

# Boerhaave's Syndrome or Spontaneous Perforation of the Oesophagus

## Review Article

A. Marinis, S. Rizos

Received 03/06/2011 Accepted 11/07/2011

### Abstract

Spontaneous perforation of the oesophagus, also called Boerhaave's syndrome, is a barogenic rupture caused by a sudden rise in intraluminal pressure, usually in the distal oesophagus. Early diagnosis of spontaneous oesophageal perforation is critical to the survival of the patient. Sudden and very intense chest or epigastric pain after an abrupt increase of oesophageal intraluminal pressure is the most characteristic clinical symptom and should raise suspicion for Boerhaave's syndrome. Plain radiographs demonstrate the presence of free air in the mediastinum, subcutaneous emphysema or air subdiaphragmatically, as well as pleural effusions. Upper gastrointestinal series reveal leakage of the contrast and confirms the location of the perforation. Similarly, chest computed tomography demonstrates the presence of an air-fluid level, pneumomediastinum and pleural effusions and, more importantly, the complications related to oesophageal perforation. Oesophageal perforation is associated with a high mortality rate, suggested to be approximately 2% for every hour after initial presentation. More specifically, diagnosis and intervention within the first 12 hours results in 80-90% survival, which falls to 70-75% between 12-24 hours; after 24 hours, survival decreases to less than 50%, and after 48 hours to less than 10%. Operative treatment of Boerhaave's syndrome mainly includes a complete debridement and lavage of infected and necrotic tissues with wide drainage of the mediastinum and pleural cavities and either primary oesophageal repair and reinforcement (if diagnosed early) or T-tube diversion or resection (if diagnosis is delayed). Early confirmation of the diagnosis is very important because it allows earlier support of the critically ill patient and prompt operative intervention when necessary. Eventful outcome of patients is related to these

early events in conjunction with the good general condition of the patient. Otherwise, the patient is virtually condemned to die..

### Key words:

Oesophageal perforation, Boerhaave's syndrome, Spontaneous, Post-emetic, Barogenic.

### Introduction

Spontaneous perforation of the oesophagus (SPE), also called Boerhaave's syndrome, is the perforation of the oesophagus after forceful vomiting and is considered as a surgical emergency. This syndrome was first described by the Dutch surgeon Hermann Boerhaave in 1724, who observed a transmural rupture of the distal oesophagus in a post-mortem examination of Baron John von Wassenaeur [1]. Though rare, this type of oesophageal perforation is life-threatening due to the development of mediastinitis and severe sepsis. Patients usually present with non-specific symptoms, such as epigastric pain and vomiting; this leads to a delay in diagnosis and appropriate treatment.

### Aetiology - Pathophysiology

Boerhaave's syndrome is a barogenic rupture caused by a sudden rise in intraluminal pressure, usually in the distal 3-6 cm of the oesophagus. It is described more commonly in males than females at a ratio of 4:1 with a median age of 64 years [2]. This syndrome generally occurs in the absence of pre-existing oesophageal pathology and is commonly associated with large meals and heavy consumption of alcohol. Pathophysiologically, the combination of a sudden rise in intra-abdominal pressure caused by vomiting or retching and neuromuscular incoordination provokes the failure of the cricopharyngeal muscle to relax, leading to a dramatic increase in the intraluminal oesophageal pressure. However, cases of SPE resulting from trauma, weightlifting, parturition, defaecation, the Heimlich manoeuvre and status epilepticus have also been reported [3]. A transmural rupture is usually observed in the lower third of the oesophagus (90%) and less fre-

A. Marinis (Corresponding author), S. Rizos  
- First Department of Surgery, Tzaneion General Hospital Piraeus  
Greece  
✉ e-mail: drmarinis@gmail.com

quently in the mid (8%) and upper (2%) thirds. The left lateral side of the lower oesophageal wall is the usual site of perforation due to anatomical weakness of this point. The length of the tear varies from 0,6-8,9 cm (average 2,2 cm) with the mucosal injury being longer than the muscular tear [4]. With the progress of time, the negative intrathoracic pressure progressively draws more air, fluids and foods from the oesophageal lumen into the mediastinum and the pleural cavities, and a chemical and septic pneumomediastinitis develops.

