

BOERHAAVE'S SYNDROME: DIAGNOSIS AND SUCCESSFUL PRIMARY REPAIR ONE MONTH AFTER THE OESOPHAGEAL PERFORATION

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SUMMARY

Boerhaave's syndrome (Spontaneous oesophageal perforation following forceful vomiting) is uncommon. However, when it occurs and the appropriate treatment is not given on time, it is fraught with early complications, leading to a very high mortality rate. This is a characteristic feature of this syndrome. Patient survival is in days. We present the case of an uncommon scenario of this syndrome in which the actual diagnosis was made one month after the oesophageal perforation, which was followed by primary repair, with a very good outcome.

Keywords: Boerhaave's syndrome, spontaneous oesophageal perforation, primary repair

INTRODUCTION

Spontaneous rupture of the oesophagus associated with forceful vomiting (Boerhaave's syndrome) is one of the most lethal diseases of the gastrointestinal tract, with a very high mortality rate. The rupture is transmural. It usually follows excessive alcohol intake or over eating, or both, because either of these can induce vomiting.

The syndrome 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", in which he discussed the case of Baron Jan van Wassenaer, a grand admiral of the Holland fleet, who after a feast, induced vomiting and consequently developed a severe left sided chest pain. He was dead within 24 hr.¹

Boerhaave's syndrome is an uncommon condition so the diagnosis can easily be missed or delayed, leading to complications like dehydration, mediastinitis, sepsis and shock. This is what accounts for the high mortality rate of 20 – 75% if appropriate treatment is not started on time. And if left untreated, the mortality reaches 100%.² Without adequate treatment, survival of Boerhaave's syndrome is in days.

CASE REPORT

A 57 year old man complained of 'violent vomiting' after a meal. Soon after the vomiting he felt a sudden onset left sided chest pain, followed by a dry cough and dyspnoea. He had not overeaten and he didn't take alcohol. The pain was constant and it radiated to the left shoulder. He was managed for food poisoning for two days in the regional hospital and was then referred to a tertiary hospital.

In the tertiary hospital, the initial impression was acute myocardial infarction, but chest x-ray done later showed massive left pleural effusion with mediastinal shift. A chest tube was inserted and it drained 1.8L of pus. Two days later he developed a fever. Subsequently, he was managed as a case of left chronic empyema thoracis, initially on broad-spectrum antibiotics and later by culture and sensitivity results.

After a month without much improvement (continuous copious daily drainage of 600 – 800ml of purulent effluent from the chest tube) he was referred to the Cardiothoracic Centre as a case of unresolving chronic empyema thoracis. He was sick and had lost weight but was in a stable condition. The wife added that the output from the chest tube seemed to increase with meals. Therefore with the suspicion of oesophageal perforation, he was given sweetened methylene blue dye orally. It appeared in the chest tube the next day.

He had thoracotomy the following morning. The findings were chronic empyema thoracis and oesophageal perforation. There were thickened parietal and mediastinal pleurae, 300ml of pus, fibrin peel entrapping the lung, and 2cm of longitudinal perforation in the distal oesophagus. The perforation edges were viable. They were neither necrotic nor oedematous.

Decortication was done; the perforation wound edges were freshened and closed primarily in two layers with vicryl 3/0 in interrupted fashion, and buttressed with the mediastinal pleura.

The pleural cavity was lavaged copiously with saline, a chest tube was inserted and the chest wall was closed in layers routinely.

Post operatively he was put on broad-spectrum antibiotics and total parenteral nutrition. On the 10th post operative day a gastrograffin swallow was done which showed free-flowing contrast from the oesophagus into the stomach, without any leakage. He was then started on oral feeding. He was discharged home on the 21st post operative day eating normal diet. There was no dysphagia. The follow-up period for over 2 years has been uneventful.



Figure 1 Chest x-ray 2nd day after the onset of the perforation showing a left pleural effusion and mediastinal shift.



