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Case report

Tension hydropneumothorax as the initial presentation of Boerhaave syndrome

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ABSTRACT

Boerhaave syndrome, a rare yet frequently fatal diagnosis, is characterized by the spontaneous transmural rupture of the esophagus. The classic presentation of Boerhaave syndrome is characterized by Mackler's triad, consisting of chest pain, vomiting, and subcutaneous emphysema. However, Boerhaave syndrome rarely presents with all the features of Mackler's triad; instead, the common presentation of Boerhaave syndrome includes chest or epigastric pain, severe retching and vomiting, dyspnea, and shock. These symptoms are typically misdiagnosed as cardiogenic in origin. Due to its atypical presentation, rarity, and mimicry of emergent conditions, diagnosis of Boerhaave syndrome is often delayed, resulting in a high mortality rate at the time of diagnosis and with a subsequent exponential increase in mortality if treatment is delayed by greater than 48 hours. Here, we report two atypical presentations of Boerhaave syndrome presenting as tension hydropneumothorax and review ten previously reported cases of Boerhaave syndrome presenting as tension hydropneumothorax. This review serves to raise clinician awareness about the expansive and elusive ways by which esophageal perforation may present, and thereby facilitate timely and potentially life-saving diagnosis.

1. Introduction

Spontaneous transmural rupture of the esophagus, or Boerhaave syndrome, was first described in 1724 by Hermann Boerhaave [1]. Mackler's triad, consisting of chest pain, vomiting, and subcutaneous emphysema represents a classic presentation. However, Mackler's triad is only present in about 5% of case, with chest pain being the most common feature [2,3]. Boerhaave syndrome itself is rare, with an annual incidence of 3.1 per 1,000,000 [4]. Coupled with its mimicry of other conditions, the diagnosis of Boerhaave syndrome is often delayed, with mortality rates up to 30% at the time of diagnosis that significantly increase by treatment delays > 48 hours [5]. We report two cases of Boerhaave syndrome with tension hydropneumothorax as the initial presentation and review other reported cases.

2. Case presentations

2.1. Case 1

A 65-year-old male with hepatitis C, alcoholism and prior small bowel obstruction (SBO) from abdominal surgery was admitted with 3 days of abdominal pain, emesis and recurrent partial SBO. The admission chest x-ray (CXR) was unremarkable. On hospital day 2, he developed acute chest pain with electrocardiogram demonstrating ST-segment elevation. Emergent cardiac catheterization revealed no obstructive coronary artery disease. Post procedure, he developed excruciating chest pain radiating to his back with tachycardia, hypotension and absent breath sounds over the left lung. CXR demonstrated interval opacification of the left hemithorax with mediastinal shift. An emergent needle thoracostomy produced air and dark fluid containing food debris, followed by tube thoracostomy which drained 4 L of bilious, enteric output. Chest computerized tomography (CT) demonstrated a large left hydropneumothorax with free air around the esophagus (Fig. 1). Esophagogastroduodenoscopy demonstrated “black esophagus” in a circumferential distal third of the esophagus consistent with necrosis and perforation. He underwent emergent surgery with cervical esophagostomy, laparotomy, and gastrostomy tube placement. His post-operative course was complicated by hypoxemic respiratory failure and on hospital day 21, massive gastrointestinal hemorrhage and death.

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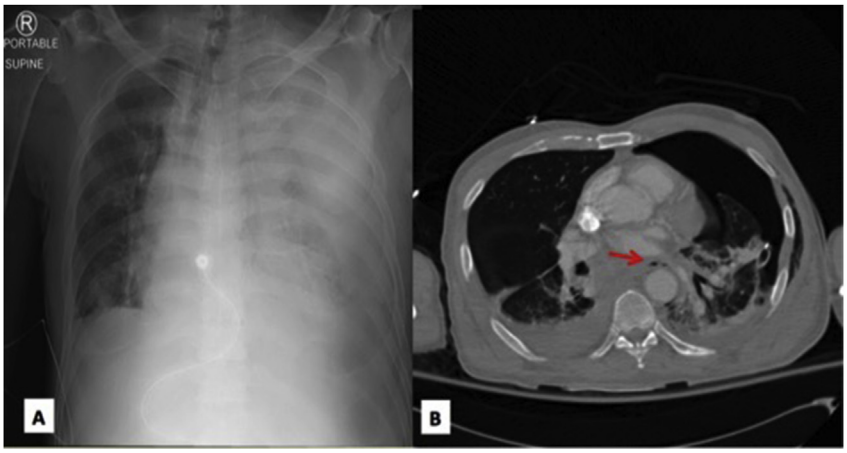

Fig. 1. A) AP Chest radiograph demonstrating complete opacification of the left lung with mediastinal shift to the right and a small right-sided pleural effusion. B) Chest CT demonstrating extra-luminal free air in the mediastinum (red arrow), left-sided hydropneumothorax and small right-sided pleural effusion. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

