

A Potential Pitfall in the Differential Diagnosis of Epigastric Pain: Boerhaave's Syndrome

Epigastrik Ağrının Ayırıcı Tamsında Potansiyel Bir Tuzak: Boerhaave Sendromu

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Abstract

Spontaneous esophageal perforation (Boerhaave's syndrome) is a very rare, life-threatening surgical emergency that should be suspected in all patients presenting with lower thoracic-epigastric pain and a combination of gastrointestinal and respiratory symptoms. A 68-years-old male presented with sudden severe atypical epigastric pain that was difficult to control. A contrast-enhanced CT scan of the chest showed bilateral pleural effusion, left pneumothorax and pneumomediastinum. Boerhaave's syndrome was diagnosed but during this time, he was unfortunately critically unwell and continued to deteriorate. He was subsequently underwent surgery but the patient died after surgery.

Keywords: Boerhaave's Syndrome, Contrast-enhanced Computerized Tomography, Esophageal Rupture

Öz

Spontan özofagus perforasyonu (Boerhaave sendromu), alt torasik-epigastrik ağrı ile gastrointestinal ve solunumsal semptomların birlikteliği ile başvuran tüm hastalarda şüphelenilmesi gereken, çok nadir, hayatı tehdit edici cerrahi bir acildir. Olgumuz, ani başlangıçlı, şiddetli ve medikal tedaviye yanıt vermeyen epigastrik ağrı ile başvuran 68 yaşında erkek hastadır. Toraksın kontrastlı tomografisinde bilateral plevral effüzyon, ve solda pnömotoraks ve pnömediastinum saptandı. Boerhaave Sendromu tanısı konan olgu, durumunun kötüleşmesi üzerine operasyona alınmasına rağmen ölümle sonuçlandı.

Anahtar Kelimeler: Boerhaave Sendromu, Kontrastlı Bilgisayarlı Özofagus Ruptürü, Tomografi

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Introduction

Perforated esophagus is the most serious, and frequently the most rapidly lethal, surgical emergency of the gastro-intestinal tract. Treatment delays and misdiagnosis of the condition contribute significantly to higher rates of morbidity and mortality (1). Diagnosis can be made earlier and more accurate with additional radiological examinations such as Computerized Tomography (CT) scan. Its sensitivity is estimated to be 92–100% (2).

Boerhaave's syndrome (BS) is first described by Dutch and Herman Boerhaave in 1724 as a very rare surgical emergency, usually diagnosed in men aged 50-70 years after heavy meal ingestion combined with abundant alcohol consumption (3). Typically, a vertical full-thickness rupture occurs on the left posterior aspect of the distal esophagus, 2-3 cm proximally to the gastroesophageal junction. The pathogenesis involves a sudden pressure increase caused by forceful vomiting against a closed glottis due to incomplete cricopharyngeal relaxation. If they are not treated, cases might rapidly progress to infectious mediastinitis and septic shock within 24-48 hours (3). BS is very rare (15% of all cases of esophageal perforation) and is commonly seen in patients aged 40-60 years old (2). Diagnostic errors are frequent, and condition can be confused with acute myocardial infarction, perforated peptic ulcers,

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and acute pancreatitis (4). If obtaining a contrast esophagogram is not possible, a contrast-enhanced CT scan of the chest should be obtained. Diagnostic thoracentesis of pleural effusion shows a presence of food particles, pH <6, or an elevated pleural fluid amylase. Even with prompt therapy and advances in surgical technique, the mortality rate can be very high, varying from 5-75%; higher rates correlate with delays in both presentation and diagnosis.

Case

A 68-years-old man presented to the emergency department with a severe epigastric pain and vomiting, preceded by a large meal. His medical history was unremarkable. He specifically denied any history of gastrointestinal disease, alcohol abuse, or cigarette smoking. On admission the patient was severely painful, blood pressure 90/50 mmHg, temperature 36.0°C, heart rate 129 beats/min, and SpO₂ 90% while breathing room air. Physical examination revealed diminished breath sounds and generalized rhonchus in both lung fields. Heart sounds were normal. Abdominal examination showed slight epigastric tenderness and normal bowel sounds. Intravenous fluids and analgesics were given to control pain. Electrocardiogram revealed atrial fibrillation. Complete blood count revealed leukocytosis of 20.000 and serum chemistry was normal. Plain radiograph of chest and abdomen was nonspecific for any disease (Figure 1). The patient didn't relieve despite proper analgesic therapy.

