Eponyms in medicine revisited

Boerhaave’s syndrome

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Boerhaave’s syndrome or spontaneous oesophageal perforation is a lifethreatening condition which demands early diagnosis and urgent management. Yet despite the pressing necessity for diagnosis, it may go unrecognised, mainly because the presenting picture mimics other common conditions and as a result, the initial crucial intervention is delayed. A careful history with a high index of suspicion of the condition and careful study of the chest film can lead to the institution of life-saving measures, with a rewarding outcome.

Recently, the present author was faced with a patient with this condition, which was missed by junior and senior colleagues from three different specialties, mainly because it simulated right-sided diaphragmatic hernia of small bowel in the chest (box 1). Early diagnosis and surgical intervention could have saved this patient’s life. The unusual modes of presentations of this condition, causing difficulty in diagnosis and management, prompted the author to review the subject in detail.

The term Boerhaave’s syndrome was introduced to distinguish the occurrence of spontaneous transmural oesophageal rupture from the Mallory–Weiss syndrome of haematemesis associated with incomplete oesophageal laceration. The distinction between these two conditions is important, since the diagnosis, management, and prognosis are different. Multiple titles have been applied to Boerhaave’s syndrome, in an effort to clarify the several different mechanisms. ‘Spontaneous rupture of the oesophagus’ is the most common although the condition is never truly ‘spontaneous’ in that there is commonly a precipitating factor. However, the adjective ‘spontaneous’ does indicate that the rupture is not a result of direct trauma, foreign body, or instrumentation. The eponym ‘Boerhaavea’s syndrome’ is still useful as a unifying term in discussion of the clinical manifestations.

History

Boerhaave’s syndrome was first described by a Dutch physician, Hermann Boerhaave in 1724. His patient was a 50-year-old Grand Admiral of The Netherlands, Baron Jan van Wassenaer, who died in 1723 after 18 hours of self-induced vomiting which resulted in oesophageal rupture. In that era it was the custom of glutons to induce vomiting with ippecac-like preparations so that they could immediately eat another meal. At autopsy, Boerhaave recognised the smell of duck flesh and found olive oil and roast duck flesh in the left pleural cavity plus a transverse tear (not the usual linear rent) in the distal oesophagus. His dramatic and memorable classic description of the syndrome enabled the condition to be recognised and he has been quoted extensively in literature. The condition remained a pathological curiosity being largely a post-mortem diagnosis and almost always fatal until the first successful drainage by Frink in 1941 and the first successful closure by Barrett in 1947, more than 200 years after the original description. Barrett, in his masterly review of the condition in 1946, concluded that, contingent on the knowledge that the accident can and does occur, knowledge of the symptomatology, and early diagnosis, surgeons should be able to save at least some of these patients.

Pathology

Most series report a substantial morbidity and mortality as without surgical intervention, spontaneous perforation of the oesophagus is virtually incompatible with life. Delayed diagnosis and different clinical variables such as localisation of perforation, pre-existing medical conditions, condition of the oesophagus, and simultaneous perforation of the parietal pleura, are related to survival. A delay of 12 hours or more between symptoms and operation is associated with a 36% reported mortality while with a delay of 24 hours this almost doubles, to 64%. The most common reason for delayed diagnosis is the fact that spontaneous rupture of the oesophagus often mimics other more common acute cardiothoracic or upper gastrointestinal conditions (box 2).

Summary

Boerhaave’s syndrome or spontaneous oesophageal perforation, is a potentially lethal and frequently elusive medical condition which presents not only a diagnostic but also a therapeutic challenge. It is insuffi

Keywords: Boerhaave’s syndrome, spontaneous oesophageal perforation

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Accepted 27 June 1996
Case report

A 92-year-old woman was admitted to an orthopaedic trauma ward as a case of right-sided intertrochanteric fracture. There were no other injuries on clinical examination. Full blood count showed a white blood cell count (WBC) of 14.2 x 10^9/L and haemoglobin of 12.6 g/dl. Chest X-ray (figure 1) and electrocardiogram on admission did not reveal any abnormal findings. She was operated on the same day and close reduction and internal fixation of the fracture with dynamic hip screw and a plate was achieved.

She had a smooth recovery from anaesthesia and remained stable on the first postoperative day. The next day a low grade fever of 38°C was noticed. Examination of the chest revealed coarse crepitations and wheezing on the right side. She was diagnosed clinically to be suffering from a chest infection. She was advised to have chest physiotherapy and a chest X-ray. Full blood count and blood and sputum cultures were done. Subsequently she had a tachypnoea and signs of peripheral circulatory failure. The blood picture showed a WBC count of 10.6 x 10^9/L, haemoglobin 8 g/dl and haematocrit 0.372. The findings on the chest X-ray were totally unexpected and were reported urgently as a small bowel herniation into the right side of the chest (figure 2).