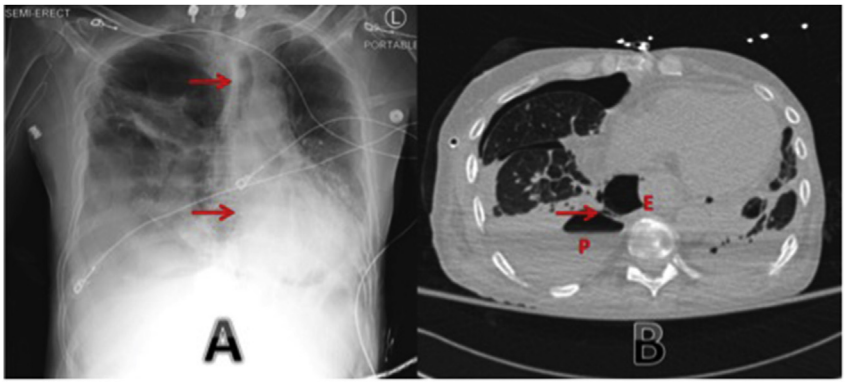

Fig. 2. A) AP Chest radiograph demonstrating a right-sided tension hydropneumothorax with mediastinal shift to the left (red arrows). B) Chest CT demonstrating free air around the esophagus (E) communicating (red arrow) with the pleural space. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Table 1
Presenting symptoms of tension hydropneumothorax in Boerhaave syndrome.

Symptoms	% of patients presenting with symptoms
Chest pain	6/12 (50%)
Epigastric pain	4/12 (33%)
Vomiting	11/12 (92%)
Dyspnea/Tachypnea	8/12 (67%)

2.2. Case 2

A 60-year-old male with alcoholism and chronic dysphagia secondary to Barrett's esophagus and achalasia was admitted with alcohol withdrawal and aspiration pneumonia. Notably, he had severe dysphagia for which he would self-induce vomiting to alleviate sensations of choking an alleviate hiccups. He failed a swallow evaluation and nasogastric tube placement was attempted but was unsuccessful. On hospital day 10, he developed tachycardia, tachypnea, hypoxemia, and chest pain, with opacification of the right hemithorax with mediastinal shift (Fig. 2). An emergent needle decompression was performed, followed by tube thoracostomy with drainage of over 1 L of exudative fluid (pH of 6.9, amylase > 6000 U/L). Chest CT confirmed free air around the esophagus in communication with the pleural space consistent with esophageal perforation. He underwent surgical repair with clinical improvement. Patient remained hospitalized undergoing physical therapy, but developed a fatal intracranial bleed on post-operative day 30.

3. Discussion

The classic presentation of Boerhaave syndrome includes chest or epigastric pain (83%), severe retching and vomiting (79%), dyspnea (39%), and shock (32%) [6]. Esophageal rupture occurs when an abrupt increase in intraluminal esophageal pressure forces gastric contents against the cricopharyngeal muscles causing transmural esophageal rupture [7]. As the esophagus ruptures, the parietal pleura can also rupture or become breached by gastric enzymes, leading to pneumothorax or hydropneumothorax. If the esophageal perforation is sealed, patients may appear deceivingly well without symptoms and/or hemodynamic instability [6].

Boerhaave syndrome presenting as tension hydropneumothorax is an extremely rare phenomenon. A MEDLINE/PubMed and Google Scholar search with search terms of “Boerhaave syndrome”, “esophageal rupture”, “esophageal perforation”, “tension hydropneumothorax”, “tension pneumothorax”, and “pneumothorax”, only identified 10 reports of tension hydropneumothorax in Boerhaave syndrome in addition to our two cases.

In patients with tension hydropneumothorax as the initial presentation of Boerhaave's syndrome, the most common symptoms were recent vomiting (11 of 12), dyspnea (8 of 12), chest pain (6 of 12), and epigastric pain (4 of 12) (Table 1). Mackler's triad of chest pain, vomiting, and subcutaneous emphysema was not seen in any of the 12 cases in our series, suggesting the classical teaching of Mackler's triad is neither sensitive nor specific for this diagnosis.

The non-specific presenting symptoms and mimicry of other emergent conditions often require additional studies to establish the diagnosis. In these 12 cases, the initial imaging modality was a CXR, which revealed tension hydropneumothorax – a rare presenting sign for esophageal rupture (Table 2). Boerhaave syndrome may also be suspected based on mediastinal widening, pneumothorax, hydrothorax, hydropneumothorax, mediastinal shift, and/or subcutaneous emphysema [8]. Esophagram or CT scan with water-soluble oral contrast is the gold standard for diagnosing esophageal perforation [2,3]. However, due to the high false negative rate of the contrast esophagram (15–25%) and

Table 2
Boerhaave syndrome with tension hydropneumothorax: reported cases.

