Spontaneous rupture of the oesophagus - Boerhaave's syndrome

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Abstract. *Spontaneous oesophageal rupture is a rare affection resulting from barotrauma to the oesophagus during forceful protracted vomiting. The clinical presentation of oesophageal rupture is dependent upon the size and the location of the injury and time course since the outset. Clinical diagnosis, in typical cases, is based on the symptom triad: effort vomiting, sudden chest pain and subcutaneous emphysema. Positive diagnosis is established by means of water-soluble contrast swallow or based on computer tomography scan. Endoscopy has a limited role in diagnosis because an endoscope and insufflation of air can extend the perforation. Boerhaave's syndrome is a surgical emergency and has a lethality rate of more than 90 % in the absence of prompt diagnosis and treatment. The authors describe the case of a 71-year-old male with atypical presentation of Boerhaave's syndrome in the lower abdominal part of oesophagus without typical clinical findings. The patient was referred for severe vomiting and for suspicion of volvulus of the stomach. The perforation of the oesophagus was identified by means of endoscopy. Surgical treatment was complicated by a postoperative leak, which was successfully managed with drainage and enteral feeding via a fine bore nasojejunal tube.*

Keywords: *spontaneous oesophageal perforation, Boerhaave's syndrome*

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Souhrn. *Spontánní ruptura jícnu je vzácná afekce vzniklá při náhlém vzestupu tlaku v jícnu v průběhu silného protrahovaného zvracení. Klinická manifestace ruptury jícnu závisí na velikosti a lokalizaci poranění a na době od vzniku ruptury. Diagnóza je v typických případech postavena na triádě příznaků: usilovné zvracení, náhle vzniklá bolest na hrudi a podkožní emfyzém. Pozitivní diagnóza je založena na pasáži jícnem s vodním kontrastem nebo na počítačové tomografii. Role endoskopie v diagnostice je limitována, protože endoskop samotný i insuflace vzduchu můžou zvětšit rozsah perforace. Boerhaaveho syndrom je chirurgickou náhlou příhodou, která má bez rychlé diagnostiky a terapie vysokou letalitu nad 90 %. Autoři popisují případ 71-letého muže s atypickou prezentací Boerhaaveho syndromu v dolní abdominální části jícnu bez typických klinických známek. Pacient byl přijat pro protrahované těžké zvracení a pro podezření na volvulus žaludku. Perforace jícnu byla diagnostikována pomocí endoskopie. Chirurgická léčba byla komplikovaná pooperační dehiscencí sutury, která byla úspěšně zvládnuta perioperačně zavedenou drenáží a úplnou parenterální, později enterální výživou pomocí tenké nasojejunální sondy.*

Klíčová slova: *spontánní perforace jícnu, Boerhaaveho syndrom*

Spontaneous rupture of the oesophagus was initially described by Herman Boerhaave in the fatal case of baron Jan van Wassenaar, Grand Admiral of the Dutch Fleet and Prefect of the Rhineland (Atrocis nec prius descripti morbi historia illustrissimi baronis Wassenariae, 1724). This condition remained a pathological curiosity until a subsequent case arose with successful surgical treatment, which was reported by Barrett in 1946 (1,5).

Case report

A 71-year-old male, heavy drinker, with a history of ischaemic heart disease and diabetes mellitus was admitted to a surgery department elsewhere because of severe protracted vomiting (later with repeated haematemesis) and abdominal pain lasting 3 days. Upper GI endoscopy done elsewhere showed a lot of blood clots in a hollow (that was considered to be an abnormal stomach), the endoscopist was unable to distinguish pylorus and the patient was transferred under suspicion of gastric volvulus to the Department of Surgery at our Teaching Hospital. On admission, the patient was in good condition in general, physical examination was normal except for tachycardia (heart rate 110/min.) and epigastric tenderness. Blood pressure was 110/60 mm Hg, body temperature was normal, laboratory tests included anaemia (haemoglobin 98 g/L and haematocrit 28.4 %), basic biochemical parameters were normal except for mild hyperglycaemia (9.5 mmol/L). Abdomen radiograph excluded pneumoperitoneum, chest radiograph was not performed. The surgeon indicated new endoscopy for bleeding from the nasogastric tube. After insertion of the endoscope, the oesophagus was conspicuously collapsed, insufflation was practically impossible, there were typical linear mucosal erosions of reflux oesophagitis in the lower part of the oesophagus (Fig. 1). A very interesting view was in the distal abdominal part of the oesophagus (at a distance of 40 centimetres from the incisors), which continued over the orifice 2 to 3 centimetres in diameter into the large cavity approximately 6 cm in diameter with a lot of blood clots. The wall of the cavity was atypical and did not reveal a view of the normal gastric wall. The endoscopic picture was under strong suspicion of oesophageal perforation. Clearly visible defect of the muscular layer of the oesophageal wall was shown in the perforation orifice after wash off (Figs. 2 to 4). By the end of examination, normal gastro-oesophageal junction and cardia were identified successfully, the gastroscope freely passed into the stomach with little quantity of sanguinolent contents, while pylorus,