In the meantime the patient further deteriorated and became agitated and dyspnoic. Her arterial blood gases were disturbed and showed a pCO₂ of 3.94 kPa, pO₂ of 8.59 kPa and bicarbonate of 25.4 mmol/l. She was put on oxygen inhalation and shown to the surgical colleagues. The common opinion was that she was suffering from spontaneous right-sided diaphragmatic hernia with bowel contents.

On the basis of her overall poor general condition and another major illness, a very high operative mortality was predicted and so a conservative approach was adopted. She did not improve and died on the fifth postoperative day. The autopsy findings showed intact diaphragms on both sides and air/fluid in the right pleural cavity with a small perforation in the lower third of the oesophagus on the right side. Detailed review of her chest X-ray revealed a right supraclavicular gas shadow which was missed by everyone and the picture of the diaphragmatic hernia on the chest film led to the diagnostic error with a fatal result.

The cause of the high morbidity and mortality is serious cardiorespiratory embarrassment and a shock-like condition, undoubtedly due to fulminating mediastinitis secondary to the accumulation within the mediastinal and pleural spaces of corrosive gastric juices, enzymes, food and bacteria. This is followed by shock, major fluid losses, and suppuration of the mediastinum and pleural cavities.4 The mediastinal pleura ruptures with the initial insult, but, if it does not, it is digested at a later stage by gastric contents13 which are then drawn into the pleural space by the negative intrathoracic pressure.21 Even if the integrity of the mediastinal pleura is not violated, a sympathetic serous effusion may develop within 48 hours. This occurs on the left side in 75–90% of cases, but can be bilateral in 5–10% of patients5,13 or rarely, as in the present case, on the right. In newborns the perforation is usually on the right, probably because the left side of the oesophagus is adjacent to the aorta in infants. It has been postulated that the oesophageal rupture is caused by a sudden rise in the intraluminal oesophageal pressure produced by actual or suppressed vomiting with full stomach contents being ejected against a closed cricopharyngeus muscle.6 Cadaveric studies confirm that a rapid rise of the order of 5 lb/in² will produce rupture and that this will occur at a lower pressure when the oesophagus is diseased.15,16 There are several reasons for the predilection of the perforation in the distal oesophagus for the left side, including thinning of the musculature of this area, segmental defects in the circular layer, weakening of the wall by entrance of vessels and nerves, anterior angulation of the oesophagus at the left diaphragmatic crus, and lack of adjacent supporting structures.17 The length of the tear varies, the average being 2.24 cm. The tear in the mucosa is usually longer than the muscle tear.18

Clinical presentation

The classic clinical presentation as usually described in the literature is of over-indulgence in food or drink with vomiting followed by severe chest pain, dyspnoea, mediastinal or subcutaneous emphysema and cardiovascular collapse (box 3)13; however, some reviews suggest that the presence of the entire
Boerhaave’s syndrome: classic presentation
- patient with a background of alcoholism or dietary excess
- Meckler’s triad of vomiting, lower chest pain and subcutaneous emphysema
- usual chest X-ray findings: pneumomediastinum and hydro-pneumothorax

Boerhaave’s syndrome: unusual clinical features
- change in voice
- extreme swelling of face and neck
- cold water polydipsia/thirst
- pericarditis
- pneumopericardium
- pneumoperitonum
- proptosis
- low pleural fluid amylase
- ruptured diaphragm with hernia*
*present case

Unusual clinical settings for the occurrence of oesophageal perforation
- absence of pain
- absence of vomiting
- during sleep
- asthma
- food binging
- Heimlich manoeuvre
- hyperemesis gravidarum
- infancy
- laughter
- lifting
- straining at stool
- prolonged coughing and hiccup
- symptomless presentation

complex of symptoms is rare, and therefore reliance on a ‘classic’ presentation may be misleading.6 Unusual clinical features (shown in box 4) should also be kept in mind while evaluating patients with suspicion of oesophageal rupture.

