Diagnosis and treatment of spontaneous esophageal rupture (Boerhaave syndrome): A case report and literature review

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Abstract

We present a case report of spontaneous esophageal rupture (Boerhaave syndrome) has been successfully treated at Viet Duc University Hospital. Through this case report, we reviewed the literatures on this rare disease's pathogenesis, diagnosis, and treatment methods.

Keywords: Boerhaave syndrome; Endoscopic Vacuum-Assisted Closure Therapy; Endoscopic Vacuum Therapy.

Introduction

Spontaneous esophageal rupture or Boerhaave syndrome is a rupture of the esophagus caused by a sudden increase in intraluminal pressure. It was first described in 1724 by Hermann Boerhaave, Professor of Medicine at Leiden University, in a publication entitled “History of a Grievous Disease Not Previously Described [1]. This rare pathology can be easily overlooked, leading to severe clinical manifestations such as mediastinitis, sepsis, and mortality up to 20-75% if its not treated promptly [2]. The diagnosis is challenging because of unspecified symptoms, so diagnostic imaging studies are necessary, and its management requires a multidisciplinary team. [3 - 5]

Case report

A 42-year-old male patient with a history of alcoholism has manifested with fever and left chest pain for a week. At the time of admission, the patient was alert, with a blood pressure of 100/60mmHg, heart rate of 93 bpm, respiratory rate of 25/min with oxygen, and oxygen saturation of 97% with diminished left lung sounds. Chest radiography showed left pleural effusion. Thoracic CT scan showed sizeable pleural effusion and pneumothorax in the left pleural cavity, with thickening of the lower third of the esophagus. A chest tube was placed in the left pleural cavity, draining 200ml of brown fluid. The patient underwent an upper gastrointestinal endoscopy and found a 20mm esophageal perforation at the lower third of the esophagus, above the esophagogastric junction. The diagnosis of Boerhaave syndrome was suggested, and surgery was indicated. During the surgery, we identified a 3 cm longitudinal perforation lesion in the lower third of the esophagus on the left border side. The edges of the esophageal perforation were debrided, and primary repair was performed with interrupted 3–0 Vicryl stitches.

A gastrostomy was performed to reduce pressure along with an jejunostomy for feeding. Extensive abdominal drainage and pleural cavity drainage were done. During postoperative follow-up, the patient developed pneumonia so he was referred to the intensive care unit for special care. Pleural drainage discharged 100-200ml/day of brown fluid with gastric juice. After seven days, we performed again an endoscopy, the perforation was still found but decreased the size to about 1.5 cm. We decided to perform E-VAC (Endoluminal vacuum-assisted closure) therapy.Repeated gastrointestinal endoscopy was indicated every seven days to evaluate the healing progress of the esophageal lesion. The patients condition gradually stabilized, finally he was discharged after five weeks of hospitalization, the esophageal rupture was completely healed.

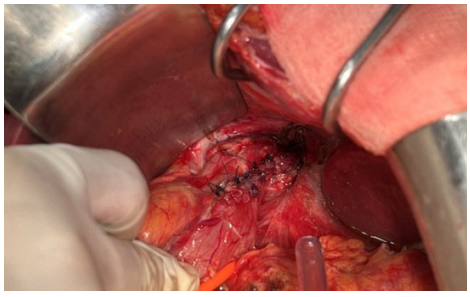

Figure 1. Primary repair of esophageal perforation

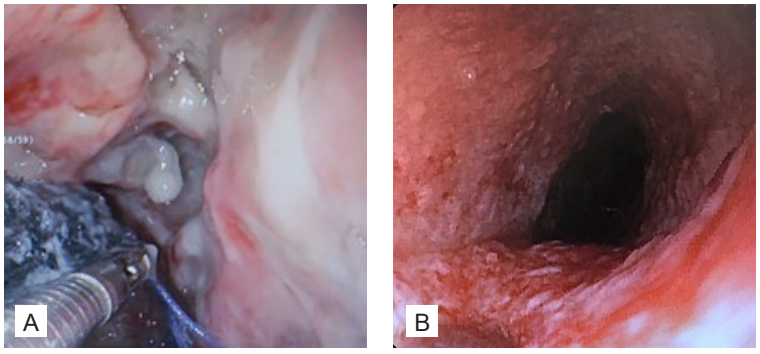

Figure 2. A: 2 weeks, B: 5 weeks (Esophageal rupture completely healed). Post EVAC-therapy endoscopy

Discussion

Through the case was successfully treated, we would like to discuss about the pathology, diagnosis and management.

Pathology

Boerhaave syndrome accounts for about 15% of all cases of esophageal rupture. This is about 3.1/1,000,000 population per [3]. The mechanism of Boerhaave syndrome is due to a sudden increase in intra-esophageal pressure combined with negative intrathoracic pressure causing complete tearing of the layers of the esophagus. Common risk factors are alcoholism, hyperemesis, prolonged cough, childbirth, and abdominal trauma. It is found that most patients are middle-aged men, possibly related to alcohol abuse. Our patient is a middle-aged male with a history of alcohol use.