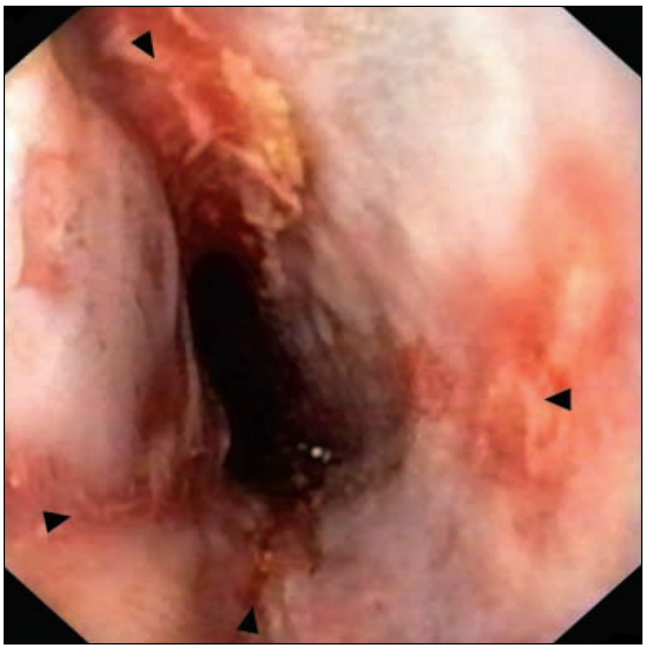

Fig. 1
Having inserted the gastroscope into the oesophagus, longitudinal mucosal erosions were found in the distal half of the oesophagus as endoscopic signs of pre-existing reflux oesophagitis (grade 2 according to Savary-Miller or grade C according to Los Angeles Classification), marked on the picture with arrowheads.

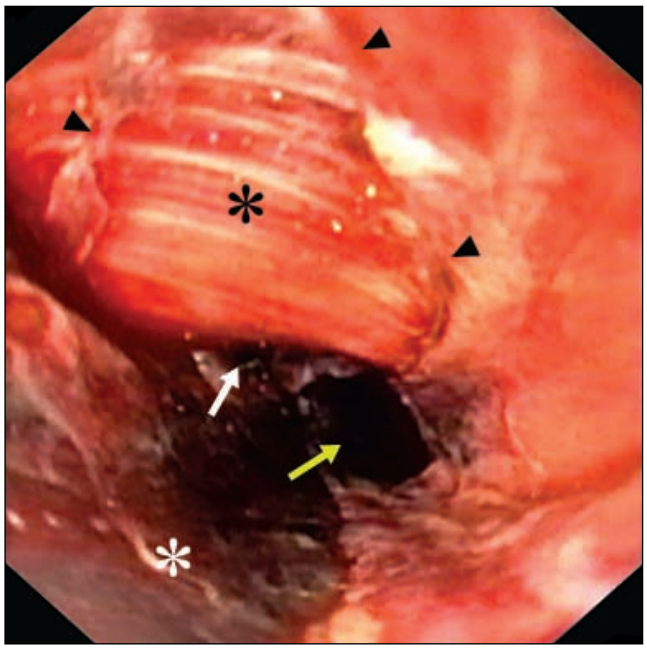

Fig. 2
After further insertion of the endoscope, an ovoid mucosal tear is seen (arrowheads) with the uncovered muscular layer of the oesophagus (black asterisk). Oesophageal rupture is marked with a white arrow, and direction to the cardia with a yellow arrow. A large clot of blood in the oesophageal lumen is clearly visible, too (white asterisk).

