Kardiologia Polska
2010; 68, 9: 1040–1042

ISSN 0022–9032

Boerhaave's syndrome — a difficult differential diagnosis of chest pain

Zespół Boerhaavego — trudna diagnostyka różnicowa bólu w klatce piersiowej

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Abstract

Boerhaave's syndrome is a very rare disease characterised by a spontaneous rupture of the oesophagus. It is often misdiagnosed and there is no consensus as to the best treatment. We describe a case of a 61 year-old man without significant previous medical history presenting in the emergency room with acute chest and back pain. Despite objective and laboratory tests negative for chest pain screening, computed tomography showed the presence of mediastinal air and pneumothorax consistent with oesophagus rupture. Urgent surgical intervention saved the patient.

Key words: Boerhaave's syndrome, computed tomography

Kardiol Pol 2010; 68, 9: 1040–1042

INTRODUCTION

Boerhaave's syndrome is a rare condition causing spontaneous oesophageal perforation, and has a high mortality rate. It was first described by Dutch physician Herman Boerhaave in 1724. The classical presentation is pain, dyspnoea and shock, followed by forceful vomiting [1]. The rupture is usually (in 90% of cases) in the lower third of the oesophagus and in the left lateral position. The mortality rate is high (20–40%) if untreated [1]. The available data are conflicting, in that they show favourable results for conservative, endoscopic as well as operative treatments. Identifying the best treatment has proved impossible.

CASE REPORT

A 61 year-old Caucasian man was assessed in the emergency room for acute chest and back pain. No significant medical history was detected; the patient took only tranquillizer domiciliary drugs. Baseline clinical examinations, ECG, chest radiogram and abdominal X-ray were normal. Laboratory tests (blood count, hepatic and kidney function, D-dimer and troponin I, blood gas analysis) excluded cardiogenic and pulmonary causes for the thoracic pain. Abdominal ultrasonography was also normal.

Because of the persistent acute chest-epigastric pain, chest and abdominal computed tomography (CT) were performed with, and without contrast. These showed pneumomediastinum and pleural effusion with subcutaneous emphysema due to an oesophagus leak localised in the highest third of the oesophagus (Figs. 1, 2).

The patient underwent urgent surgical repair of the oesophagus perforation, along with the cardias, anterior patch sec, and trans diaphragmatic mediastinal debridement and drainage.

The post-operative course was complicated by sepsis and acute respiratory distress syndrome, which were treated with antibiotic therapy and mechanical ventilation. At 3-month follow-up, the patient is asymptomatic and in a good condition.

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Received: 01.12.2009 **Accepted:** 16.12.2009