Esophageal rupture in Boerhaave syndrome can occur at any site. However, the most common is the left posterior-lateral wall of the lower third of the esophagus because this is the weak part of the esophagus [6]. Diagnosis of Boerhaave syndrome is difficult due to its non-specific manifestations. The clinical symptoms of Boerhaave's syndrome depend

on the location of the esophagus rupture, degree of rupture, and time of disease detection. Cervical esophageal rupture is more often localized because of the limited ability to spread to the mediastinum through the retroesophageal space of digestive fluid, and symptoms may include neck pain, dysphagia, or dysphonia. Intrathoracic esophageal rupture can easily lead to mediastinal abscess or pneumomediastinum, with common symptoms such as vomiting, dyspnea, chest pain, and subcutaneous emphysema. Lower esophageal rupture can cause peritonitis. However, one-third of the patients dont have these symptoms3. The patient in our clinical report was hospitalized with fever and left chest pain with dyspnea for about one week, which was atypical for Boerhaave syndrome and can be confused with many other medical conditions.

Diagnosis

Diagnostic tests include contrast-enhanced esophageal radiography, which helps detect contrast leaks out of the esophagus. Water-soluble contrast media is preferred over barium because leakage of barium is more likely to cause mediastinitis and subsequent fibrosis. Chest CT scan is a useful, more sensitive tool than contrast-enhanced esophageal radiograph, allowing the assessment of abscesses and adjacent organs and for differential diagnosis with other esophagus lesions such as esophageal cancer. Gastrointestinal endoscopy can be used with caution because it can cause more severe damage to the esophagus, increase the volume of fluid air into the mediastinum or pleural cavity, and affect the patient's condition. In some patients with pleural drainage, a methylene blue test may be indicated [6]. Our patient's chest CT scan only detected indirect signs such as pneumothorax in the left pleural cavity, air in the mediastinum, and abnormal thickening of the lower third of the esophagus, but no sign accurately showed esophageal perforation. The patient underwent a gastrointestinal endoscopy and found a perforated lower third of the esophagus, helping to make a definitive diagnosis.

Treatment

Esophageal perforations and postoperative esophageal anastomotic leaks are still a life-threatening condition; the reported mortality ranges from 10 to 25%, when therapy is started within 24 h, and from 40 to 60%, when the treatment is delayed. Regarding treatment principles, patients fast and compensate with adequate fluid, the abscess is extensively drained, broad-spectrum antibiotics are used, and the esophageal wall should be repaired if possible. The choice of treatment depends on many factors such as the patient's general condition, the location and extent of the esophageal rupture, the time of diagnosis, and the integrity of the esophageal wall. For cases diagnosed early (before 24 hours), primary esophagus repair is considered the gold standard for treatment [4], [5], [10]. The result is usually good. There are controversies among authors about late cases (diagnosed after 24 hours) because the esophageal walls are often edematous or fibrotic; it is difficult to suture the esophagus, with a high failure rate. The choice of open or laparoscopic surgery depends on the patients condition and the surgeons experience. There have been reports of performing esophageal suture laparoscopically [8]. However, in our opinion, open surgery should be performed to facilitate the esophagus suture and extensive drainage of the cavity. It does not prolong operation time in an emergency setting. Some authors may choose esophagectomy or cervical esophagostomy, but esophagectomy at this time may aggravate the patient's condition; cervical esophagostomy requires re-operation for esophageal reconstruction. In addition to surgery, endoscopic interventions also help support the treatment of this disease. Esophageal stenting is indicated for treatment in some patients with late arrival or severe systemic condition that does not allow surgery [7 - 10]. However, esophageal stent placement does not resolve mediastinal infection.

EVAC (Endoscopic vacuum-assisted closure) therapy or EVT (Endoscopic Vacuum Therapy) firstly reported in the treatment of an anastomotic leak following a rectal surgery in 2003 [11]. Since then, EVT has been used in the adult population

