlike patients with Mallory-Weiss tear or gastro-duodenal ulceration, bleeding is generally not a feature in Boerhaave syndrome.

Subcutaneous emphysema

Rupture of the oesophagus results in the leakage of air and oesophageal or even gastric contents into the mediastinal space.[3] Each inspiration creates a negative intra-thoracic pressure, air and oesophageal contents are drawn into the potential space via the rupture site, resulting in pneumomediastinum.[19] Classical signs of pneumomediastinum are subcutaneous emphysema and Hamman's sign.[15] The subcutaneous emphysema is caused by air dissecting into the interstitial space of the dermis whilst Hamman's sign describes audible crepitations over the precordium.[20] Once pneumomediastinum has developed, subcutaneous emphysema can be found in all of the cases if thoroughly examined and Hamman's sign is present in approximately 50–80% of the cases.[21] Nevertheless subcutaneous emphysema only becomes apparent an hour after initial insult.[5]

As air dissects superiorly along the interstitial space, some patients may present with puffiness of the neck and face, or even proptosis.[12] Other unusual signs include rhinolalia or hoarseness of the voice if recurrent laryngeal nerve is involved.[7]

Dyspnoea

Dyspnoea is a prominent symptom which may result from the pain or from hydropneumothorax.[1] Signs of pneumothorax, pleural effusion or hydropneumothorax may be evident on chest examination especially affecting the left side.[1] However the examination may be entirely normal if presentation is very early.[1]

Shock and sepsis

As mediastinitis sets in, secondary to the presence of gastric contents in the mediastinal space, pyrexia is seen in most cases.[1,18] The inflammatory process often continues involving the pleural or even peritoneal space. Gastric contents and enzymes may allow inflammation and infection to seep into the pleural space. Reactive serous pleural effusion is present in most cases even if leakage is contained within the mediastinal space.[22] With a combination of respiratory compromise, fluid loss and widespread inflammation patients with Boerhaave syndrome often progress to severe sepsis and shock if diagnosis or treatment is delayed.

Investigations

Routine blood tests for patients with Boerhaave's syndrome typically show leucocytosis, left shift and raised haematocrit.[1,5,7] These findings are the results of the inflammatory process and fluid loss but are non-specific. Changes of the axis and decrease in voltage may be seen on the ECG.[18]

Erect chest radiograph

This is the most important initial investigation: findings may vary depending on the site of the perforation as well as the interval between the time of the event and the investigation but approximately 90% of the chest radiographs are abnormal.[3,5] The most common sign seen on the chest x-ray is unilateral (or occasionally bilateral) pleural effusion, which is reported in 90% of the cases.[1,6] Pneumothorax may present concomitantly with pleural effusion.[1] If the x-ray is taken at least one hour after the rupture, signs of pneumomediastinum and subcutaneous emphysema in the neck or chest region may also be apparent. The V-sign of Naclerio is another specific finding although it is only present in 1/5 of the cases.[7] As the name implies, it represents the dissection of air in the retrocardiac fascial plane, which creates a V shaped widening of the mediastinum.[1,7]

Pleural aspirate

Pleural fluid aspirate with a pH of less than 6.0 with food particles as well as an elevated amylase suggests oesophageal rupture.[1,5,7] As the serum amylase level is typically normal in Boerhaave's syndrome, the diagnosis of pancreatitis can be easily excluded. Cytology of the pleural fluid may also show squamous cells of gastro-intestinal tract origin.

Oesophageal contrast studies

The gold standard investigation is contrast oesophagogram. Despite some controversy over the choice of contrast, most of the literature suggests a water-soluble contrast medium is preferable e.g. Gastrografin. It is felt that a water-soluble contrast agent when compared with barium does not provoke or exacerbate the inflammation within mediastinum or pleural cavity.[7] If there is a risk of aspiration into the lungs Gastrografin should be avoided as it may induce pulmonary oedema due to its hypertonic nature. In this situation Iopromide would be a safer alternative, although sensitivity may be lower (60–75%) due to its rapid flow rate.[7] If no leakage is seen during initial oesophagogram, an attempt to obtain oblique or lateral view or to reposition the patient in the left and right lateral decubitus position may improve the sensitivity of the study.[7] A repeat study after an hour, or a barium contrast oesophagogram, which is more sensitive (around 90%) could be performed if the index of suspicion remains high.[1,7] The size of the tear may not correlate with the degree of extravasation on the oesophagogram, as in some cases the perforation site may be oedematous or blocked temporarily, for example, with blood clot.[1]