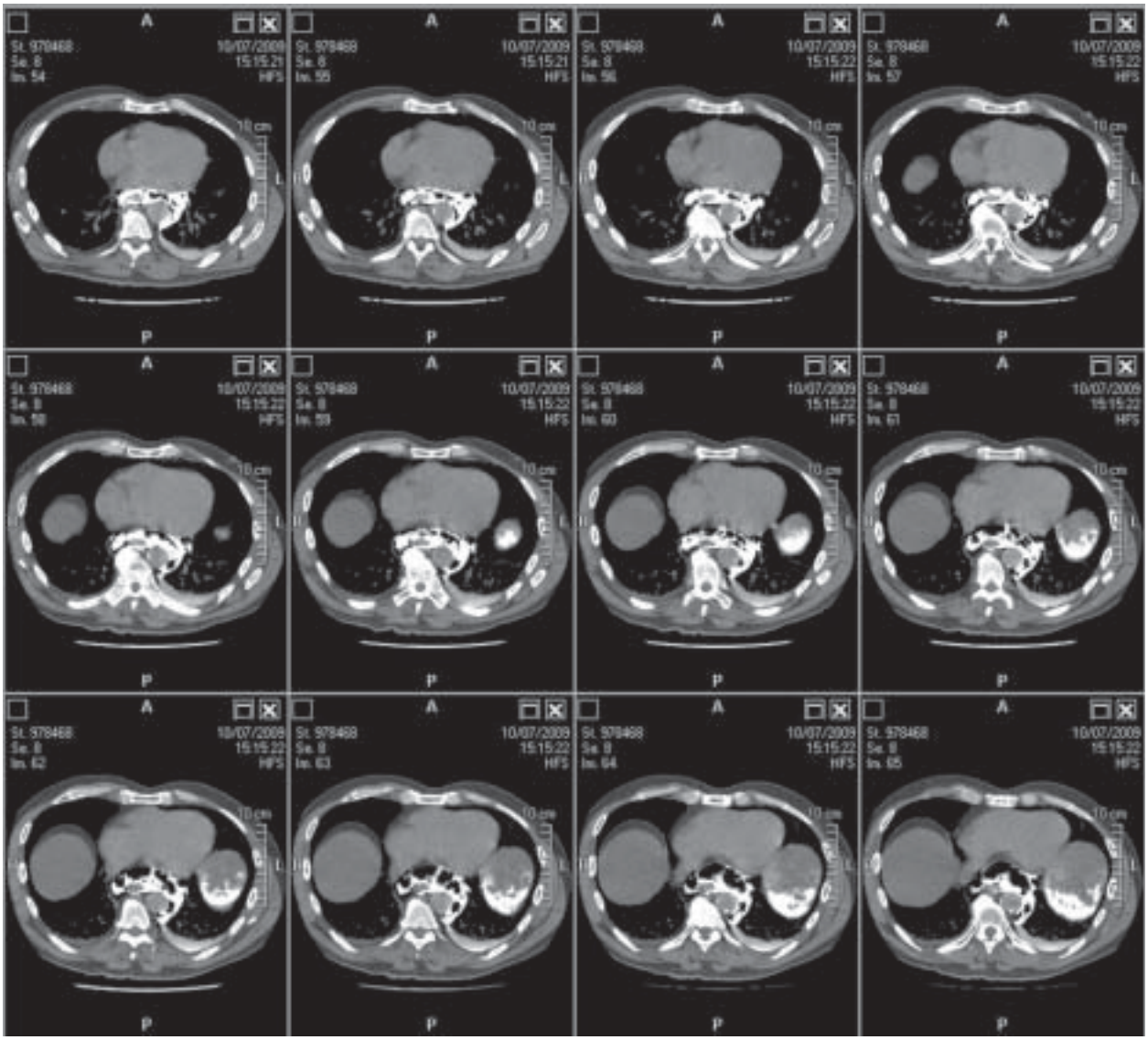

Figure 1. Thorax and abdomen CT with contrast shows pneumomediastinum and pleural effusion with subcutaneous emphysema

DISCUSSION

Due to the rarity of the condition, Boerhaave's syndrome often presents a diagnostic challenge and delayed diagnosis. The commonest misdiagnoses are: perforated ulcer, myocardial infarction, pulmonary embolism, dissecting aneurysm and pancreatitis [2].

The mortality rate is 20–40% and the factors that influence the prognosis are the time interval between onset of injury and primary repair, as well as the underlying physical status of the patient [2]. The rupture is usually (in 90% of cases) localised in the lowest third of the oesophagus and in the left lateral position [3]. Diagnosis can be made using a chest X-ray (although often the result will be normal), contrast oesophagogram and CT [1, 2, 4].

No consensus exists as to the most suitable treatment, but there are four aims in management strategy: 1. Direct repair if possible. If not possible, functional or surgical isolation of the oesophagus from the stomach. 2. Adequate drainage. 3. Appropriate antibiotics. 4. Adequate feeding [2].

Primary repair is usually advocated and seems to be the best treatment, if presentation is not delayed longer than 24 hours; the longer the delay, the more the tissue necrosis and oedema are present, perhaps preventing successful repair [5–7]. Jougon et al. [8] showed, however, that even a long interval before treatment does not preclude primary oesophageal repair. A conservative strategy is feasible when the perforation has existed for at least five days, there are no signs of severe sepsis, an oesophageal barium study shows a wide-mouthed cavity draining freely back into the oesophagus, and the pleural space is not contaminated. Endoscopic treatment is often advocated but should only be used where there is no sepsis and in cases with very early diagnosis [1].