### Clinical presentation

The classic clinical presentation of Boerhaave's syndrome is the Mackler's triad, consisting of vomiting or retching, severe lower chest pain and cervical subcutaneous emphysema. In a large series, however, this classic triad was present only in the 14% of the patients [5]. The most important clinical feature is a sudden, often "dramatic" in intensity, chest pain following an episode of raised intra-abdominal pressure, most commonly after forceful vomiting. Pain is located retrosternally and is usually exacerbated by movement and respiration. Typically, haematemesis is absent, which distinguishes it from the Mallory-Weiss tear, and swallowing precipitates cough and pleuritic pain because of communication of the oesophagus with the pleural cavity. Clinical examination reveals a patient with tachypnoea and orthopnoea, tachycardia, fever, diaphoresis and hypotension, with bilateral pleural effusions. Mediastinal emphysema can be audible on auscultation or visible on chest x-rays and precedes subcutaneous emphysema, which is seen in 28-60% of patients at initial presentation. Another rare presentation is Anderson's triad, referring to subcutaneous emphysema, rapid respirations and abdominal rigidity. Abdominal pain and tenderness is found in about 60% of patients, leading to a negative laparotomy in 9% of cases [6].

### Differential diagnosis, errors and prognosis

Diagnostic errors are actually very frequent with the most common misdiagnosis being the perforated ulcer, followed by myocardial infarction, pulmonary embolism, dissecting aneurysm and pancreatitis [7]. The diagnostic error is generally over 50%, with a delay of more than 12 hours in the majority of cases, with only 5% diagnosed initially at presentation and about 35% correctly diagnosed pre-mortem [8]. SPE is associated with a high mortality rate, suggested to be approximately 2% for every hour after initial presentation. More specifically, diagnosis and intervention within the first 12 hours results in

80-90% survival. This rate falls to 70-75% between 12-24 hours; after 24 hours survival, it decreases to less than 50% and after 48 hours to less than 10% [9-11].

### Diagnostic work-up

Diagnostic workup includes chest x-rays, a water-soluble contrast swallow, chest computed tomography (CT) and upper gastrointestinal (GI) endoscopy.

### Radiology

- A plain chest x-ray usually shows pneumomediastinum in 10-20% of cases (Naclerio's V-sign), subcutaneous emphysema or air subdiaphragmatically and pleural effusions (Fig. 1), commonly left-sided, which develop immediately from the oesophageal leakage, or at a later phase sympathetically from adjacent mediastinitis.



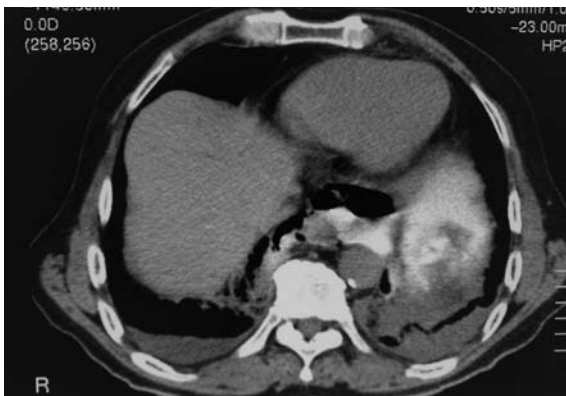
**Fig. 1** Chest radiograph showing a large left pleural effusion (from our archives).

- Oesophagogram is performed using barium for a suspected thoracic perforation and Gastrografin® for an abdominal perforation. Barium is inert in the chest but causes peritonitis in the abdomen, whereas aspirated Gastrografin® can cause life-threatening pneumonitis. Usually the contrast media is extravasated into the pleural cavity (Fig. 2), showing the location of perforation and facilitating the decision concerning the surgical approach (abdominal vs. thoracic). However, contrast studies are associated with a negative false rate of 27-66% [12].
- Chest CT demonstrates an air-fluid level at the site of perforation, pleural effusions, pneumomediastinum and subcutaneous emphysema, highly suggesting the site of perforation (Fig. 3). This diagnostic modality, however, is commonly used post-operatively in order to assess the development of complications (pneumonitis, mediastinitis, empyema, abscesses) and to control the adequacy of therapeutic interventions (resolving mediastinitis,

adequate drainage of the mediastinum and pleural cavities, etc).



