BOERHAAVE SYNDROME: CASE REPORT

MARCUS ALMEIDA; LEANDRO MOURA; TIMÓTEO VERÍSSIMO; RODRIGO CARVALHO; GUILHERME SOUZA; RENATA GUERRA; GUILHERME ARANTES; LEONARDO SANTOS; BRUNO SILVA; HENRIQUE LIMA

ABSTRACT

INTRODUCTION: Boerhaave Syndrome (BS) is defined as a spontaneous rupture of the esophagus, a rare but potentially fatal condition with a high morbidity and mortality rate, constituting the most lethal perforation of the gastrointestinal tract.

CASE REPORT: Male patient, 49 years old, alcoholic, presents with chest pain that has lasted for five days, triggered by emesis due to alcoholic beverages. He is normotensive, normocardic, afebrile and eupneic. Preserved chest expansion, left thoracic and cervical subcutaneous emphysema, and reduction in ipsilateral breath sounds. Computed tomography of the chest was requested and evidenced soft tissue emphysema in the cervical spaces, pleural effusion on the left and pneumomediastinum. Patient underwent double-mouth cervical esophagostomy, conventional gastrostomy, closed pleural orientation on the left and complementary therapy with endoscopic vacuum. There was good postoperative evolution with esophageal transit remaining during the same hospitalization.

DISCUSSION: The diagnosis of BS represents a real challenge, and its treatment, whether surgical or not, must be instituted immediately given the high morbidity and mortality. Although Non-Operative Treatment is reserved for selected patients, there is a tendency to adopt aggressive Operative Treatment for patients with late diagnosis, local or systemic complications. Complementary therapies such as esophageal stent or endoscopic aspirator present promising results with less invasiveness and reduced mortality rate.

CONCLUSION: SB should be remembered as a differential diagnosis of chest pain in the emergency department. Despite the scarce case series, early diagnosis and treatment constitute the most relevant prognostic factor.

KEYWORDS: BOERHAAVE SYNDROME; ESOPHAGEAL PERFORATION; MEDIASTINITIS.

INTRODUCTION

Boerhaave syndrome is defined as the spontaneous rupture of the esophagus, a rare but potentially fatal condition. Although it occurs at a low incidence rate of 3.1 per 1,000,000, its mortality rate is high, ranging from 35% to 40%, making it the most lethal perforation of the gastrointestinal tract.[1]

It was first described in 1724 by the Dutch surgeon Hermann Boerhaave, who reported in his monograph the symptoms experienced by Admiral Von Wassenauer: sudden and excruciating chest pain after vomiting following a copious meal, which led to his death. The pathology was confirmed by autopsy, which revealed an esophageal perforation.[2]

The pathophysiological mechanism involved in this condition is due to a sudden increase in intraluminal esophageal pressure with the absence of relaxation of the upper esophageal sphincter, leading to a complete rupture of all layers of the esophagus (Haba et al, 2020). The main reported symptoms are chest pain, vomiting, fever, subcutaneous emphysema, and dyspnea. They are classically described by the Mackler triad composed of vomiting, chest pain, and subcutaneous emphysema.[1]

The diagnostic workup requires a clinical history that includes risk factors for BS with suggestive manifestations. Among the complementary tests, one can use chest X-ray (CXR), which is a method with high sensitivity, up to chest and abdominal CT with oral contrast, which is the most specific method, capable of identifying the site of esophageal perforation, as well as its extension and associated complications. Other tests such as contrast esophagogram and upper gastrointestinal endoscopy (UGIE) may be used for selected patients.[3]

The management can be instituted by ST or NST, although endoscopic therapies have stood out more recently, especially those using vacuum therapy or stent placement, corresponding to the main or adjuvant therapy.1

Early diagnosis is a real challenge and is one of the main prognostic factors, especially those diagnosed within the first 24 hours, as the mortality rate increases considerably in those with late evolution, reaching up to 65% mortality due to mediastinitis and rapid progression to septic conditions.[4]

CASE REPORT

Male patient, 49 years old, asthmatic and alcoholic, was referred for evaluation by the general surgery team due to ventilator-dependent chest pain and thoracic subcutaneous emphysema within five days of evolution. Symptoms

1. Hospital Estadual de Aparecida de Goiânia.

ADDRESS
MARCUS ALVES CAETANO DE ALMEIDA
Endereço: Av. Diamante, s/n - St. Conde dos Arcos, Aparecida de Goiânia - GO, 74969-210.
Email: marcuscaetanomed@gmail.com

were triggered after vomiting due to alcohol intake. Absence of dyspnea, nausea, vomiting, or fever on admission. Patient undergoing antimicrobial therapy in the originating unit.

