Boerhaave's Syndrome: Early versus Delayed Intervention

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INTRODUCTION

Boerhaave's syndrome is a spontaneous transmural esophageal rupture caused by sudden increase in intraluminal pressure. It accounts for around 15-30% of cases of spontaneous esophageal rupture (Law, T.T. 2017; & Ali, J.T. 2017). It is associated with high fatality with a mortality rate of 20-40% in patients with early presentation and intervention (Curci, J.J. 1976). In untreated patients with delayed presentation, the disease is nearly fatal with mortality of 50-100% (Curci, J.J. 1976; Turner, A.R. 2019; Brauer, R.B. 1997). Boerhaave's syndrome can be treated with conservative, endoscopic or surgical approach. Survival rates being 75%, 100% and 81% respectively in case of early presentation without complications (Schipper, J.P. de. 2009) with no consensus on the most suitable treatment option. Studies have suggested endoscopic management in patients with early presentation without complications (Schipper, J.P. de. 2009). For patients with delayed presentation, studies suggest early conservative and delayed surgical approach (Schipper, J.P. de. 2009). We report two cases of Boerhaave's syndrome with delayed and early presentation respectively in the order of presentation to our hospital.

Abstract: Boerhaave's syndrome is a rare but lethal disease involving the gastrointestinal tract. It is a spontaneous esophageal perforation with complete transmural tear caused by a sudden increase of intraluminal pressure in the esophagus. The disease is associated with high mortality of more than 40% with early surgical intervention. Delayed surgical intervention more than 7 days after the initial presentation is associated with nearly 100% mortality. The classical Mackler's triad of symptoms is rare. The reported rate of misdiagnosis is high. Delayed presentation, misdiagnosis, and intervention is associated with poor healing, increased complications, prolonged hospital stay, and increased mortality. We report two cases of Boerhaave's syndrome. One case presented early within 24hours with classical symptoms, while another presented late after 7 days with atypical symptoms and a delayed diagnosis elsewhere. Both the cases where managed by recommended standard practices. Despite minimal complications, the patient with early presentation had a comparatively good healing, early recovery, and short hospital stay. The patient with delayed presentation had already developed multiple complications at presentation to our setup. Though this patient required a prolonged hospital stay, an intensive, imaginative, and aggressive multidisciplinary care resulted in a good recovery. A high index of suspicion and early intervention helped in the recovery and prevention of complications in patients presenting early. Good multidisciplinary care resulted in a good recovery in the patient with delayed presentation. With a good and imaginative multidisciplinary care, even a condition with a mortality rate as high as 100% can be managed successfully.

Keywords: Boerhaave Syndrome, Mediastinitis, Esophageal Perforation.
CASE DESCRIPTION

Case 1

60 year old male with no comorbidities was admitted to a nearby tertiary hospital with complaints of cough with expectoration for two days and one episode of forceful vomiting followed by chest pain and tightness in left side of the chest. He was found to have left sided pleural effusion which required intercostal drain (ICD) placement with foul smelling purulent collection drainage. During this period, he became hemodynamically unstable with worsening oxygenation and acute kidney injury requiring intubation, mechanical ventilation, vasopressor support and hemodialysis. His blood and endotracheal aspirate was growing Candida tropicalis. Computed Tomography (CT) of chest showed pneumothorax and esophagitis. Upper GI endoscopy showed distal esophagus transmural perforation. He was shifted to our hospital for further management. Repeat endoscopy showed long linear lower esophageal tear which was approximated by clipping and self expandable metal stent (SEMS). Patient was started on naso-gastric tube feeds and continued on vasopressors; antibiotic support and renal replacement therapy. Repeated cultures showed presence of XDR Klebsiella in blood, endotracheal aspirate and pus requiring polymyxin antibiotics. Throughout the period, the patient had multiple episodes of atrial fibrillation managed with amiodarone and beta blockers. In view of continued fever, video assisted thoracoscopic surgery(VATS) with decortication and tracheostomy was done. Despite this, the patient had persistent fever. We repeated CT of chest which showed contrast extravasation suggestive of esophageal leak. Hence, left thoracotomy, cervical esophagostomy and feeding jejunostomy was done. Patient remained in septic shock in spite of supportive measures and antibiotic optimisation. Repeat CT of chest showed left sided lung volume loss; collapse, consolidation of lung; bronchiectatic changes and pleural effusion. Intrapleural amikacin was given to control infection and patient was gradually weaned to high flow oxygen device through tracheostomy. Definitive surgery with Right thoracotomy, esophagectomy with substernal cervico esophagogastric anastomosis was done. Extensive mediastinitis involving entire esophagus and periesophageal dense adhesions were noticed. Small tracheal rent of 5-6mm which occurred intraoperatively was sutured. Post operatively, in view of persistent leak noticed on the ventilator, nasal endoscopy guided placement of tracheostomy tube beyond tracheal rent was done. As ventilatory leak persisted, bronchscopy was done which showed posterior tracheal wall linear fistula less than 1 cm². Adjustable flange tracheostomy tube was placed with cuff beyond fistula opening. With time, ICD drain output reduced and all ICDs were gradually removed. Repeat bronchoscopy showed a healed tracheal rent. Patient was gradually weaned from ventilator; however, the decannulation attempt of tracheostomy failed. Repeat CT showed left lung collapse, pleural thickening and hydropneumothorax. Patient was slowly mobilised with active chest and limb physiotherapy throughout this period. After 6 months of ICU stay, patient was discharged with tracheostomy tube and home nursing care support. The patient had multiple episodes of admissions to the hospital over time with dyspnea, lung collapse due to secretions and tachyarrhythmias. Although the patient failed multiple attempts of decannulation over time, normal oral feeding was established with no hemodynamic issues; and now, he is mobile with continued rehabilitation at home.