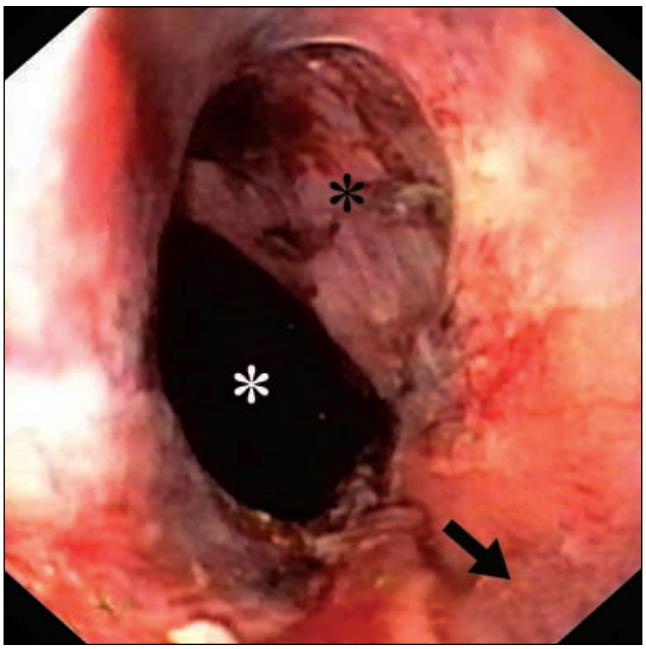

Fig. 3
Oesophageal rupture. Endoscopic view of perforation at the abdominal part of the oesophagus. A large clot of blood in the perforation cavity (white asterisk), muscular layer of the oesophagus (black asterisk), and direction to the cardia (black arrow).

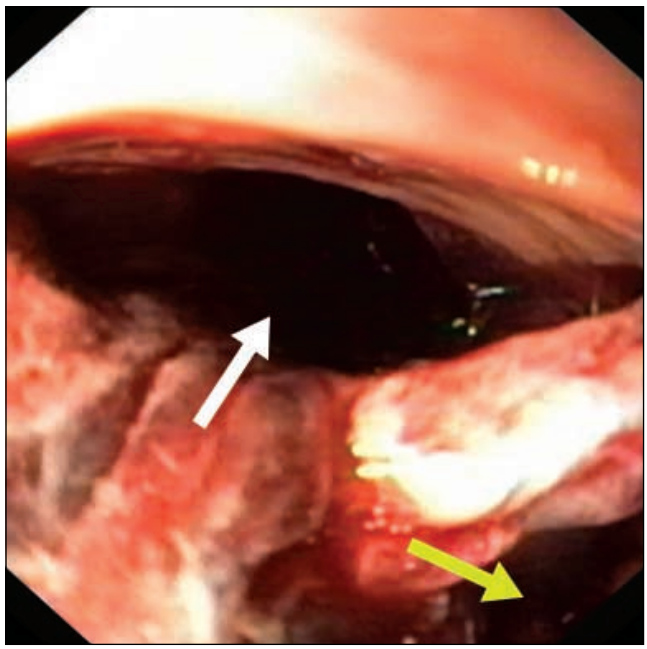

Fig. 4
Detailed endoscopic view of the oesophageal rupture. Orifice of the perforation cavity (white arrow), direction to the cardia (yellow arrow).

duodenal bulb and further duodenum were normal. The perforation cavity was localized to the left back of the abdominal oesophagus and posterior gastric wall and with regard to absence of peritoneal signs, it had to be a covered perforation without direct communication either to the lesser sac or peritoneal cavity. Surgical intervention confirmed large perforation of the abdominal part of the oesophagus 5 centimetres in length and large haematoma close to the stomach. On surgery, the rupture was closed with a primary suture without flap reinforcement and the wound was drained with two drains. During initial postoperative days, the course was without meaningful complications except for severe acute alcoholic delirium. Complex intensive care was provided, including continuous nasogastric suction, intravenous broad-spectrum antibiotics (cefoxitin) and total parenteral nutrition. On the 7th postoperative day, a water-soluble contrast swallow showed a leak from the suture area (Fig. 5) but local drainage was sufficient. That is why no further surgery was done. A CT scan showed the leak, too, found pneumomediastinum, and detected small effusions in both pleural spaces (Figs. 6 to 8).