Some patients are too ill to tolerate contrast oesophagogram. For these cases, CT scan may prove to be useful in identifying a peri-oesophageal fluid collection or air track. It may not, however, demonstrate the exact location of the rupture.[5,7,12]

Management and Treatment

The immediate management of patients with Boerhaave's syndrome should ensure adequacy of the airway, breathing and circulation in an attempt to buy time for surgical intervention. This is especially important for patients who are in shock, either due to hypovolaemia or septicaemia. Patients should also be kept nil by mouth and commenced on intravenous broad-spectrum antibiotics (e.g. cefuroxime 750mg tds and metronidazole 500mg tds).

Patients who are severely shocked or appear to be in a moribund state should still be considered for urgent surgical intervention, as clinical improvement and recovery can sometimes occur after appropriate surgical management.[6]

Non-operative treatment

It was commonly believed that surgical intervention was the only option to ensure survival until the first case of successful non-operative treatment of Boerhaave's syndrome was reported in 1994.[24]

Non operative treatment comprises intravenous fluid and antibiotics. A nasogastric tube should be inserted, with suction to decompress the stomach, along with drainage of leakage or abscess via a thoracostomy tube. The patient will need to remain nil-by-mouth for around 7 days, and feeding should be provided either enterally, via a jejunostomy or parenterally.[7]

According to a meta-analysis of 86 patients with oesophageal perforation regardless of aetiology over 15 years, non-operative management may be suitable for the following patients:

- patients with a small oesophageal leak,
- leakage draining back into the oesophagus,
- patients with late presentation
- patients with no evidence of septicaemia
- patients without underlying oesophageal disease.[23]

It was also noted in this study that most perforations treated non-operatively were located in upper or middle third of the oesophagus, and were generally the result of iatrogenic injury. This group appears to have a better prognosis.[23] Appropriate selection of patients for non-operative treatment is not easy as the size of the perforation is difficult to determine with certainty except at operation. Early discussion with senior surgical colleagues, preferably in a specialist cardiothoracic centre, is recommended.

Operative treatment

Operative treatment remains the mainstay of treatment for the majority of patients with Boerhaave syndrome.[15,25] Several surgical approaches have been documented in Boerhaave's syndrome. A conservative surgical approach with routine mediastinal and pleural cavity toilet, debridement and drainage, together with gastrostomy and jejunostomy insertion is generally used in patients with delayed intervention where the edges of the perforation are too friable to suture or high risk of dehiscence if reconstruction is attempted[23,26] (cf. our patient). T-tube insertion has been reported to establish a controlled fistula in this group of patients.[23]

A combination of optimal primary repair with drainage is believed to offer the best chance of survival, if it is carried out within 24 hours of rupture.[7] However, occasionally primary repair may be successful even after 72 hours.[28] Finally resection of oesophagus may be necessary if underlying oesophagus is severely diseased.[7]

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A recent case in our department

A 76 year old woman gave a history of retrosternal burning pain 3 days before admission, associated with coughing and retching. Despite taking antacid the retching continued for about 2 hours. During the period before admission she ate small amounts of a soft diet with some discomfort. At the time of admission the retrosternal burning discomfort had increased in intensity, she also complained of right-sided pleuritic chest pain, difficulty in swallowing and breathlessness. Furthermore she noticed some facial and neck swelling. There was a previous history of asthma for which she took Salbutamol and Beclomethasone inhalers. She was normally independent in activities of daily living and did not smoke or drink alcohol.

On examination she appeared alert but in discomfort. Her temperature was 38°C, respiratory rate was 28 per minute, oxygen saturation 92% on air, heart rate of 116 beats per minute and blood pressure 140/85 mmHg. Chest examination revealed bilateral wheeze with reduced breath sounds in the lower zones particularly on the right. There was subcutaneous emphysema on the anterior aspect of the chest wall on the right. Her abdomen was soft with normal bowel sounds. Otherwise no abnormality was detected.

An initial diagnosis of acute exacerbation of asthma with a possible pneumothorax was made. She became more unwell, her heart rate increasing to 140 beats per minute, blood pressure falling to 114/64 mmHg and her abdomen became distended, although soft and bowel sounds remained audible. The white cell count was 19.1 (neutrophils 18) and C reactive protein 513. Her chest x-ray showed a moderate sized right sided pleural effusion, surgical emphysema on the right and a hiatus hernia. (Figure 1).