The physical examination revealed a patient in fair overall condition, with good skin color, well-hydrated, normotensive, normocardic, afebrile, and eupneic in ambient air. On examination of the respiratory system, preserved chest expansibility was noted, along with subcutaneous emphysema on palpation, predominantly in the thoracic and left cervical regions, and reduced vesicular murmur on auscultation ipsilaterally. Neurological, cardiovascular, and abdominal systems showed no alterations.

In the face of the diagnostic hypothesis of BS, a CT scan of the neck, chest, and abdomen with intravenous contrast was requested, which revealed marked soft tissue emphysema in the anterior and paravertebral cervical spaces (Figure 01), as well as left pleural effusion associated with pneumomediastinum (Figure 02). The abdominal CT scan was normal. Laboratory tests showed leukocytosis (15,370/mm³), with normal band neutrophil count, and elevated C-reactive protein (65.1 mg/L).

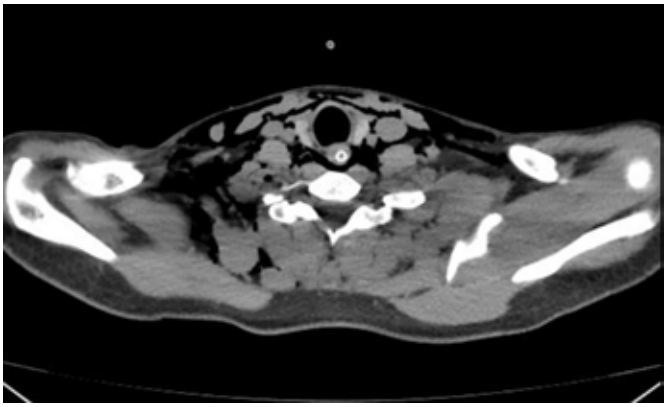

Figure 01 - Extensive soft tissue emphysema in cervical and paravertebral compartments.
Source: Personal archive.

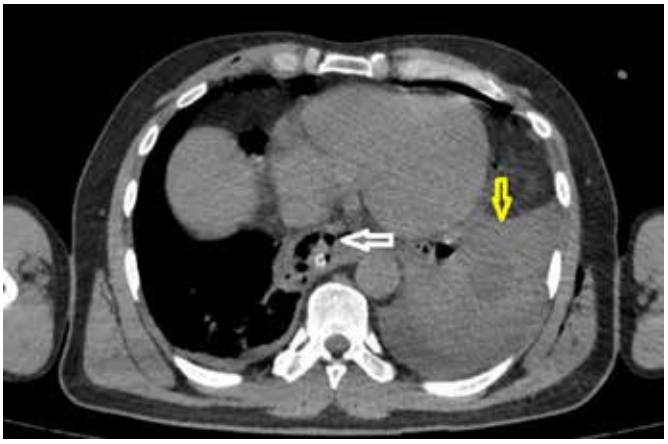

Figure 02 - Tomographic signs of pneumomediastinum, white arrow. Large left pleural effusion, yellow arrow. Source: Personal archive.

After clinical reassessment, closed water seal pleural drainage was indicated on the left, followed by urgent upper gastrointestinal endoscopy, which showed a 2 cm diameter esophageal perforation in the left lateral wall, located 40 cm from the upper dental arch and 1 cm from the esophagogastric junction. It communicated with the left pleural space, which showed extensive contamination (Figure 03).

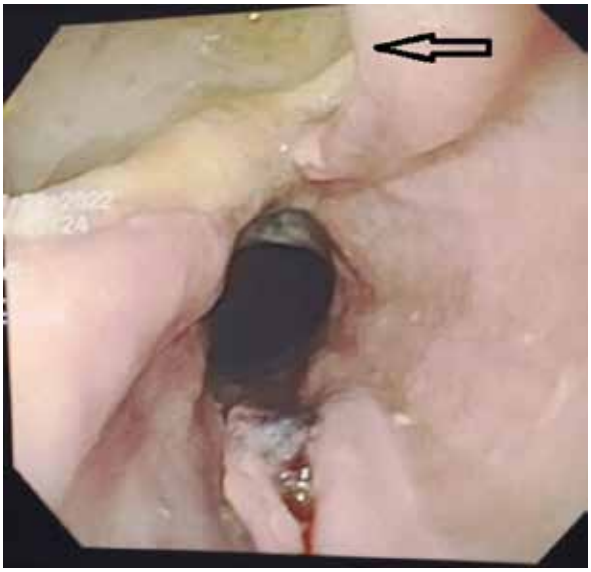

Figure 03 - Endoscopic evidence of transmural esophageal injury on the left lateral wall, indicated by the black arrow. Source: Personal archive.