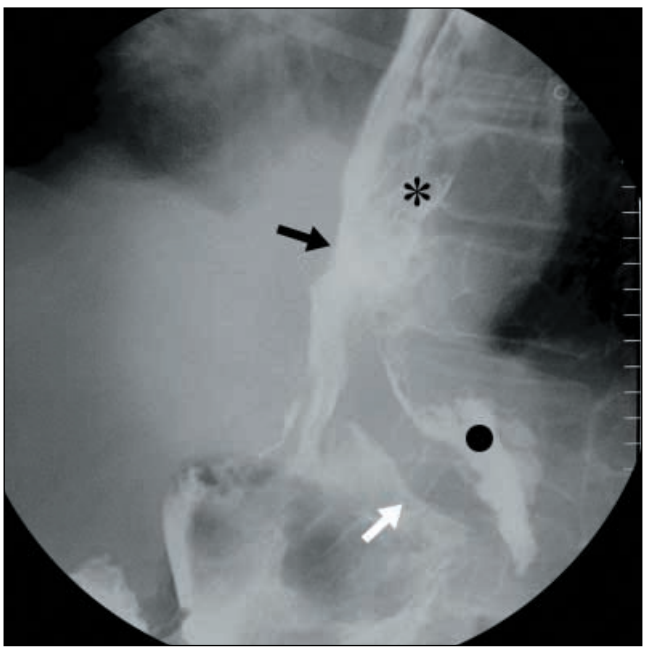

Fig. 5
Water-soluble contrast swallow shows a leak from the suture area (black arrow) on the 7th postoperative day. Contrast media flow along the oesophagus into the mediastinum (asterisk). The perforation cavity is filled with contrast media (closed circle), the cavity disfigures the fornix of the stomach (white arrow).

Intensive metabolic care was continued without any further complication. Parenteral nutrition was shifted to total enteral nutrition via a fine bore nasojejunal tube (using a polymeric formula). One month after surgery a check oesophageal contrast swallow was done with quite normal findings. After gradual realimentation, the patient was discharged six weeks after surgery in good general condition. At the present time, six months later, the patient is symptom free.

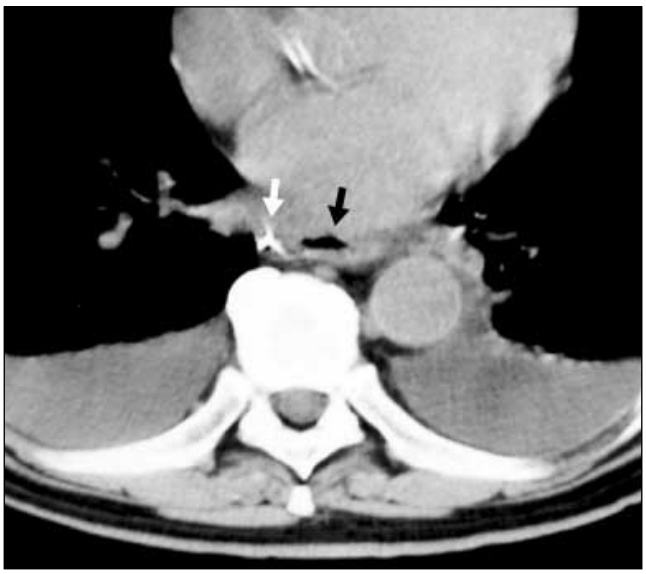

Fig. 6
Native CT scan of the mediastinum (9th postoperative day). Pneumomediastinum shown at a detailed view. A strip of gas is seen at the posterior part of mediastinum left of the oesophagus (black arrow). The nasogastric tube in the oesophagus is marked with a white arrow.