**Fig. 2** Swallow with a non-ionic water-soluble contrast agent (Iopamiro®) showing leakage from the lower thoracic esophagus and the development of an air-fluid level (from our archives).



**Fig. 3** Chest computed tomography demonstrating the leakage of the contrast into the lower mediastinum, the concomitant presence of free air and bilateral pleural effusions (from our archives).

### Endoscopy

Upper GI endoscopy with flexible endoscopes is currently considered to have a higher diagnostic sensitivity than contrast radiology in Boerhaave's syndrome, allowing accurate assessment of the location of perforation and mucosal extent of the tear, as well as safe placement of a nasojejunal feeding tube for enteral feeding.

Finally, preoperative investigations can include the oral ingestion of dyes, such as methylene blue, as well as thoracocentesis, which reveal frank gastric contents on aspiration or a pleural fluid with a low pH (less than 6.0) and a high amylase level.

### Management

Resuscitation of the patient with spontaneous oesophageal perforation includes airway control and proper oxygenation, large-bore intravenous catheters and aggressive fluid resuscitation, invasive haemodynamic monitoring (central venous and arterial catheters), inotropic agents, and monitoring of urine output. Management can be either non-operative or surgical, depending on time delay in presentation and diagnosis, extent of perforation and the overall general condition of the patient.

modynamic monitoring (central venous and arterial catheters), inotropic agents, and monitoring of urine output. Management can be either non-operative or surgical, depending on time delay in presentation and diagnosis, extent of perforation and the overall general condition of the patient.

### Non-operative management

A contained perforation diagnosed early, with limited soiling and without any distal GI obstruction, in a patient without signs or symptoms of mediastinitis or sepsis and without other medical co-morbidities, and a delayed diagnosis which is well tolerated, are the usual indications for non-operative treatment [13, 14]. The latter involves aggressive resuscitation and close vital sign monitoring in an intensive care unit (ICU) environment, along with measures such as: nil per os (NPO), intravenous anti-secretory agents (H2-receptor antagonists or proton pump inhibitors), broad-spectrum antibiotics and anti-fungal agents, naso-gastric decompression through a nasogastric tube safely placed during endoscopy and adequate chest drainage of pleural effusions. Successful attempts to seal the oesophageal leak with covered, self-expandable stents endoscopically placed have been described in the literature [15].

### Operative management

The primary goal of operative intervention is to manage the perforation site in order to prevent ongoing spillage of oesophageal contents, to completely debride the infected and necrotic tissues with concomitant wide drainage of the mediastinum and pleural cavities and, finally, to restore oesophageal integrity and gastrointestinal continuity. Interestingly, thorough debridement and irrigation are more important manoeuvres in terms of survival than the type of reconstruction [16]. In general, primary repair and reinforcement usually follow if diagnosis is established early and t-tube diversion and resection if diagnosis is delayed beyond 24 hours.

- *Primary repair and reinforcement.* Primary repair, first employed by Barrett in 1947 [17], consists of a precise two-layer closure of the perforation with 2/0 or 3/0 interrupted absorbable sutures, followed by reinforcement of the suture line with adjacent healthy tissue (pleura, lung, diaphragm, intercostal muscle, gastric fundus, omentum) or even absorbable mesh [18]. However, primary repair is associated with a high leakage rate of up to 23% if treatment is delayed beyond 24 hours, rising to 50% in the presence of sepsis [19, 20].

- *T-tube diversion.* Stable patients presenting late in the course of the disease with an established

oesophago-pleural communication and localized sepsis are not usually treated by primary repair, but can be managed instead by diverting the oesophageal contents via a T-tube in a controlled manner [21]. The tube is left in place for 3-6 weeks until a fistula tract has developed.

- *Oesophagectomy*. In delayed diagnosis with concomitant sepsis resection of the severely damaged oesophagus with immediate (if contamination is minimal) or delayed reconstruction is the method of choice. This operative approach is associated with a high morbidity and mortality rate and reserved for critically ill septic patients. Techniques that simply exclude and divert the injured organ proximally and distally, leaving in situ the oesophagus, have been abandoned and are now historical.

