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CASE REPORT

BOERHAAVE'S SYNDROME: A DIAGNOSTIC DILEMMA IN THE EMERGENCY ROOM

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Boerhaave's syndrome is a potential lethal condition which presents not only a diagnostic but also a therapeutic challenge. Errors in diagnosis are usually caused by unawareness of its varied and atypical presentations. All clinicians need to be aware of this lethal disease, its frequent unusual presentations and the importance of early diagnosis.

Keywords: Boerhaave's Syndrome, oesophageal rupture,

INTRODUCTION

Boerhaave's syndrome or spontaneous esophageal rupture is a rare, but potentially lethal condition. It usually follows forceful vomiting with subsequent mediastinitis and in the absence of therapy is associated with high morbidity and mortality. Because of the lack of awareness about this rare condition, it may go unrecognized.[1] The presenting picture mimics other common conditions and as a result crucial intervention is delayed. Mortality rates as high as 100 percent have been reported in some case-series after one week of nonsurgical medical care.[2,3] We present the case of a patient with spontaneous esophageal rupture in which the diagnosis was delayed because of unusual presentation.

CASE REPORT

An 82-year-old retired electrical engineer presented to the emergency room with severe left sided chest pain and difficulty breathing for the past three hours. His past history included mild asthma and ischemic heart disease. He had had coronary artery bypass surgery about 15 years back. His attendant, gave the history that while having lunch, the patient had three episodes of vomiting followed by intense chest pain and shortness of breath. He was managed as angina in another hospital, and was treated with sublingual nitroglycerine without any relief in symptomatology.

Physical examination revealed an anxious gentleman in mild respiratory distress. Vital signs revealed blood pressure of 136/70 mm Hg, pulse rate 88 beats per minute, respiratory rate 28/min, and temperature of 36.5° C. Chest auscultation revealed diminished breath sounds, egophony, decreased tactile fremitus and dullness to percussion at the left lung base. The abdominal examination revealed mild epigastric tenderness but no guarding or rebound tenderness. Laboratory studies showed a white blood cell count of 11,000 cells per micro liter, with 65% neutrophils and 26.3% lymphocytes. Electrocardiogram showed no new ischemic changes. The chest x-ray revealed left-sided alveolar infiltrates along with pneumothorax (Figure-1).The arterial blood gases on 3 liters of oxygen, revealed a PaO_2 of 51.1 mmHg, $PaCO_2$ of 34.2 mmHg, pH of 7.44 and arterial oxygen saturation of 87.7%.

Figure 1 Chest radiograph showing left sided infiltrates with pneumothorax

Based on clinical symptomatology and radiological findings, the diagnosis of left sided aspiration pneumonia with small pneumothorax was made and patient was admitted under pulmonology service. Ultrasound guided thoracentesis was suggested but the attendant refused it. The patient was started on empiric antibiotic therapy with high flow supplemental oxygen. The patient's condition deteriorated the next morning. He was found to be hypotensive and in respiratory distress. He was intubated with size 8.0 endotracheal tube and a left chest tube was urgently placed. The chest tube drained about 400 ml of dirty grey, foul smelling fluid which also contained food particles. The biochemistry of fluid showed glucose of 15mg/dl, protein of 788mg/dl, lactate dehydrogenase of 4650

IU/L and amylase of 65770 IU/L. The pleural fluid was sent for culture and sensitivity.

Cardiothoracic surgery was consulted and the patient was urgently taken to the operating room. He underwent a left posterolateral thoracotomy and the pleural cavity was entered through the sixth intercostal space. Approximately one liter of greyish brown fluid was drained. A 6-cm perforation was identified just proximal to the gastroesophageal junction and extensive mediastinal necrosis was seen. The perforation was closed primarily in two layers with an intercostal flap overlay. Diversion surgery by esophagostomy was also done and a feeding jejunostomony was created (Figure-2).