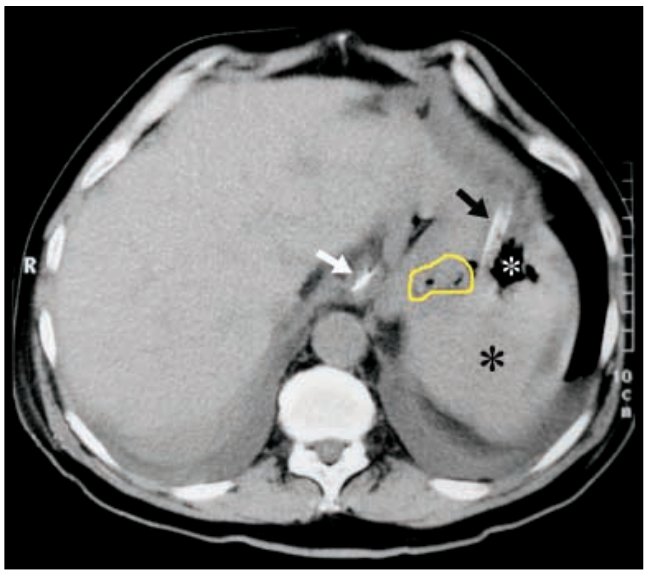

Fig. 7
Native CT scan (9th postoperative day). The perforation cavity filled with gas (white asterisk) is placed below the left diaphragm between the abdominal part of the oesophagus with the nasogastric tube (white arrow) and the upper pole of the spleen (black asterisk). The surgically introduced drain is marked with a black arrow. The gastric fornix is circled with a yellow line.

Discussion

Spontaneous rupture of the oesophagus (Boerhaave's syndrome) is a rare, life threatening condition. The classic clinical presentation is usually introduced by repeated episodes of vomiting, typically in a middle-aged man with recent excessive dietary and alcohol intake. This is followed by a sudden onset of severe chest pain. Typical clinical features consist of effort vomiting, sudden chest pain and subcutaneous emphysema (the Mackler triad) (10,23).

Because it is usually associated with a forceful emesis, Boerhaave's syndrome is not truly spontaneous. However, the term is useful for distinguishing it from iatrogenic perforation, which accounts for 85 - 90 % of cases of oesophageal rupture (3,23).

Oesophageal rupture in Boerhaave's syndrome is postulated to be the result of a sudden rise in intraluminal oesophageal pressure produced during vomiting as a result of neuromuscular incoordination causing failure of the cricopharyngeus muscle to relax. The syndrome is commonly associated with overindulgence in food and alcohol, alcoholism being a primary risk factor (23). We report a case of oesophageal rupture in a heavy drinker presented with protracted vomiting (later with repeated haematemesis) and abdominal pain lasting for 3 days. There were neither chest pain and subcutaneous emphysema nor less typical findings (cervical vein distension, hoarseness, tachypnea, breathlessness, peripheral cyanosis) or later stages of illness (sepsis, haemodynamic instability). There was no radiation of abdominal pain, without abdominal rigidity. Swallowing did not aggravate pain or precipitate coughing. Boerhaave's syndrome can be associated with pre-existing oesophageal