It was indicated, in the same surgical procedure, the performance of cervical esophagostomy with double stoma for transit diversion associated with conventional gastrostomy tube for enteral nutrition (figure 04).

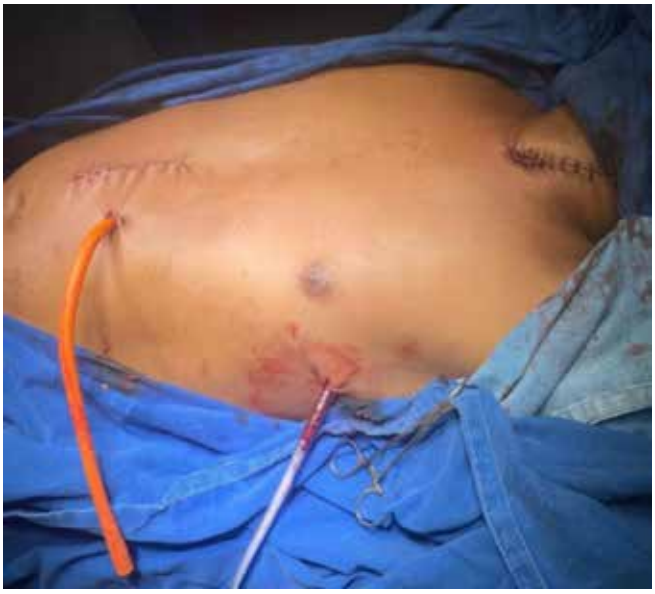

Figure 04 - Patient in immediate postoperative period. Source: Personal archive.

In the postoperative course, the thoracic drain showed enteral-like output, indicating the presence of a left esophago-pleural fistula. Due to its persistence, on the 12th postoperative day (POD), endoscopic vacuum therapy was indicated along with Total Parenteral Nutrition (TPN) and decompressive GT. After four days of this therapeutic approach, there was a significant reduction in thoracic drain output, followed by resolution of the fistula, leading to the early removal of the thoracic drain and endoscopic vacuum therapy.

Underwent a follow-up EGD on the 31st POD, which showed a healed esophageal lesion in good condition. Consequently, within 48 hours, esophageal transit reconstruction was performed with primary end-to-end anastomosis in a single plane associated with cervical laminar drainage. The patient was discharged 72 hours after the procedure following a negative methylene blue test.

DISCUSSION

BS is a rare cause of esophageal perforation in the general population but is associated with known risk factors, which involve increased esophageal intraluminal pressure. These include uncontrollable vomiting, often resulting from alcohol consumption, asthma attacks, straining during bowel movements, and heavy lifting.[5] In this case study, at least two of these factors are identifiable: episodes of vomiting and asthma, in addition to the significant association with alcohol consumption.

Despite the high mortality rate associated with delayed diagnosis of BS, this case demonstrates a prolonged clinical course, contrasting with the natural history of the disease. The patient presented to the emergency department with a stable clinical condition and no signs of sepsis based on the quick Sequential Organ Failure Assessment (qSOFA) score, despite at least 96 hours of esophageal perforation evolution. This outcome may be attributed to the intravenous antimicrobial therapy initiated at the referring facility, as well as clinical support and likely delayed but progressive contamination of the mediastinal and pleural compartments. In a classic progression, rapid onset of sepsis due to mediastinitis is expected within approximately 12 to 24 hours, with progressive organ dysfunction beyond 48 hours of evolution. However, the patient's presentation with thoracic pain, dyspnea, and subcutaneous emphysema made spontaneous esophageal rupture a feasible diagnostic hypothesis.