Figure 2 Barium swallow 10 days after the primary repair. There was no extravasation seen.

DISCUSSION

Boerhaave's syndrome can be difficult to diagnose because there are no classic symptoms, and the perforation can also masquerade many clinical conditions like acute myocardial infarction, acute aortic dissection, tension pneumothorax³ or pneumoperitoneum.⁴

However, the most consistent feature in most cases is over indulgence in food and/or alcohol, followed by vomiting, then sudden onset severe left sided chest pain. The pain occasionally radiates to the left shoulder giving the false impression of acute myocardial infarction.

Since Boerhaave's syndrome and acute myocardial infarction have a lot in common regarding the clinical presentation, in most cases acute myocardial infarction is actually the first diagnosis considered. The common features are the age range (50 -70 years), the male gender, the sudden nature of the left sided chest pain, and the radiation to the left shoulder. Indeed, acute myocardial infarction was the first diagnosis considered when this patient arrived in the tertiary hospital.

Pathophysiologically, Boerhaave's syndrome is due to a sudden rise in intraluminal oesophageal pressure during vomiting, in the presence of a closed glottis. There is failure of the cricopharyngeous muscle to relax due to neuromuscular incoordination. Age is an important prerequisite factor for this neuromuscular incoordination. Anatomically the tear is located at the left posterolateral wall in the lower third of the oesophagus (in the chest). Free fluid and air leak into the posterior mediastinum and then into the left pleural cavity.

The second commonest site of rupture is subdiaphragmatic, leading to abdominal symptoms. When the syndrome is suspected the first investigation of choice is a contrast oesophagogram, preferably using gastrograffin, showing extravasation at the point of the perforation. Even though barium is superior in demonstrating small perforations its use is not advised since the extravasation leads to severe mediastinitis with consequent fibrosis, making later surgery difficult.

Alternatively, if the patient has a chest tube inserted (with the initial diagnosis of pleural effusion) then the methylene blue dye test can be done. When sweetened methylene blue is taken orally, it gives a bluish discoloration to the chest tube effluent within 12 – 24 hours. This is what was done for this patient.

Another investigation is a chest x-ray. It may show a left pleural effusion, pneumomediastinum or pneumothorax. CT scan may be useful in patients too ill for contrast studies. It can localize fluid collections and periesophageal air tracks suggestive of perforation but it cannot precisely localize the site of the perforation. Oesophagoscopy is not encouraged because it can widen the perforation and also introduce more air into the mediastinum and the left pleural cavity.

Concerning management, perforations diagnosed within 12 – 24 hours are considered early perforations whilst those diagnosed after 24 hours are late perforations.^{4,5} Early perforations have the best outcome because most complications have not yet set in. They are treated by reinforced primary repair.^{5,6}

The problem (and the controversy), is with the late perforations. By the time diagnosis is made the wound edges have become oedematous, stiff or friable rendering primary repair risky due to the high rate of breakdown. Usually, there is also associated mediastinitis and empyema thoracis. Late perforations are therefore managed by debridement of the pleural cavity and mediastinum, oesophagectomy, cervical oesophagostomy, and feeding gastrostomy.^{5,7}

Oesophageal replacement is done after 6 weeks, usually with the colon or the stomach.⁶ On the contrary, other authors⁸ advocate that reinforced primary repair can be done for most late perforations. Jougon et al even advocate that all oesophageal perforations can have primary repair no matter the time between perforation and treatment.⁹ This approach might not be very safe. We are of the opinion that not all but some of the late perforations may benefit from primary repair. Our patient was diagnosed one month after the perforation but since the perforation edges were viable he had reinforced primary repair with a very good outcome.

The peculiar features in this patient are the unusual one month long interval between perforation and diagnosis, being in a stable state for surgery, finding viable perforation edges at surgery, having primary repair, and having a good outcome.

CONCLUSION

This is an unusual case of Boerhaave's syndrome that was diagnosed one month after the perforation. And unlike in most cases, he had primary repair with a very good outcome.

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