The most striking feature is the excruciating pain which is poorly relieved by narcotics. Usually it is a pleuritic left-sided chest pain which may radiate to the substernal area, epigastrium or back. Swallowing exaggerates the pain and it may cause coughing if there is a pleural tear. Sometimes the patient may collapse and dyspnoea usually develops after the onset of pain. The contributing factors for respiratory impairment are the pain itself, splinting and hydro-pneumothorax or a tension pneumothorax. In reports by Walker et al7 and Michel et al8 pain was a complaint in all cases, but an abdominal location was just as common as the chest. Vomiting is usually thought to cause the perforation, but actual vomiting occurs as a consequence of any episode of Boerhaave’s syndrome, as in the present case. Although only three out of 14 patients in the series of Walker et al complained of dyspnoea, 12 had a history of severe vomiting. Another clinical presentation is the presence of subcutaneous emphysema. Although very helpful, it is rarely present initially, either because perforation has occurred directly into the pleural cavity or because there was no time for it to develop radiographically. Its presence on physical examination varies from 25% to 65% in various reports.45 If pneumomediastinum is present, mediastinal cracking coincident with each heart beat known as Hamman’s crunch may be heard in 20% of cases with a stethoscope and can be mistaken for a friction rub of pericarditis.9 The patient may also present initially with spontaneous pneumothorax with all its characteristic signs. As the inflammatory process subsequently starts in the mediastinum and pleural cavities, most patients become febrile, accompanied by signs of sepsicaemia and haemodynamic instability; however, because of the lack of specificity in the diagnosis of oesophageal perforation is often delayed and/or missed.6 According to Keighly, the incidence of diagnostic error can be as high as 50% as it was in his series of 12 cases.10

Differential diagnosis

The truism that a diagnosis will only be made if it is borne in mind, applies with particular force to this life-threatening condition. The most crucial aspect of clinical diagnosis is to obtain a rapid but complete and accurate history, but not all patients are able to provide one. In such circumstances, the relatives in attendance and other people around should be interviewed, but the history may be misleading, without an episode of preceding emesis, and the rupture can occur in patients while asleep, bending over, watching television, or drinking water.22 Therefore it is always helpful in diagnosis to be aware of the diversity of circumstances that may predispose to oesophageal perforation (box 5).

The main cause of diagnostic error is the failure to consider oesophageal perforation in the diagnostic hypothesis. The condition can easily mimic other acute abdominal and cardiothoracic conditions (box 2). The most frequently mimicking diagnosis is the perforated peptic ulcer which can be differentiated from Boerhaave’s syndrome by a history of ulcer, pneumoperitoneum and a gradual increase in the severity of abdominal symptoms in most cases, compared to the chest findings. Both types of perforation require surgery, but ulcer disease demands laparotomy while oesophageal perforation is best approached through the chest. The other common diagnostic error is to confuse oesophageal rupture with myocardial infarction.25,26 In the case described by Mansour and Teaford,19 there was oesophageal rupture into the pericardium. It has been remarked by Hampton25 in a discussion of chest pain and breathlessness, that a coronary care unit can be a dangerous place for a patient with chest pain that is not due to ischaemic heart disease. The coronary care unit staff must continually be reminded of the need to monitor noncardiac problems. The high incidence of coronary artery disease and the general awareness that chest pain may be harbinger of sudden cardiac death may divert attending physicians from the rigorous formulation and concurrent testing of other diagnostic hypothesis. The presentation of Boerhaave’s syndrome can also be easily confused with acute pancreatitis which is often associated with alcohol abuse, vomiting, left pleural effusion, and chest and abdominal pain. A normal serum amylase makes pancreatitis unlikely.

It is also important to differentiate pericarditis and pulmonary embolism from Boerhaave’s syndrome. Hamman’s ‘mediastinal crunch’ of Boerhaave’s syndrome can sound like a pericardial friction rub and patients with pericarditis may also report chest pain when sitting forward as well as with pericarditis. The manifestations of pulmonary embolus (dyspnoea, tachycardia, cyanosis, chest pain and circulatory collapse) have striking similarity to the presentation of oesophageal rupture and represent

Postgrad Med J: first published as 10.1136/pgmj.73.859.265 on 1 May 1997. Downloaded from http://pmj.bmj.com/ on May 21, 2022 by guest. Protected by copyright.
a very dangerous trap. Other thoracic diseases such as dissecting aortic aneurysm, spontaneous pneumothorax and pneumomediastinum should be ruled out since thoracotomy plays a limited role in their usual management. Clinical findings and radiological investigations allow them to be differentiated. The list of differential diagnoses is vast but a limited study of the upper gastrointestinal tract with contrast material in the emergency room can quickly separate Boerhaave's syndrome from other more common disease entities.