From an anatomical standpoint, it is noteworthy that during the endoscopic examination, the esophageal perforation occurred in the left lateral wall, located one centimeter from the esophagogastric junction, with a length of two centimeters. These data reinforce and corroborate with the literature, except for the significant proximity to the esophagogastric junction.[2]

Given the presented case, it is evident that the adoption of NST is not feasible due to late clinical presentation, with at least five days of evolution, extensive contamination of the mediastinal and pleural compartments, as well as signs of ventilatory discomfort and chest pain. Based on objective criteria, as proposed by the Pittsburgh score, a sum of six points is calculated, including leukocytosis, pleural effusion, uncontained fistula, and delayed diagnosis, which also supports the need for aggressive surgical treatment.[3,6]

Therefore, a GT feeding tube was constructed as a method of long-term enteral nutrition, along with double-lumen cervical esophagostomy as a principle of contamination control, in addition to closed left pleural drainage. Although not performed, Nissen fundoplication with perforation blocking was also a feasible option due to its proximity to the esophagogastric junction.[4]

During the postoperative management, despite the surgical therapy instituted, persistence of the left esophagopleural fistula was evidenced, leading to the possibility of using endoscopic vacuum therapy, which represents a minimally invasive option with results comparable to primary closure, despite the low number of cases. In current literature, success rates range from 70 to 100%, with mortality rates of 7 to 18% and complication rates of 10 to 14%.[7] After 96 hours of application, aiming at reducing contamination and conditioning the lesion, there was closure of the fistula and healing of the perforation, objectively assessed by follow-up endoscopy.[8]

The reconstruction of the esophageal tract occurred during the same hospitalization and represented an important milestone for the patient's quality of life in the postoperative period, as oral diet became feasible.

CONCLUSION

SB, despite its rarity, should be considered as a differential diagnosis in cases of chest pain in the emergency department, especially in patients with known risk factors.

There is much debate about the ideal therapeutic modality, and among its possibilities, it is convenient to aim for risk stratification, with recommended scores such as the Pittsburgh score to define surgical therapy or not. Regardless of the therapeutic approach, it is necessary to follow principles for the treatment of this serious condition, which include treatment and control of local contamination, enteral nutrition, and wide drainage if necessary.

The gold standard surgical treatment is primary repair of the lesion for eligible patients. Other alternative modalities, such as exclusion and diversion of transit, are options in unfavorable clinical scenarios. Endoscopic treatment, either vacuum therapy or self-expanding stent placement, has gained prominence, with some studies showing outcomes similar to the gold standard treatment. Finally, despite the limited literature and case studies on BS, it is demonstrated that the most relevant prognostic factor is early diagnosis and treatment.

REFERENCES

1. Allaway MGR, Morris PD, B. Sinclair J, Richardson AJ, Johnston ES, Hollands MJ. Management of Boerhaave syndrome in Australasia: a retrospective case series and systematic review of the Australasian literature. ANZ Journal of Surgery. 2020 Dec 14;91(7-8):1376–84.
2. Salvador-Ibarra IJ, Pizaña-Davila A. Síndrome de Boerhaave: reporte de un caso y revisión de la literatura. Cirugía y Cirujanos. 2021 Dec 21;89(92).
3. Chirica M, Kelly MD, Siboni S, Aiolfi A, Riva CG, Asti E, et al. Esophageal emergencies: WSES guidelines. World Journal of Emergency Surgery. 2019 May 31;14(1).
4. Dias P, Martins De Freitas V, Medina Da Fonseca C. REVISÃO DE LITERATURA: MANEJO NA PERFURAÇÃO DE ESÔFAGO. REVISTA EMESCAM DE ESTUDOS EM SAÚDE. 2019 Jan;1(1).
5. Haba Y, Yano S, Akizuki H, Hashimoto T, Naito T, Hashiguchi N. Boerhaave syndrome due to excessive alcohol consumption: two case reports. International Journal of Emergency Medicine. 2020 Nov 30;13(1).
6. Schweigert M, Santos Sousa H, Solymosi N, Yankulov A, Fernández MJ, Beattie R, et al. Spotlight on esophageal perforation: A multinational study using the Pittsburgh esophageal perforation severity scoring system. The Journal of Thoracic and Cardiovascular Surgery. 2016 Apr;151(4):1002–11.
7. Pattynama LMD, Eshuis WJ, van Berge Henegouwen MI, Bergman JJGHM, Pouw RE. Vacuum-stent: A combination of endoscopic vacuum therapy and an intraluminal stent for treatment of esophageal transmural defects. Frontiers in Surgery. 2023 Feb 22;10.
8. Loske G, Schorsch T. Endoskopische Vakuumtherapie beim Boerhaave-Syndrom. Der Chirurg. 2016 Jun 3;87(8):676–82.