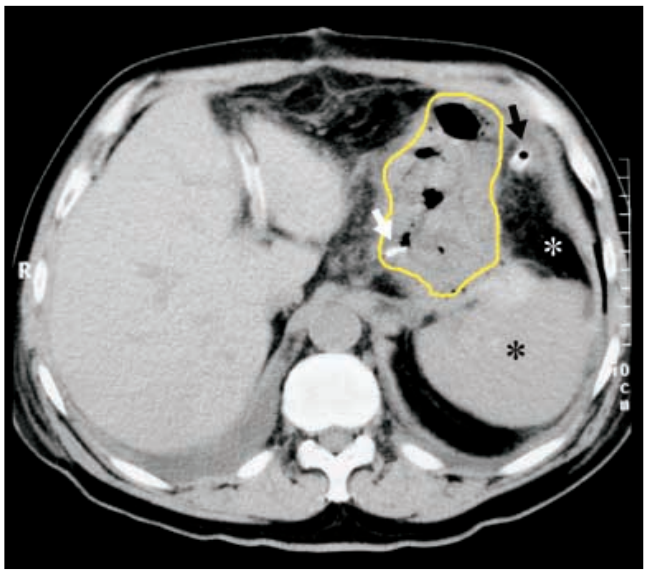

Fig. 8
Native CT scan (on the 9th postoperative day). The perforation cavity is seen, mostly filled with gas (white asterisk) close to the stomach (circled with a yellow line). The nasogastric tube (white arrow), upper pole of the spleen (black asterisk). The surgically introduced drain is marked with a black arrow.

disease, like gastro-oesophageal reflux disease, Barrett's oesophagus or oesophageal carcinoma (7,8,16). There was pre-existing reflux oesophagitis (as a manifestation of gastro-oesophageal reflux disease) in our case. Recurrent spontaneous rupture of the oesophagus has been reported only exceptionally (20,27).

Diagnosis of Boerhaave's syndrome is based on upright chest radiograph (pneumomediastinum, unilateral pleural effusion, mediastinal widening), water-soluble contrast swallow and CT scan (extra-oesophageal air, peri-oesophageal fluid, air and fluid in the pleural spaces, retroperitoneum or lesser sac) (2,14,16,23). Endoscopy is not commonly used for diagnosis. Furthermore, it may be associated with additional risks as both the endoscope and insufflation of air can extend the perforation and introduce air into the mediastinum (13). However, endoscopy may be useful when perforation is only suspected or even not expected. In our case, the patient was referred for upper GI endoscopy because of vomiting blood with suspicion of volvulus of the stomach. Oesophageal rupture was recognized only by means of endoscopy.

Boerhaave's syndrome is a transmural perforation of the oesophagus to be distinguished from the more common Mallory-Weiss syndrome, a non-transmural mucosal tear of distal oesophagus or proximal part of the gastric body, also associated with vomiting (23). The most common anatomical location for perforation in Boerhaave's syndrome is at the left posterolateral wall of the lower third of the oesophagus (at its weakest point) (23). This part of the abdominal oesophagus was affected in our case, too.

Laboratory findings are often nonspecific. Patients may be presented with leukocytosis, half of them having a higher haematocrit value due to fluid loss into mediastinum and pleural spaces (23). We found minor bilateral pleural effusion and the CT scan revealed pneumomediastinum. There were no crackling sounds upon chest auscultation (known as the Hamman crunch).

In differential diagnosis of Boerhaave's syndrome, it is necessary to distinguish local oesophageal affections, such as Mallory-Weiss mucosal tear, spontaneous intramural oesophageal perforation and intramural haematoma (oesophageal apoplexy) as well as extra-oesophageal disease (acute myocardial infarction, aortic dissection, acute pancreatitis, aspiration pneumonia and tension pneumothorax) (10,16,23).

Delay in the diagnosis of Boerhaave's syndrome is frequent because a traumatic episode may occur during alcoholic drunkenness or the presented signs and symptoms are nonspecific and atypical (one third of all cases) (3,23). Delay in rupture recognition results in a significant increase in complications (25). Overall lethality of spontaneous oesophageal rupture is about 35 %. The best outcomes are associated with early recognition and appropriate surgical treatment within 12 hours of rupture. After 24 hours the lethality rate rises up to 50 % and to nearly 90 % after 48 hours (10,23,25). Nowadays, modern surgical techniques and intensive care management have significantly reduced lethality and improved prognosis in this syndrome (19,26,34,35).

The majority of oesophageal perforations require urgent surgical treatment (17,21,30,31,33,35). As food, microorganisms, gastric acid and digestive enzyme contaminate the mediastinum in spontaneous perforation of the oesophagus, surgery should be performed within 24 hours if possible (19). Within 12 hours of perforation, a polymicrobial infection is common, Staphylococcus, Pseudomonas, Streptococcus and Bacteroides organisms are common pathogens (18). Surgery consists of primary repair of the ruptured oesophagus, mediastinal debridement and pleural drainage (15,19,24). Thoracoscopic repair of the oesophageal rupture as a mini-invasive approach was also published (29). Oesophageal resection and delayed reconstruction may be an option where primary repair is impossible or as a salvage procedure where other methods have failed. The poor quality of life after oesophagectomy is improved by subsequent reconstruction (32). Complications of surgery in general include persistent oesophageal leak, mediastinitis and sepsis. Reinforcement of the suture line with a fundic wrap, diaphragmatic, pleural or omental flap appears to reduce the incidence of postoperative leak (15,28,34). Late complications of surgical intervention may include empyema and oesophago-tracheal fistulas (23).

