Boerhaave Syndrome Presenting as Abrupt Onset of Massive Hydropneumothorax

Boerhaave syndrome is a rare and life-threatening disease that often presents a diagnostic challenge. It is usually confused with critical but more prevalent diseases such as acute myocardial infarction, perforated peptic ulcer, and acute pancreatitis. Boerhaave syndrome is caused by forceful vomiting resulting in a full-thickness tear of the middle or lower esophagus, typically an area of natural narrowing and at the esophagogastric junction and the left atrium. Because of these anatomic sites, hydropneumothorax, hemopneumothorax and pneumopericardium can occur. We report a case of a 48-year-old chronic alcoholic man presenting with abrupt onset of massive bilateral hydropneumothorax. In this case, it was hard to take a medical history from the patient due to sudden respiratory arrest when he arrived at the emergency room. Despite ongoing chest tube drainage, hydropneumothorax didn't improve. Pleural fluid amylase level was increased. Because of the possibility of esophageal rupture, esophagography was performed. As a result of the esophagography, he was diagnosed as Boerhaave syndrome with pneumopericardium. If massive hydropneumothorax of unknown cause presents abruptly, Boerhaave syndrome should be suspected as one of its causes. We recommend that pleural fluid amylase levels to be checked and if it is elevated, esophagography should be performed immediately.

INTRODUCTION

An esophageal perforation is a serious condition with significantly high morbidity and mortality rates despite advances in diagnostics, surgery, and critical care. Iatrogenic and spontaneous perforations are two general etiologies. Other less common causes include perforations secondary to malignancy or trauma. Medical procedures cause over half of all perforations and can be caused by any instrument that enters the esophagus. Out-of-hospital perforations are typically traumatic or spontaneous. Among these, the presence of a spontaneous perforation is called Boerhaave syndrome, because it was first described in 1724 by the Dutch physician Herman Boerhaave at Leiden University. Boerhaave syndrome most commonly results from a sudden increase in intraluminal esophageal pressure combined with negative intrathoracic pressure after excessive vomiting and retching. The most common anatomical location for a tear in patients with Boerhaave syndrome is the left posterolateral wall of the lower third of the esophagus, 2~3 cm above the stomach. Typically, Meckler's triad of chest pain, vomiting, and subcutaneous emphysema are characteristic features of Boerhaave syndrome. However, occasionally, nonspecific symptoms may contribute to a delay in diagnosis and a poor outcome. A delayed diagnosis is usually caused by a lack of knowledge and ambiguous clinical symptoms. Because of this delay in diagnosis and a lack of suspicion, the mediastinitis and sepsis caused by the combination of gastric acid and digestive enzymes can contribute to fatal conditions. Here, we report the
case of a 48-year-old man with chronic alcoholism and no history of pulmonary or cardiac diseases, presenting with abrupt onset massive bilateral pleural effusion. After check-up of elevated pleural amylase level and esophagography findings using a Levin tube, he was finally diagnosed with Boerhaave syndrome and an esophago-pericardial fistula.

CASE REPORT

A 48-year-old man with chronic alcoholism was referred to our emergency room (ER) with a diagnosis of bilateral pleural effusion on a chest X-ray after 2 visits to other hospitals 24 hours and 3 hours ago. During his first visit, he complained of epigastric pain and general weakness. He was alert at the time, and had stable vital signs. Only a mild costophrenic angle blunting was found on his chest X-ray (Fig. 1). During his second visit at another local hospital, he went into respiratory arrest immediately after giving his name, and cardiopulmonary resuscitation (CPR) was performed for 10 minutes. After CPR, his condition improved and he was diagnosed with a hemopneumothorax as a result of the chest computed tomography (CT). Subcutaneous emphysema with crepitus was detected during physical examination. The chest tube drainage was 1.8 L (left) and 1.6 L (right), respectively. The drained fluid was dark-colored, and his serum hemoglobin (Hb) was 16.7 g/dL. He was transferred to the ER under mechanical ventilation with stable vital signs. Laboratory findings were leucopenia (white blood cells, 1,400/μL) with a marked elevation in C-reactive protein (26.21 mg/dL) and thrombocytopenia (117,000/μL) without anemia (Hb, 16.0 g/dL). A pleural effusion analysis after a thoracentesis showed characteristic bloody exudates with a marked elevation in pleural amylase level (993 IU/L). According to a later report, pleural effusion cultures and a blood culture revealed methicillin-sensitive Staphylococcus aureus, and Streptococcus mitis/oralis. During his first hospital visit, approximately 24 hours earlier, a chest X-ray was nearly normal,
except for mild left costophrenic angle blunting (Fig. 1A). During the second visit, approximately 3 hours earlier, he had a large bilateral pneumomediastinum with bilateral chest tube insertion (Fig. 1B). Although the chest tube drainage was ongoing, he had a persistent bilateral pneumothorax on chest X-ray. An enhanced chest CT image showed a bilateral hemothorax and mediastinal air (Fig. 2). Side effects from the CPR are possible. A chest CT of the lung revealed a peripneumomediastinum and a large bilateral pneumothorax (Fig. 3). Because of the characteristic increase in pleural amylase, we suspected Boerhaave syndrome, so an esophagography via a Levin tube using nonionic and iodinated water-soluble contrast agent (Ultravist: Bayer, Berlin, Germany) was performed, which demonstrated marked extravasations from the left distal esophagus to the pericardium (Fig. 4). With the abrupt onset of massive pleural effusion and no medical history, he was diagnosed with Boerhaave syndrome and an esophago-pericardial fistula. Emergency primary esophageal repair was performed on a 2.5 ~ 3 cm longitudinal laceration in the lower esophagus towards the right mediastinal pleura. The esophagogastric junction was repaired, but he finally died of sepsis postoperatively.