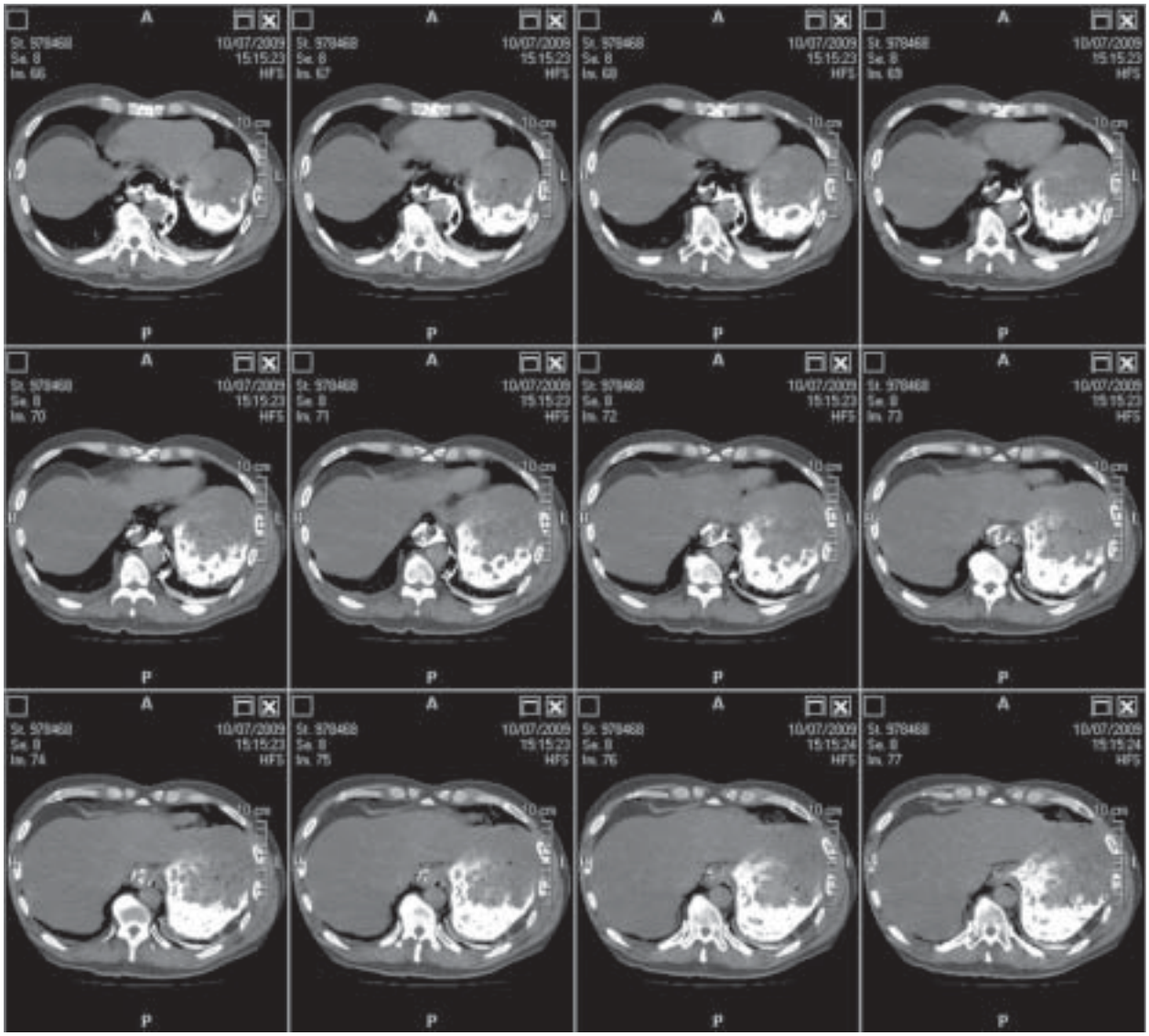

Figure 2. Continued from Figure 1

Our case underlines the difficulty of diagnosing Boerhaave's syndrome due to the rarity of the condition and the non-specificity of its clinical manifestation. This led to a delay in recognising the pathology and treatment. We have to take into account the possibility of meeting in a first aid situation such a patient, and so we must include Boerhaave's syndrome in chest pain differential diagnosis. The CT scan was the diagnostic tool that helped us to make the correct diagnosis. Furthermore, as well documented in literature data, our case stresses the importance and accessability of CT for diagnosing spontaneous oesophageal perforation thanks to the relatively fast visualisation and high quality of the images.

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