for closure of esophageal, gastric (most commonly after bariatric surgery), small bowel, pancreatic, and colorectal defects, with success rates above 70%. Recently it has been utilized to treat cases of upper gastrointestinal fistula, with good results [12]. In 2017, Kuehn et al [13] published a MEDLINE analysis of 11 case series with over 210 patients with upper GI tract defects treated with E-Vac. In this review, success rate was 90 and 96% for anastomotic leakages and esophageal perforations, respectively. Principle of the EVAC method: A suction tube fitted with a sponge is inserted into the abscess (intracavitary) or into the gastrointestinal tract (intraluminal) under endoscopic guidance. Then, it is continuously suctioned with a pressure of 100-125mmHg. Necrotic tissue, pus and bacteria are sucked out to help clean the abscess through this negative pressure suction, and it also stimulates the development of granulation tissue to facilitate wound healing. The procedure is easy to perform, does not require complicated equipment, and the procedure time is short. The authors suggest that the endoscopy should be repeated after 3-5 days to evaluate the lesion, and shouldnt be left more than 7 days because it will be difficult to remove the suction tube and easily cause bleeding. One advantage of the EVAC is that it can help control infection in critically ill patients; or in patients with gastrointestinal lesions in locations where surgical intervention is difficult (eg, esophageal anastomotic fistula located in the mediastinum), however if the patient's symptoms dont improve, surgical intervention may be required [12]. Disadvantage of the EVAC method is that the treatment stay usually lasts 4-5 weeks, increasing the patient's hospital costs. Currently, there are still no standardized indications for EVAC treatment. Most authors believe that lesion such as: anastomotic leakage, perforation (acute or chronic) of the upper or lower gastrointestinal are candidates for EVAC method. Common EVAC application sites are the esophagus (esophageal perforation due to trauma or pathology; esophageal anastomotic fistula), stomach (gastric anastomotic fistula), rectum (anastomotic leak and fistula). Our personal opinion, in cases of large perforations > 5cm, surgery and the EVAC method should be combined to increase the rate of success and shorten the patient's hospital stay.

Our patient was actively resuscitated; we actively debrided the edges of the esophageal perforation, and we decided to suture the esophagus primarily. After surgery, the patient still had an esophageal fistula. To resolve this situation, we combined with a digestive endoscopy doctor to set up EVAC at the ICU. To reduce treatment costs, every 7 days we performed the endoscopy again to evaluate the healing progress of the esophageal lesion and replace the sponge. After 5 weeks, the perforation was completely healed, the patient gradually stabilized and could be discharged. EVAC therapy helped the patient avoid re-operation.

Conclusion

Boerhaaves syndrome still represents a challenge in diagnostic and therapeutic with high mortality. Early diagnosis and management, including thoracic drainage and surgery, are essential to improve prognosis and reduce mortality. Management requires a multidisciplinary team for better results. We suggest EVAC as an essential component in the treatment algorithm for Boerhaave syndrome.

References

1. C. Michael Gibson, M.S., M.D.Boerhaave syndrome overview.https://www.wikidoc.org/index.php/Boerhaave_ syndrome_overview.
2. Turut H, Gulhan E, Adams P Y, Cetin G. Successful conservative management of Boerhaaves syndrome with late presentation. *J Natl Med Assoc*. 2006; 98(11): 1857–1859
3. Turner AR, Turner SD. Boerhaave Syndrome. [Updated 2022 Dec 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-.Available from: https://www.ncbi.nlm.nih.gov/books/NBK430808/
4. Kollmar O, Lindemann W, Richter S, Steffen I, Pistorius G, Schilling MK. Boerhaave's syndrome: primary repair vs. esophageal resection--case reports and meta-analysis of the literature. *J Gastrointest Surg*. 2003; 7(6):726-34

5. Han D, et al. The role of operation in the treatment of Boerhaave's syndrome. *BioMed Research International, Volume 2018*, Article ID 8483401, 5 pages.https://doi.org/10.1155/2018/8483401
6. Chew FY, Yang ST. Boerhaave syndrome. *CMAJ 2021*; 193:E1499.
7. Kopelman Y, MD. Boerhaave syndrome in an elderly man successfully treated with 3-month indwelling esophageal stent. *Radiology Case Reports* 1 3 (2018) 1084–1086
8. Hager Aref et al, Laparoscopic Management of Boerhaaves syndrome: a case report with an intraoperative video.. *BMC Surgery* (2019) 19:109 https://doi.org/10.1186/s12893-019-0576-7
9. Rubicondo et al. Endoluminal vacuum-assisted closure (EVAC) therapy for postoperative esophageal fistula: successful case series and literature review *World Journal of Surgical Oncology* (2020) 18:301 https://doi.org/10.1186/s12957-020-02073-6
10. Jougon J, McBride T, Delcambre F, Minniti A, Velly JF. Primary esophageal repair for Boerhaave's syndrome whatever the free interval between perforation and treatment. *Eur J Cardiothorac Surg*. 2004; 25(4):475-9.
11. Weidenhagen R, Spelsberg F, Lang R, Jauch KW. New method for sepsis control caused by anastomotic leakage in rectal surgery: the Endo-VAC. *Color Dis* 2003; 5: 1–4
12. Diogo Turiani Hourneaux de Moura. Role of endoscopic vacuum therapy in the management of gastrointestinal transmural defects. *World J Gastrointest Endosc 2019* May 16; 11(5): 329-344.
13. Kuehn F, Loske G, Schiffmann L, Gock M, Klar E. Endoscopic vacuum therapy for various defects of the upper gastrointestinal tract. *Surg Endosc*. 2017 Sep;31(9):3449-3458. doi: 10.1007/s00464-016-5404-x. Epub 2017 Jan 11. PMID: 28078463.