Case 2

50 year old male was admitted with sudden onset chest discomfort and severe back pain of one hour duration following an episode of forceful vomiting. CT chest showed lower esophageal rupture with large hydropneumothorax, compressive collapse of left lung with mediastinal and surgical emphysema. Emergency left posterolateral thoracotomy with primary esophageal repair was done after intercostal drain insertion on the day of admission. Patient was extubated on second post operative day. By post operative day 6, the patient developed fever. The CT gastrograffin study showed smooth contrast flow across anastomotic site with large loculated collection in posterior pericardial recess. Empirical antibiotic was started as per hospital antibiotic policy. Patient continued to have fever and repeat CT chest showed contrast extravasation near gastro esophageal junction. The Upper Gastro-intestinal endoscopy showed fistula at lower end of esophagus. Pericardiocentesis was done for the collection shown on CT in view of persistent fever and hemodynamic instability. Patient condition gradually improved and he was discharged home after starting oral feeds after one month of hospital stay.

DISCUSSION

Boerhaave's syndrome was first described in 1724 by Herman Boerhaave (Nakano, T. 2018). The classical presentation of Boerhaave's syndrome is known as Meckler's triad which includes chest pain, vomiting and subcutaneous emphysema. Though combination of these symptoms are rarely seen posing a diagnostic challenge (Han, D. 2018). It is commonly misdiagnosed as perforated ulcer, myocardial infarction and pancreatitis (Schipper, J.P. de. 2009). A chest X-ray can be normal in 10-15% cases (Ferreiro-Iglesias, R. 2017). Oesophagogastroduodenoscopy(OGD) has sensitivity and specificity of 100% and 83% respectively. In case of high index of suspicion a CT chest with oral contrast should be done. Conservative, endoscopic and surgical approach have been used previously (Law, T.T. 2017; Schipper, J.P. de. 2009). But these are more effective with early presentation without mediastinitis and other complications.
The first case presented to our hospital after nine days of his first symptom. By the time he presented to us, he was in septic shock with candidemia, mediastinitis, esophagitis, ventilator acquired pneumonia and acute kidney injury. After initial stabilization, he was taken for urgent endoscopic stenting. In view of persistent infection and mediastinitis, he underwent VATS and multiple ICD placement. The patient developed tracheal tear during definitive surgery because of dense adhesions. Imaginative thinking on the part of intensivists, pulmonologist and otolaryngologist and management with long flange tracheal tube helped in healing of tear. A good nutrition management, thromboprophylaxis, continued nursing and physiotherapy care with a motivated patient and family support help in recovery of the patient.

The second case presented within an hour after vomiting. With our previous experience with Boerhaave's syndrome an urgent CT chest helped in early diagnosis. He was taken for immediate surgery in view of lung collapse and hydropneumothorax after placing ICD. Anastomotic site leak has been reported in 17-32% cases post surgical repair. This patient also had some anastomotic leak which was effectively managed.

**CONCLUSION**

With our limited experience, we conclude that early presentation of Boerhaave syndrome has a better outcome compared to late presentation. However, a good outcome can be seen in delayed presentation of Boerhaave’s syndrome with an effective multidisciplinary approach; patient motivation, perseverance and family support. Clinicians experience in managing such conditions helps in improving outcome, reducing morbidity and mortality.

**Clinical Significance:** This article highlights the fact that a high index of suspicion even in cases with atypical presentation can help in early diagnosis. Clinician experience and multidisciplinary approach improves patient outcome.

**REFERENCES**