### Conclusions

Early diagnosis of spontaneous oesophageal perforation is critical to the survival of the patient. Sudden and very intense chest or epigastric pain after an abrupt increase of oesophageal intraluminal pressure is the most characteristic clinical symptom and should raise suspicion for Boerhaave's syndrome. Early confirmation of the diagnosis allows earlier support of the critically ill patient and prompt operative intervention when necessary. Eventful outcome of patients is related to these early events in conjunction with the good general condition of the patient. Otherwise, the patient is virtually condemned to die.

### Conflict of interest

The authors declare that they have no conflict of interest.

### References

- Derbes VJ, Mitchell RE. Hermann Boerhaave's Atrocis, nec Descripti Prius. *Morbi Historia: the first translation of the classic case report of rupture of the esophagus, with annotations*. Bull Med Libr Assoc. 1955;43:217.
- Brauer RB, Liebermann-Meffert D, et al. Boerhaave's syndrome: analysis of the literature and report of 18 new cases. *Dis Esophagus* 1997;10:64-8.
- Mackler S. Spontaneous rupture of the oesophagus; an experimental and clinical study. *Surg Gynecol Obst* 1952;93:345-56.
- Kossick PR. Spontaneous rupture of the oesophagus. *S Afr Med J* 1973;47:1807-9.
- Griffin SM, Lamb PJ, Shenfine J et al. Spontaneous rupture of the esophagus. *Br J Surg* 2008; 95:1115-20.
- Shenfine J, Dresner SM, Vishwanath Y, et al. Management of spontaneous rupture of the oesophagus. *Br J Surg* 2000;87:362-73.
- Symbas PN, Hatcher CR, Harlaftis N: Spontaneous rupture of the esophagus. *Ann Surg* 1978;187:634-9.
- Abbott OA, Mansour KA, Logan WA Jr, et al. Atraumatic 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.
- Brewer LA, Carter R, Mulder GA, Stiles QR. Options in the management of perforations of the esophagus. *Am J Surg* 1986;152:62-9.
- Henderson JA, Peloquin AJ. Boerhaave revisited: spontaneous esophageal perforation as a diagnostic masquerader. *Am J Med* 1989;86:559-67.
- Richardson JD, Martin LF, Borzotta AP, Polk HC Jr. Unifying concepts in treatment of esophageal leaks. *Am J Surg* 1985;149:157-62.
- Buecker A, Wein BB, Neueberg JM, et al. Esophageal perforation: comparison of use of aqueous and barium-containing contrast media. *Radiology* 1997; 202: 683-6.
- Ivey TD, Simonowitz DA, Dillard DH, Miller DW Jr. Boerhaave syndrome. Successful conservative management in three patients with late presentations. *Am J Surg* 1981;141:531-3.
- Cameron JL, Kieffer RF, Hendrix TR, et al. Selective non-operative management of contained intrathoracic esophageal disruptions. *Ann Thorac Surg* 1979;27:404-8.
- Yuasa N, Hattori T, Kobayashi Y, et al. Treatment of spontaneous esophageal rupture with a covered self-expanding metal stent. *Gastrointest Endosc* 1999;49:777-80.
- Altorjay A, Kiss J, Voros A, et al. The role of esophagectomy in the management of esophageal perforations. *Ann Thorac Surg* 1998; 65:1433-6.
- Barrett N. Report of a case of spontaneous perforation of the oesophagus successfully treated by operation. *Br J Surg* 1947; 47: 216-8.
- Bardaxoglou E, Manganas D, Meunier B, et al. New approach to surgical management of early esophageal thoracic perforation: primary suture repair reinforced with absorbable mesh and fibrin glue. *World J Surg.* 1997 ;21:618-21.
- Wright CD, Mathisen DJ, Wain JC, et al. Reinforced primary repair of thoracic esophageal perforation. *Ann Thorac Surg* 1995;60:245-9.
- Lawrence DR, Ohri SK, Moxon RE, et al. Primary esophageal repair for Boerhaave's syndrome. *Ann Thorac Surg* 1999;67:818-20.
- Mansour KA, Wenger RK. T-tube management of late esophageal perforations. *Surg Gynecol Obstet* 1992; 175:571-2.