**Diagnosis**

Once oesophageal perforation is suspected, X-ray is the most valuable modality and the simple erect film of the chest yields the most information (box 6), although 12% of patients may have normal findings. The most common error in diagnostic work up is the failure to obtain a chest X-ray when Boerhaave's syndrome presents as an 'acute abdomen.' The most common findings are pleural effusions and pneumothorax with incidences of 91% and 80%, respectively. Pneumothorax is usually associated with pleural effusion and is unilateral but can be bilateral in a few cases. Subcutaneous emphysema in the soft tissue of the neck or chest wall and the mediastinal air is seen in 66% of patients. Mediastinal air should be carefully looked for, as it is easily missed if it is retrocardiac. Widening of the mediastinum is occasionally seen and the so-called 'V-sign of Nacelero' is another subtle X-ray finding which is easily overlooked. In reponse air in the fascial planes of the mediastinum and diaphragmatic pleura behind the heart and is caused by dissection by mediastinal emphysema. A postero-anterior view of the mediastinum may show air to be present before other abnormalities are detectable. Therefore it is an early and reliable finding as it occurs in 20% of cases. In addition to these findings on the erect chest film, a perforated duodenal or gastric ulcer must be excluded, although Boerhaave's syndrome has been reported in association with pneumoperitonium and pneumopericardium.

Mediastinal emphysema takes at least one hour to develop and the chest X-ray will remain normal in 10–12% of patients. Therefore, whether or not mediastinal emphysema is detected, it is advisable to do a contrast study of the oesophagus if there is clinical suspicion of rupture. Contrast studies are positive in 75% of cases, however, so a lack of extravasation does not necessarily exclude a perforation. In such cases the examination should be repeated if clinically advisable. It is believed that false negative oesophagograms occur because the material may be too viscous (eg, barium) to leak out, passage is too rapid (eg, gastrograffin), or the perforation is blocked by oedema, clots, or food particles. One should always obtain oblique views if the antero-posterior and lateral projections are negative to ensure that a leak is not being hidden on the film by contrast material within the lumen.

The majority of patients will show hydropneumothorax on chest film. In such patients, thoracocentesis will be diagnostic if food particles or gastric juice with a pH less than 6 in aspirate with a high amylase content or squamous cells from saliva is found. Drainage through a chest tube of previously swallowed 'tracer' fluids such as methylene blue or barium may confirm the diagnosis in some patients. In patients in whom oesophageal perforation is suspected clinically but the contrast studies are negative, computed tomography (CT) can be a good and useful diagnostic adjunct. Moreover thoracic CT can be complementary to a positive oral contrast study by localising collections of fluid for surgical drainage and it can also be valuable in patients too ill to cooperate in an oral contrast study. Its principal weakness is an inability to locate the exact site of perforation. CT findings suggestive of oesophageal perforation are listed in box 7. CT scans are also useful in follow up after initiation of therapy and in the evaluation of patients who fail to improve despite either operative or nonoperative management.

A perplexed and worried physician attending a very ill patient with an uncertain diagnosis usually has two recourses: to consult with colleagues and to turn to the published literature. With regard to the first option, it is of interest that three clinical specialists and three residents had been consulted in the present case, without the correct diagnosis being reached. It is important to realise that a diagnosis of oesophageal rupture is best considered during the initial contact with the patient as it may become more elusive with the passage of time. In 1965, Levine and Kelley noted that fewer than 35% of reported cases of spontaneous oesophageal perforation had been diagnosed before death and that many were not even suspected prior to post-mortem examination.
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**Factors to be considered in selecting appropriate therapy**

- size and location of perforation
- presence of underlying oesophageal disease
- interval between perforation and diagnosis
- condition of oesophagus
- extent of soilage or injury to adjacent organs and tissues
- age and general condition of the patient

**Principles of operative management**

- wide debridement of oesophageal and mediastinal tissue
- proper and thorough cleansing of pleural cavity
- correction or elimination of any distal obstruction
- two-layer closure of perforation, if necessary reinforce with other tissues
- provision of broad-spectrum antibiotic cover and elective ventilation, if required.