Source	Age	Sex	Initial imaging modality	Confirmatory imaging	Thoracic drainage	Mortality
Current report–Case 1	65	M	CXR - Complete opacification of lung with mediastinal shift to the right, small right-sided pleural effusion	CT Chest - Extraluminal free air in the mediastinum, left-sided hydropneumothorax and small right-sided pleural effusion	4 L bilious, enteric output	Expired
Current report-Case 2	60	M	CXR - Right-sided tension hydropneumothorax with mediastinal shift to the left	CT Chest - Free air around the esophagus communicating with the pleural space, left pleural effusion	1 L exudative fluidwith high amylase, pH 6.9	Expired
Nguyen Ho 2016 [11]	48	M	CXR - Left-sided hydropneumothorax; Subsequent CXR - Large pleural effusion in left hemithorax	Chest CT - Left pleural effusion and mediastinal air in distal esophagus	Pleural fluid with amylase 9713 U/L	Survived
Vallabhajosyula 2016 [2]	43	F	CXR - Right-sided tension pneumothorax, pleural effusions	CT Chest with water soluble oral contrast - Long segment tear of right anterolateral distal esophagus with adjacent contrast leak into mediastinum and right pleural cavity	Turbid yellow fluid with high neutrophils and protein, normal amylase, cholesterol, and LDH	Unknown
Ijaz 2015 [12]	21	M	CXR - Left-sided hydropneumothorax	CT Chest - Left pneumothorax, no mediastinal air; Subsequent CT Chest 12 days later - direct communication between esophagus and left pleural space	500 mL purulent, brown-green fluid - exudative, neutrophilic. Subsequent drainage chylous – triglycerides 1041 mg/dl, repeat pleural fluid analysis with amylase 12238 U/L	Expired
Tanaka 2014 [13]	45	M	CXR - Massive left-sided pleural effusion, left-sided tension pneumothorax	CT Chest - Esophageal wall thickening, displacement of lower esophagus with mild surrounding mediastinal emphysema	2 L green digestive juice	Survived
Veno 2013 [14]	67	M	CXR - Tension pneumothorax with right-sided mediastinal shift, pleural effusion	Esophagram - Contrast extravasation from esophagus on left inferior side	1.5 L white fluid, Subsequent thoracotomy with gastric content in thoracic cavity and empyema	Survived
Keane 2012 [15]	30	M	CXR on admission - Right-sided pneumonia, pleural effusion, small pneumothorax. Repeat CXR after 48 hrs - sustained right-sided pneumothorax, complete opacification of the lung	CT Chest- Hydropneumothorax with mediastinal shift and contrast leak between distal esophagus and right pleural cavity	Brown thick fluid aspirate	Unknown
Suzuki 2012 [10]	80	F	CXR - Right sided mediastinal shift	CT Chest - Left-sided pneumothorax, left pleural effusion, pneumomediastinum at lower level of esophagus	Food debris (broccoli)	Survived
Doherty 1996 [16]	47	M	CXR - Tension pneumothorax, left-sided effusion, pneumomediastinum, cervical subcutaneous emphysema	CT Chest - Gastrograffin entering left pleural cavity, mediastinal emphysema, left pneumothorax, total collapse of LLL and RLL	Needle decompression - “brownish fluid”; Tube thoracostomy drained > 5L of fluid (pH not acidic)	Unknown
Onyeka 1999 [17]	72	F	None - Clinical diagnosis	Gastromiro study - Right lower linear esophageal rupture	Needle decompression; Tube thoracostomy draining 200 mL of bile-stained fluid containing food debris, pH 6.0.	Expired
Zamir 1995 [18]	68	M	CXR - Left-sided tension pneumothorax and large left pleural effusion	Gastrograffin study - Left supradiaphragmatic extravasation	Unspecified	Survived

Abbreviations: Chest roentgenogram (CXR), computed tomography (CT).

tenuous nature of esophageal rupture, chest CT is a more appropriate modality for diagnosis. Extraluminal and/or mediastinal free air are the most common findings, occurring in 92% of cases [2,3]. Additional chest CT findings that are suspicious, but not specific for esophageal rupture include esophageal thickening, mediastinal fluid, or pleural effusions [9].

Tube thoracostomy and drainage of pleural effusions can rapidly diagnose Boerhaave syndrome. Food particles or biliary contents with pH < 6, high amylase content, salivary squamous cells, and polymicrobial gram stain or cultures from the pleural fluid are all diagnostic of esophageal communication with the intrathoracic space [10].

Boerhaave syndrome requires aggressive surgical or endoscopic therapy. The best predictor of mortality is the timing of diagnosis. Shaker and colleagues noted diagnosis and surgery within 24 hours of presentation decreased mortality from 44% to 5.5% in 27 patients with esophageal rupture [5]. A delay between symptom onset and procedure of ≥12 hours appears to be associated with a 36% mortality rate, while a delay of ≥24 hours can increase mortality rates up to 80% [5,10]. Prompt management of Boerhaave syndrome is instrumental for survival.

Mackler's triad is rarely encountered in Boerhaave syndrome, and in patients with hydropneumothorax; vomiting, chest pain and dyspnea are the most frequent symptoms. In addition to imaging, thoracentesis and/or tube thoracostomy are essential in establishing this diagnosis and should be performed expeditiously to minimize delays in treatment. The rapid appearance of a large pleural effusion and hydropneumothorax in a patient with chest pain is another finding that can suggest this diagnosis.

Authorship statement

All individuals who quality as authors have been listed. Each has participated in the conception and writing of the manuscript, the collection and analysis of data, and the critical editing of the manuscript.

Disclosure statement

The authors declare no conflicts of interest.

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