DISCUSSION

A spontaneous esophageal perforation is challenging territory for clinicians who have specialty in gastrointestinal disturbances. This condition was described for the first time in 1724. Spontaneous esophageal perforation’s incidence is known to be 1 in 6,000 patients. Bang et al. first reported Boerhaave syndrome in Korea. Pleural effusion presenting as a first symptom of Boerhaave syndrome has been reported very rarely, but Kang et al. did report Boerhaave syndrome presenting as recurrent pleural effusion in one case of Boerhaave syndrome in 2000. The occasional nonspecific nature of the symptoms may contribute to a delayed diagnosis and high mortality rate. An esophageal perforation, caused by forceful vomiting, results in a full-thickness tear of the middle or lower esophagus. The site is typically an area of natural narrowing at the aortic arch, the
carina, left atrium, or the esophagogastric junction. In this case, the rupture site was a longitudinal laceration of the lower esophagus and esophagogastric junction. Mediastinitis or sepsis often occurs as a result of the anatomical position of the rupture site. Thus, an esophageal rupture is often associated with high morbidity and mortality rate. Clinical suspicion and an early diagnostic procedure for Boerhaave syndrome are important because of high mortality rate due to a delayed diagnosis from atypical symptoms and the anatomical position of the rupture site. A late diagnosis, of more than 24 hours after perforation, is associated with increased morbidity and mortality rates. However, an early diagnosis is often difficult and requires high suspicion, particularly in patients with an atypical presentation.

The classic clinical trial in Boerhaave syndrome consists of forceful vomiting, chest pain, and subcutaneous emphysema. Pain is the most common symptom and is usually localized to the perforation site. Emesis was documented in fewer than 80% of cases in a large series. In this case, we did not know whether he was suffering from emesis, because he was referred to our hospital with a mechanical ventilator and could not explain himself properly. This case shows the importance of information about a patient’s medical history. Recognition of previous respiratory or cardiac diseases could help identify Boerhaave syndrome from sudden respiratory arrest and abrupt onset of massive pleural effusion. Above all, when Boerhaave syndrome is suspected, an esophagography with a water-soluble contrast agent such as Ultravist can confirm the diagnosis and define the perforation site. In this case, the perforation site was confirmed by both esophagography and enhanced chest CT. Because of the location of the perforation site, rapid spread of fluid into the mediastinal and pleural cavity due to negative intrathoracic pressures lead to mediastinitis, pleural empyema, and eventually death. Outcome certainly depends on the extent and location of the perforation, but the single most important factor remains treatment timing. Management of a spontaneous esophageal perforation is controversial, and early surgical intervention is preferred in most cases of a thoracic esophageal rupture.

Based on this case, we recommend that if an abrupt onset of pleural effusion with no history or cause occurs, it is necessary to suspect an esophageal rupture. The pleural amylase level should be checked, and esophagography should be performed.

We report the case of a 48-year-old man with chronic alcoholism, accompanied by an abrupt onset of massive pleural effusion with no medical history who was diagnosed with Boerhaave syndrome and an esophago-pericardial fistula by pleural amylase level and esophagography with a Levin tube.

REFERENCES