Non-surgical treatment includes intravenous volume resuscitation, continuous nasogastric suction, intravenous broad-spectrum antibiotics and total parenteral or later enteral nutrition by nasojejunal tube (3,23,30). Endoscopic therapy for Boerhaave's syndrome has been utilizing new, covered, self-expanding metallic stents (4,6,9,12,22) but their precise role remains to be determined in patients with

delayed diagnosis, who are at high surgical risk and exhibit persistent oesophageal leak despite conservative treatment (11).

Boerhaave's syndrome is a surgical emergency, its prognosis depends on prompt diagnosis and adequate treatment. Ideal management involves a combination of both surgical intervention and intensive medical care.

REFERENCES

1. Agarwal PK, Miller SE: Spontaneous rupture of the oesophagus: case report of a delayed diagnosis and subsequent management. J Roy Soc Med 1995; 88: 149 - 150.
2. Backer CL, Lo C, Hartz RS: Computed tomography in patient with oesophageal perforation. Chest 1990; 98: 1078 - 1080.
3. Baehr PH, McDonald GB: Oesophageal disorders caused by infection, systemic illness, medications, radiation, and trauma (p. 519 - 539). In: M Feldman, BF Scharschmidt, MH Sleisenger, eds: Sleisenger & Fordtran's Gastrointestinal and Liver diseases. 6th ed. Philadelphia: WB Saunders, 1999.
4. Bak YT, Kwon OS, Yeon JE, Kim JS, Byun KS, Kim JH, Kim JG, Lee CH, Choi YH, Kang DH: Endoscopic treatment in a case with extensive spontaneous intramural dissection of the oesophagus. Eur J Gastroenterol Hepatol 1998; 10: 969 - 972.
5. Bjerke HS: Boerhaave's syndrome and barogenic injuries of the oesophagus. Chest Surg Clin N Am 1994; 4: 819 - 825.
6. Chung MG, Kang DH, Park DK, Park JJ, Park HC, Kim JH: Successful treatment of Boerhaave's syndrome with endoscopic insertion of a self-expandable metallic stent: report of three cases and a review of the literature. Endoscopy 2001; 33: 894 - 897.
7. Craven GG, Whittaker MG: Boerhaave's syndrome as a complication of pre-existent gastrointestinal disease. Ir J Med Sci 1992; 161: 670 - 674.
8. Davis R: The Boerhaave syndrom associated with carcinoma of the oesophagus. South Afr Med J 1973; 47: 2269 - 2272.
9. Davies AP, Vaughan R: Expanding mesh stent in the emergency treatment of Boerhaave's syndrome. Ann Thorac Surg 1999; 67: 1482 - 1483.
10. Duehring GL: Boerhaave syndrome. Radiol Technol 2000; 72: 51 - 55.
11. Dumonceau JM, Deviere J, Cappello M, van Gossum A, Cremer M: Endoscopic treatment of Boerhaave's syndrome. Gastrointet Endosc 1996; 44: 477 - 479.
12. Eubanks PJ, Hu E, Nguyen D, Procaccino F, Eysselein VE, Klein SR: Case of Boerhaave's syndrome successfully treated with a self expandible metallic stent. Gastrointest Endosc 1999; 49: 780 - 783.
13. Gubbins GP, Nensey YM, Schubert TT, Batra SK: Barogenic perforation of the oesophagus distal to a stricture after endoscopy. J Clin Gastroenterol 1990; 12: 310 - 312.
14. Han SY, McElvein RB, Aldrete JS, Tishler JM: Perforation of the oesophagus: Correlation of site and cause with plain film findings. Am J Roentgenol 1985; 145: 537 - 540.