Figure-2: Chest radiograph after surgical intervention

The patient was transferred to the intensive care unit. The post operative course was complicated by severe sepsis / shock requiring multiple inotropic drugs. Blood cultures grew E.Coli. Pleural fluid grew gram negative rods and gram positive cocci. After 3 days, he developed acute renal failure requiring dialysis. His condition deteriorated further and he developed multi organ failure. He died after 20 days in the ICU.

DISCUSSION

This syndrome was first described by Herman Boerhaave (1669-1738) from Leiden, The Netherlands (Atroscis, nec descripti pruis, morbid historica).[4] His patient was a 50-year-old Grand Admiral who died in 1723 after 18 hours of self induced vomiting which resulted in esophageal rupture. Since then, the term Boerhaave's syndrome is used to describe a full thickness perforation of the esophagus not associated with external trauma, instrumentation or foreign bodies.

The diagnosis of this uncommon condition is often missed or delayed, because of a non-specific presentation in the majority of cases. This syndrome can be confused with pneumonia, pneumothorax, lung abscess, myocardial infarction, pancreatitis, dissecting aortic aneurysm, pulmonary embolus, pericarditis or perforated ulcer.[1] Early diagnosis and prompt surgical intervention is life saving. A delay of 12 hours or more between symptoms and operative repair is associated with a reported 36 percent mortality; with a delay of 24 hours this almost doubles to 64%.[5] It may approach nearly 100 percent without any surgical intervention.[2] Boerhaave's syndrome is traditionally associated with the triad of vomiting, chest pain and subcutaneous emphysema as described by Meckler[6], but the patient may present with atypical symptoms so reliance on a 'classic' presentation may be misleading[7]. Unusual clinical features should also be kept in mind while evaluating patients with a suspicion of esophageal rupture.

The mechanism of injury as elucidated by Rogers and associates [8] is an increase in intragastric pressure coupled with opening of lower esophageal sphincter to allow entry of gastric contents into the lower esophagus. If the upper sphincter does not open, hydrostatic pressure within the esophagus rises to a point when the wall gives way at its weakest point; the posterolateral wall on the left. Spillage of corrosive gastric acid, enzymes, food and bacteria into the mediastinum leads to severe mediastinitis and empyema causing a sepsis syndrome and shock.

Chest radiograph may show a pleural effusion, pneumothorax or the 'V' sign of Naclerio, a density behind the left cardiac border. Pate et al[2] in their series found that the initial chest roentgenogram was abnormal in 97% of the patients but was interpreted as compatible with perforation of esophagus in only 27%. Contrast studies are positive in 75 percent of cases; however, a lack of extravasation does not necessarily exclude a perforation. Pleural fluid examination can be an adjunct to diagnosing Boerhaave's syndrome as described by Drury et al.[9] Their patient had a negative barium swallow but subsequent pleural fluid testing showed an acidic pH with an increased amylase level of salivary origin. Pleural effusion cytology showed undigested food particles. The role of upper GI endoscopy in the presence of esophageal rupture is controversial because some authorities believe that insufflation of air can extend the perforation.

Various treatment options are available, both surgical and non-surgical. Non-surgical treatment is recommended for patients with small perforations, contamination confined to the mediastinum and late recognition (more than 24 hours) of an esophageal perforation, as the surgical mortality at 24 hours becomes equal to that of a more conservative approach.[10]

Surgical intervention which includes open thoracotomy, debridement and surgical closure of the perforation is recommended if the diagnosis is made early. Diversion surgery by esophagostomy, esophagectomy or T-tube diversion[11] is usually reserved for critically ill patients. Use of self expanding metallic stent has been reported to be successful if diagnosis is made within 24 hours.[12]

The truism that a diagnosis will only be made if it is borne in mind applies with particular force to this life threatening condition that characterizes spontaneous esophageal perforation. All clinicians need to be aware of the manifestations of Boerhaave's syndrome and the importance of early diagnosis.

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