**Learning points**

- spontaneous oesophageal perforation is a life-threatening condition and if diagnosed early, life may be saved with urgent and aggressive management
- a careful history with a high index of suspicion and careful study of chest film will give important clues to diagnosis
- a delay of 12 hours or more between symptoms and operation is associated with a 36–64% mortality
- the common reason for delayed diagnosis is that it always mimics other more common acute cardiothoracic or upper gastrointestinal conditions
- total reliance on a ‘classic’ presentation may be misleading
- limited erect chest film and limited study of upper gastrointestinal tract with contrast material can quickly separate Boerhaave’s syndrome from other more common disease entities
- in patients with suspicion of oesophageal perforation but negative contrast studies, thoracic CT is a useful diagnostic modality
- in most patients improved overall results can only be attained by early diagnosis and aggressive surgery

**Treatment**

Despite numerous reports documenting a wide range of experience in the treatment of oesophageal perforation, recommendations regarding treatment remain controversial. Selected patients can be treated nonoperatively but, in most patients, improved overall results can only be attained by aggressive surgery utilizing one of a variety of techniques. Shock is not a contraindication to operation in these patients; rather, it underscores the necessity for surgical intervention, for upon opening the mediastinum and release of tamponade, blood pressure is often revived. The patients selected for conservative treatment are those with small perforations, contamination confined to the mediastinum, and late recognition (more than 24 hours) of an oesophageal perforation, as the initial mortality rate is no more than 24 hours.41 The more conservative approach.40 Drainage of the mediastinum by per oesophageal nasogastric tube through the oesophageal rent, using it as a drain and means of antibiotic delivery, and other more elaborate tube systems have been described. Rare patients have demonstrated adequate internal drainage of a mediastinal abscess back into the oesophagus and have recovered without tube thoracostomy.49

Once the diagnosis of spontaneous oesophageal perforation is established, several factors are critical in determining the appropriate operative treatment (box 9). Hence the management must be individualised. The different options for operative management are: primary closure, reinforced primary closure, resection of the oesophagus, drainage alone, T-tube drainage, exclusion and diversion, and intra-luminal stent. Whichever mode of treatment is selected, the primary aim of the treatment must be to prevent further soilage from the perforation, to eliminate the infection produced by soilage, to restore the integrity and continuity of the gastrointestinal tract, and to restore and maintain adequate nutrition. Furthermore, irrespective of the operative technique adapted for the closure of perforation there is no doubt that it should be accompanied by thorough cleansing of the pleural cavity, debridement of devitalized tissues, correction or elimination of any distal obstruction, prolonged gastric drainage (preferably through gastrostomy rather than nasogastric tube), and expansion of the lung by pleural drainage, supplemented if necessary with elective ventilation and decortication. As a basic principle, samples of pleural and mediastinal fluid should be taken for bacteriological culture and antibiotic sensitivity, and nutrition maintained preferably via an enteral route, such as jejunostomy, rather than total parenteral nutrition.

Most spontaneous perforations occur in the distal oesophagus on the left side, therefore preferable exposure is by left thoracotomy in the 7th or 8th intercostal space.4 Once the oesophagus is exposed, localisation of subtle perforations may require instillation of methylene blue into the oesophageal lumen or the insufflation of air into the oesophagus immersed in saline solution. The mucosal layer usually remains healthy. However, a myotomy may be necessary to visualise the full extent of the mucosal injury and to facilitate its repair. Inadequate exposure and repair of the mucosal defect can predispose to leakage from the closure. Therefore the length of the tear in each component of the oesophageal wall should be separately established and repair carried out in two layers.

The most common and feared complication of oesophageal closure is leakage from the suture line with subsequent empyema or oesophagocutaneous fistula. The factors contributing to this complication are a poor blood supply, absence of a protective omentum, lack of a serosal layer, and friable submucosa. It is for this reason that many surgeons have suggested buttressing the primary repair in some way. In Reeder et al’s series of 33 patients, however, no specific therapy was clearly superior. The tissues used to reinforce the suture line have included pleural flap,46 omentum, a pedicled intercostal muscle flap,47 pericardial fat pad flap,48 flap of diaphragm,49 fundus of stomach50 and even portions of lung.51 Regardless of the type of material used, secure closure necessitates suturing the reinforcing tissue closely, as for an anastomosis, rather than just simple ‘taking’. More complex and invasive techniques to minimize the leakage, like T-tube insertion into the site of rupture and a variety of defunctioning procedures have been described but they incur the trauma of major surgery and all rely upon natural closure of the oesophageal tear. Urschel and others have described exclusion and diversion or even oesophageal resection in the treatment of complicated oesophageal perforations.52,53

In spite of all the operative measures taken to prevent perforations, if the suture line, it can still occur with the formation of empyema or oesophagocutaneous fistula. If it does occur, it will usually close if there is no distal obstruction, local infection, foreign body, malignant change, or epithelialisation of the tract. The
progress of the oesophageal fistula may be followed by daily drainage and serial contrast swallows. Operative intervention is necessary if healing has not started after several weeks; the leaking segment should then be by-passed and excluded or resected, as appropriate.