15. Henderson JA, Péloquin AJ: Boerhaave revisited: Spontanoeous oesophageal perforation as a diagnostic masquerader. Am J Med 1989; 86: 559 - 567.
16. Jagminas L, Silverman RA: Boerhaave's syndrome presenting with abdominal pain and right hydropneumothorax. Am J Emerg Med 1996; 14: 53 - 56.
17. Kallis P, Belsham PA, Pepper JR: Spontaneous of the oesophagus (Boerhaave's syndrome): conservative versus surgical management. J Roy Soc Med 1991; 84: 690 - 691.
18. Katsetos MC, Tagbo AC, Linberg MP, Rosson RS: Oesophageal perforation and mediastinitis from fish bone ingestion. South Med J 2003; 96: 516 - 520.
19. Lawrence DR, Ohri SK, Moxon RE, Townsend ER, Fountain SW: Primary oesophageal repair for Boerhaave's syndrome. Ann Thorac Surg 1999; 67: 818 - 820.
20. Lujan HJ, Lin PH, Boghossian SP, Yario RF, Tatooles CJ: Recurrent spontaneous rupture of the oesophagus: an unusual late complication of Boerhaave's syndrome. Surgery 1997; 122: 634 - 636.
21. Marshall WB: Boerhaave syndrome: a case report. Am Assoc Nurs Anesth J 2002; 70: 289 - 292.
22. Mason GR: Oesophageal perforations, anastomotic leaks, and strictures: the role of prostheses. Am J Surg 2001; 181: 195 - 197.
23. Murphy M, Kalapatapu V, Ramanathan P: Boerhaave syndrome. http:// www.emedicine.com/med/topic233.htm.
24. Ojima H, Kuwano H, Sasaki S, Fujisawa T, Ishibashi Y: Successful late management of spontaneous oesophageal rupture using T-tube mediastinoabdominal drainage. Am J Surg 2001; 182: 192 - 196.
25. Pate JW, Walker WA, Cole FH, Owen EW, Johnson WH: Spontaneous rupture of the oesophagus: A 30-year experience. Ann Thorac Surg 1989; 47: 689 - 692.
26. Reeder LB, De Filippi VJ, Ferguson MK: Current results of therapy for oesophageal perforation. Am J Surg 1995; 169: 615 - 617.
27. Reeder LB, Warren SE, Ferguson MK: Recurrent spontaneous perforation of the oesophagus. Ann Thorac Surg 1995; 59: 221 - 222.
28. Safavi A, Wang N, Razzouk A, Gan K, Sciolaro C, Wood M, Vyhmeister EE, Miranda R, Ahn C, Gundry SR: One-stage primary repair of distal oesophageal perforation using fundic wrap. Am Surg 1995; 61: 919 - 924.
29. Scott HJ, Rosin RD: Thoracoscopic repair of a transmural rupture of the oesophagus (Boerhaave's syndrome). J Roy Soc Med 1995; 88: 414 - 415.
30. Sleeboom C, Slors JF: Spontaneous rupture of the oesophagus. Neth J Surg 1990; 42: 151 - 154.
31. Váňa J, Celec J, Kačo Ľ, Bízik I: Primárne nepoznaný Boerhaave syndrom. Rozhl Chir 2002; 81: 51 - 53.
32. Varghese D, Patel H, Waters R, Dickson GH: Quality-of-life study on four patients who underwent oesophageal resection and delayed reconstruction for Boerhaave's syndrome. Dis Oesophagus 2000; 13: 314 - 316.
33. Vysloužil K, Král V, Neoral Č, Klementa I, Blahut L, Konečný M: Chirurgická léčba spontánní perforace jícnu - Boerhaave syndrom. Rozhl Chir 2000; 79: 521 - 523.
34. Wright CD, Mathisen DJ, Wain JC et al: Reinforced primary repair of thoracic oesophageal perforation. Ann Thorac Surg 1995; 60: 245 - 248.
35. Záhořák K, Šimkovič D, Mašurka V: Naše zkušenosti s diagnostikou a terapií Boerhaaveho syndromu. Rozhl Chir 1989; 68: 457 - 463.

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