A contrast-enhanced CT scan of the abdomen was taken to exclude perforation, pancreatitis and

other abdominal pathologies. Normal pancreatic and liver tests made acute pancreatitis and cholecystitis unlikely. A normal serum chemistry and abdomen CT made an abdominal source of symptoms unlikely. An initial diagnosis of peptic ulcer was made but his condition deteriorated and he desaturated through the night. The patient was dyspneic and tachypneic. We administered oxygen via face mask to maintain SpO₂: 90%.

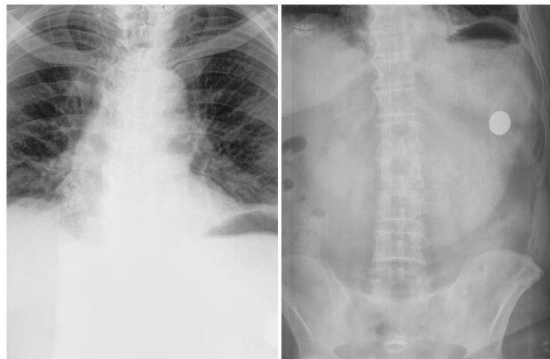


Figure 1. Plain radiograph of chest and abdomen

A contrast-enhanced CT scan of the chest was taken and CT confirmed left sided pneumothorax, revealed the presence of pleural effusion and pneumomediastinum (Figure 2), and allowed us to exclude other thoracic causes of pain, dyspnea, and respiratory distress (eg, aortic dissection, pulmonary embolism). We decided to drain the left pleural space, and immediately after chest-tube insertion we retrieved air bubbles and 1200 cc pleural fluid containing food particles. We placed a nasogastric tube to clear gastric contents and limit further contamination. BS was diagnosed. We applied suction to the chest tube and started intravenous antibiotics. At this stage the patient was critically unwell and continued to deteriorate. He was intubated and subsequently underwent surgery but the patient died after surgery.

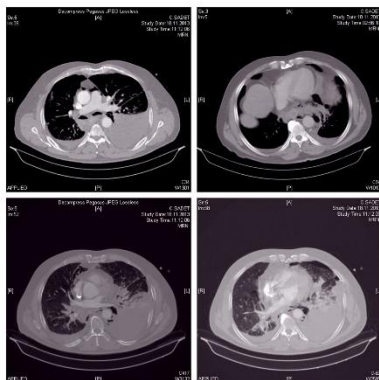


Figure 2. Contrast-enhanced CT scan of the chest

Discussion

If it is not thought in the differential diagnosis, BS is often unsuspected or misdiagnosed. Its manifestations closely mimic more common intrathoracic diseases such as myocardial infarction,

pericarditis, acute aortic disease or abdominal emergencies such as perforated peptic ulcer or acute pancreatitis. Unfortunately, diagnostic work-up delays may prevent treatment on time and can cause a negative effect on patients' outcomes (3).

Plain films of the thorax and abdomen are the initial imaging tests in emergency departments to investigate abdominal pain and vomiting. Unfortunately, they have a limited diagnostic value. The usual although unspecific radiographic features include mostly left-sided pleural effusion, lung infiltrates and atelectasis, whereas more specific signs such as pneumothorax, pneumomediastinum and pneumopericardium are rarely detected and not obvious (3).

The "classic" clinical presentation of BS includes episodes of vomiting (often preceded by excessive food and alcohol intake), followed by a sudden onset of upper abdominal and lower chest pain (4). Vomiting, lower thoracic pain, and subcutaneous emphysema (found in 14–30% of patients) are known as the Mackler triad, which is very suggestive of BS (2). Up to one third of all patients have an atypical presentation (2). The differential diagnosis of BS includes various thoracic and abdominal conditions. Diagnosis of BS can be difficult because often no classic symptoms are present and delays in presentation for medical care are common (4).

Blood laboratory tests are rarely beneficial in diagnosis. Normal pancreatic and liver tests may help to exclude acute pancreatitis and cholecystitis. Normal troponin activity make acute myocardial infarction unlikely. In patients with pleural effusion a thoracentesis is more informative than other laboratory tests. Macroscopic assessment often reveal undigested food particles, that confirms gastrointestinal-tract perforation (5). The chemistry of the pleural fluid usually shows low pH and high amylase level.

Our case highlights the difficulty of diagnosing BS due to the rarity of the condition. This leads to a delay in recognizing the pathology and treatment. Epigastric pain is one of the most common causes of admission to emergency departments. So we must think BS in epigastric pain with a differential diagnosis. The CT scan was the diagnostic tool that helped us to make the correct diagnosis. In addition, as well documented in literature data, our case highlights the importance of CT for diagnosing spontaneous esophageal perforation.

Informed Consent: Written informed consent was obtained from patient who participated in this case (18.11.2013).

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