## Σύνδρομο Boerhaave ή Αυτόματη Ρήξη του Οισοφάγου

### Άρθρο Ανασκόπησης

Αθανάσιος Μαρίνης, Σπύρος Ρίζος

#### Περίληψη

Ως αυτόματη ρήξη του οισοφάγου, επίσης γνωστή ως σύνδρομο Boerhaave, θεωρείται η βαρο-τραυματική ρήξη που συμβαίνει μετά από αφνίδια αύξηση της ενδοαυλικής πίεσης συνήθως στο περιφερικό τμήμα του οισοφάγου. Η πρόωμη διάγνωση της αυτόματης ρήξης του οισοφάγου αποτελεί τη πιο κρίσιμη παράμετρο που καθορίζει την επιβίωση του ασθενή. Η εμφάνιση αφνίδιου και ιδιαίτερα έντονου θωρακικού ή επιγαστρικού άλγους μετά από βίαιη αύξηση της ενδο-οισοφαγικής πίεσης είναι το πιο χαρακτηριστικό σύμπτωμα και θα πρέπει να θέτει την υπόνοια του συνδρόμου Boerhaave. Οι απλές ακτινογραφίες αναδεικνύουν τη παρουσία ελεύθερου αέρα στο μεσοθωράκιο, υποδόριο εμφύσημα ή ακόμα κι αέρα υποδιαφραγματικά, καθώς και πλευριτικές συλλογές. Η διάβαση του ανώτερου πεπτικού αναδεικνύει τη διαφυγή του σκιαγραφικού από τον οισοφάγο καθώς και την εντόπιση του πιθανού σημείου ρήξης. Παρομοίως, η αξονική τομογραφία θώρακος επιβεβαιώνει τη παρουσία υδραερικού επιπέδου, πνευμομεσοθωρακίου και πλευριτικών συλλογών, αλλά εξίσου σημαντική είναι και η συμβολή της στη διάγνωση των επιπλοκών απότοκων της ρήξης του οισοφάγου. Η ρήξη του οισοφάγου σχετίζεται με υψηλή θνητότητα, η οποία αυξάνεται κατά περίπου 2% για κάθε ώρα από την έναρξη των συμπτωμάτων. Ειδικότερα, η πρόωμη διάγνωση κι έγκαιρη παρέμβαση εντός των πρώτων 12 ωρών συνδέεται με επιβίωση 80-90%, η οποία όμως ελαττώνεται στο 70-75% όταν η παρέμβαση γίνει στο διάστημα 12-24 ωρών. Δυστυχώς η επιβίωση ελαττώνεται κάτω από 50% μετά τις 24 ώρες, ενώ μετά από 48 ώρες πέφτει κάτω από 10%. Η χειρουργική θεραπεία του συνδρόμου Boerhaave περιλαμβάνει κυρίως τον επαρκή καθαρισμό κι έκπλυση των επιμολυσμένων και νεκρωτικών ιστών, με ταυτόχρονη ευρεία παροχέτευση του μεσοθωρακίου και των υπεζωκοτικών κοιλιοτήτων, καθώς και τη πρωτογενή συρραφή με ενίσχυση της

γραμμής συρραφής (όταν η διάγνωση γίνεται νωρίς) ή την εκτροπή με σωλήνα T και την οισοφαγεκτομή (σε καθυστερημένη διάγνωση). Η πρόωμη επιβεβαίωση της διάγνωσης ακολουθείται από ταχύτερη υποστήριξη του βαρέως πάσχοντος ασθενή και από άμεση χειρουργική παρέμβαση όταν κρίνεται σκόπιμη. Η καλή πρόγνωση του ασθενή σχετίζεται με αυτά τα πρώτα γεγονότα, σε συνδυασμό με τη καλή γενική κατάσταση του ασθενή. Ειδικά, ο ασθενής είναι σχεδόν καταδικασμένος να πεθάνει.

#### Λέξεις κλειδιά

Διάτρηση οισοφάγου, Σύνδρομο Boerhaave, Αυτόματη διάτρηση, Μεθεμεικική διάτρηση, Βαρότραυμα.