Spiral CT of the thorax, with water soluble oral contrast showed extensive surgical emphysema over the right anterior chest wall together with mediastinal emphysema and contrast external to the bowel lumen, within the mediastinum at and below the level of the carina. (Figure 2).

The patient was started on intravenous cefuroxime and metronidazole and following discussion was transferred to the tertiary cardiothoracic surgical unit where she underwent surgery after 24 hours. Oesophagoscopy revealed a perforation in the distal oesophagus on the right. Limited thoractomy was performed using a right postero-lateral approach. The pleural cavity and mediastinum contained a large amount of bile, gastric juice and pus.

Direct repair of the oesophagus was not feasible, so all effort was made to achieve satisfactory mediastinal and pleural drainage and toilet together with insertion of a chest drain. A laparotomy was also performed to insert a draining gastrostomy and feeding jejunostomy.

Post operatively the patient was treated on the intensive care unit with a prolonged course of antibiotics for the sepsis secondary to mediastinitis and ventilation for respiratory failure – this required prolonged weaning.

At 54 days post-operation her contrast swallow scan showed a persistent oesophageal defect with a blind ended tract but contrast flowed freely into the stomach, and oral fluid was therefore re-introduced. She made a gradual but steady recovery and is currently at home doing well.

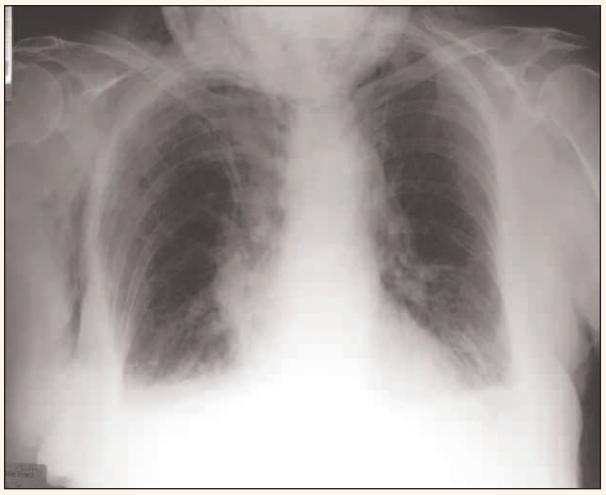

Figure 1. Chest X-ray.

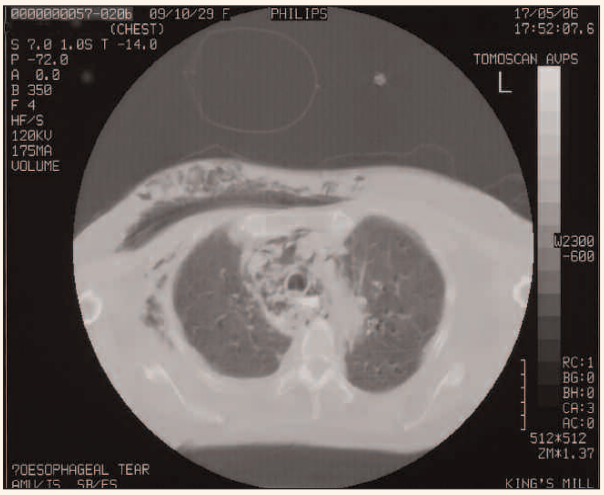

Figure 2. CT scan of the thorax.

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Reference:

1. Curci JJ, Horman MJ. Boerhaave's syndrome: the importance of early diagnosis and treatment. *Ann Surg* 1976; **183**: 401–8.
2. Abbott OA, Mansour KA, Logan WD Jr, Hatcher CR Jr, Symbas PN. A traumatic so-called 'spontaneous' rupture of the esophagus: a review of 47 personal cases with comments on a new method of surgical therapy. *J Thorac Cardiovasc Surg* 1970; **59**: 67–83.
3. Jones WG, Ginsberg RJ. Esophageal perforation. A continuing challenge. *Ann Thorac Surg* 1992; **53**: 534–43.
4. Cirocco WC. Alexander the Great may have died of postemetic esophageal perforation (Boerhaave's syndrome). *Journal of clinical gastroenterology* 1998; **26**: 93–4.
5. Corey M, Ugo AE. Esophageal perforation, rupture and tears [online] 2006 [cited 2007 feb 28]. Available from: URL: http://www.emedicine.com/E,ERG/topic176.htm.
6. Klin B, Berlatzky Y, Uretzky G. Boerhaave's syndrome: case report and review of literature. *Isr J Med Sci* 1989; **25**: 113–5.
7. Praveen KR, Mahammed O. Boerhaave syndrome [online] 2006 [cited 2007 Feb 28]. Available from: URL: http://www.emedicine.com/med/topic233.htm
8. Brauer RB, Liebermann MD, Stein HJ, Bartels H, Siewert JR. Boerhaave's syndrome: analysis of the literature and report of 18 new cases. *Diseases of the oesophagus* 1997; **10**: 64–8.
9. Bldergroen MR, Lowe JE, Postelthwait RW. Diagnosis and recommend management of esophageal perforation and rupture. *Ann thorac Surg* 1986; **42**: 235–9.
10. Lyon WS, Seremetis M, deGuzman VC, Peabody JW. Rupture and perforation of the oesophagus, the case for conservative supportive management. *Ann Thorac Surg* 1978; **25**: 346–50.
11. Tilanus HW, Bossuyt P, Schattenkerk ME, Obertop. Treatment of oesophageal perforation: a multivariate analysis. *Br J Surg* 1991; **78**: 582–5.
12. Janjua KH. Boerhaave's syndrome. *Postgrad Med J* 1997; **73**: 265–70.
13. Callaghan J: The Boerhaave Syndrome. *Br J Surg* 1972; **59**: 41.
14. Bennett DJ, Deveridge RJ, Wright JS. Spontaneous rupture of the esophagus: a review with reports of six cases. *Surgery* 1970; **68**: 766.
15. Kallis P, Belsham Pa, Pepper JR. Spontaneous rupture of the esophagus (Boerhaave's syndrome): conservative versus surgical management. *Journal of the Royal Society of Medicine* 1991; **84**: 690–1.
16. Walker WS, Cameron EWJ, Walbaum PR. Diagnosis and management of spontaneous transmural rupture of the oesophagus (Boerhaave's Syndrome). *Br J Surg*; **72**: 204–7.
17. Jougon JB, Ballester M, Delcambre F, Bride TM, Dromer CEH, Velly JF. Assessment of spontaneous pneumomediastinum: Experience with 12 patients. *Ann Thorac Surg* 2003; **75**: 1711–4.
18. Maunder RJ, Pierson DJ, Hudson LD. Subcutaneous and mediastinal emphysema pathophysiology, diagnosis and management. *Arch Intern Med* 1984; **144**: 1447–53.
19. Newcomb AE, Clarke CP. Sponetaneous pneumomediastinum; a benign curiosity or a significant problem? *Chest* 2005; **128(5)**: 3298.
20. Carolan PL, Vaughan DJ. Pneumomediastinum [online] 2006 [cited 2007 Feb 28]. Available from: URL: http://www.emedicine.com/ped/topic1832.htm
21. Munsell WP: pneumomediastinum. *JAMA* 1967; **202**: 129–33.
22. Bobo WO, Billups WA, Hardy JD. Boerhaave's Syndrome: a review of six cases. *Ann Surg* 1970; **172**: 1034.
23. Altorjay A, Kiss J, Voros A: Non operative management of esophageal perforation. Is it justified? *Ann Surg* 1997 Apr **225(4)**: 415–21.
24. Troum S, Lane CE, Dalton ML. Surviving Boerhaave's syndrome without thoracotomy. *Chest* 1994; **106**: 297–9.
25. Govil Y, Kowalski TE. Esophageal rupture [online] 2004 [cited 2007 Feb 28]. Available from: URL : http://www.emedicine.com/med/topic2811.htm.
26. Thal AP, Hatafuku T. Improved operation for esophageal rupture. *JAMA* 1964; **188**: 826.
27. Keighley MRB, Girdwood RW, Ionescu MI, Wooler GH. Spontaneous rupture of the esophagus. *Br J Surg* 1972; **59**: 649.
28. Sakamoto Y, Tanaka N, et al: Surgical management of late esophageal perforation. *Thorac Cardiovasc Surg* 1997; **45(6)**: 269–72.