Boerhaave's syndrome : successful conservative treatment in two patients

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Abstract

The Boerhaave syndrome is a spontaneous, post-emetic rupture of the esophagus and a rare but potentially fatal cause of upper gastrointestinal bleeding. There are currently no guidelines on the optimal treatment of these patients, although there is a strong tendency towards a surgical approach. We present 2 cases of male patients, 66- and 77-year old respectively, both admitted to the emergency department with hematemesis. Unexpectedly, these turned out to be caused by the Boerhaave syndrome. Based on the severity of presentation, either a conservative or endoscopic treatment was adopted, both with good outcome. (Acta gastroenterol. belg., **2020**, 83, **654-656**).

Key words : Boerhaave's syndrome, conservative, endoscopy.

Introduction

A specific type of spontaneous rupture of the esophagus, the Boerhaave syndrome, was first described in 1724 by the Dutch physician Hermann Boerhaave (1,2). It describes the rupture of the distal esophagus caused by a rapid increase in intra-abdominal pressure, as seen in heavy vomiting or retching (1). Boerhaave's syndrome is rarely encountered and symptoms are aspecific (3). Consequently, diagnosis is often delayed, which leads to an increased risk of complications and mortality. The syndrome can cause severe septic shock, (multiple) organ failure (MOF) and is associated with high mortality rates (20-40%). If left untreated, it may have a fatal outcome (1,2,4,5). Therefore, a high index of suspicion is warranted in patients presenting with pain, fever or upper gastrointestinal bleeding after forceful vomiting. There is no consensus on optimal treatment, although the majority of authors do suggest a surgical approach. Only in selected cases a conservative treatment is possible (2). We present two cases of Boerhaave's syndrome. In the first case a patient was treated conservatively, with a good clinical outcome. Given the more severe presentation in our second case, this patient was treated endoscopically with stent placing.

Case 1

A 66-year-old man presented at the emergency department with vomiting and acute epigastric pain. After a normal lunch he had two episodes of vomiting with significant amounts of bright red blood and a feeling of dizziness. He had no other complaints, however there was a longer existing history of dysphagia for meat, though never associated with hematemesis. There was no recent use of non-steroidal anti-inflammatory (NSAID), anti-platelet or anticoagulant drugs. Vital signs were as follows : blood pressure 97/72 mm Hg, pulse rate 103 beats/min and body temperature of 37.8°C. Abdominal examination showed epigastric pain on palpation, without guarding or rebound tenderness. Further clinical examination was unremarkable. Laboratory evaluation revealed an elevated white blood cell count (with a left shift), normal CRP and discrete liver function abnormalities. Abdominal ultrasound was normal and showed no signs of free fluid. After intravenous (IV) fluids and proton pump inhibitors (PPI's), an urgent

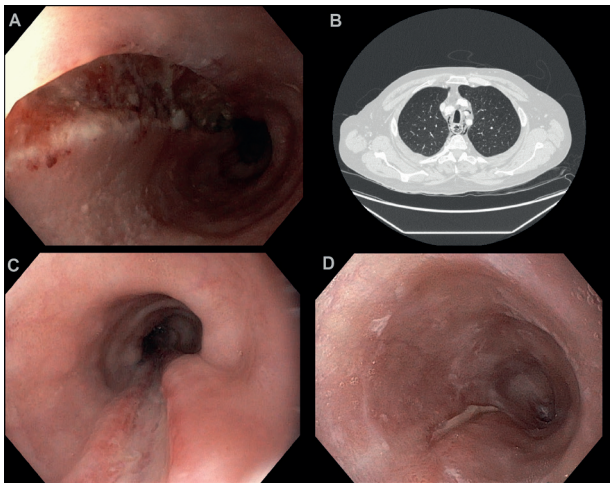

Figure 1. — A. Gastroscopy on admission demonstrated an esophageal laceration over 10 cm. B. CT scan on admission showed mediastinal emphysema surrounding the esophagus. C. Gastroscopy after five days demonstrated the partially healed laceration. D. Gastroscopy after 6 weeks showed almost complete healing of the laceration.

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gastroscopy was performed. This revealed an impressive laceration of the esophagus over a distance of 10 cm (Figure 1A).

Since there was no active bleeding, a conservative strategy was elected without endoscopic therapy. To exclude a full thickness perforation, a complementary CT scan was carried out. This showed the presence of mediastinal emphysema and a small amount of free fluid in the lower mediastinum, without significant pleural fluid or mediastinal abscess formation (Figure 1B). This implicates that a transmural perforation was present.

Treatment consisted of IV fluids, broad spectrum antibiotics Meropenem, Amikacin and antifungal therapy Fluconazole, fasting, anti-emetics and IV PPI's. Initially a hemodynamic impact was seen, but during treatment vital signs improved and the patient remained hemodynamically stable. Throughout the hospitalization an increase of the inflammatory markers was seen : CRP increased up to 166 mg/L (normal : < 5 mg/L), however the patient remained stable and did not develop any fever. The inflammatory markers declined gradually, there was no evolution to mediastinitis nor septic shock. Oral fluids and nutrition were slowly reintroduced with success.

Gastroscopy was repeated after 5 days, which showed the prominent laceration, with signs of partial healing (Figure 1C). Oral antibiotics were continued and our patient was discharged after six days.

A control gastroscopy after six weeks showed adequate healing of the laceration with presence of residual ulcer (Figure 1D). No other esophageal abnormalities were detected. With the longer existing history of dysphagia for meat an additional esophageal manometry was performed, without evidence for underlying motility abnormalities.

Case 2

A 77 year old male patient was seen in the emergency department because of hematemesis. He had a history of abdominoperineal resection of the rectum (APR) because of a locally advanced rectum carcinoma 17 years before. Furthermore he was a diabetic type 2 patient, suffered from Parkinson's disease and had a history of permanent atrial fibrillation for which he was on a direct oral anticoagulant rivaroxaban.

He now presented after vomiting multiple times, of which the first episode was associated with hematemesis. Afterwards he began to experience right thoracic pain, more with movement and with coughing. His colostoma output had not changed in the last few days. Physical examination showed normal blood pressure (139/76 mm Hg), a slightly elevated and irregular pulse (103 bpm), no fever (temperature 36.4°C) and normal oxygen saturation levels. His abdomen was not distended and on abdominal palpation he showed no tenderness or signs of peritonitis. There was no pain on palpation of the right hemithorax, nor was there subcutaneous emphysema palpable.

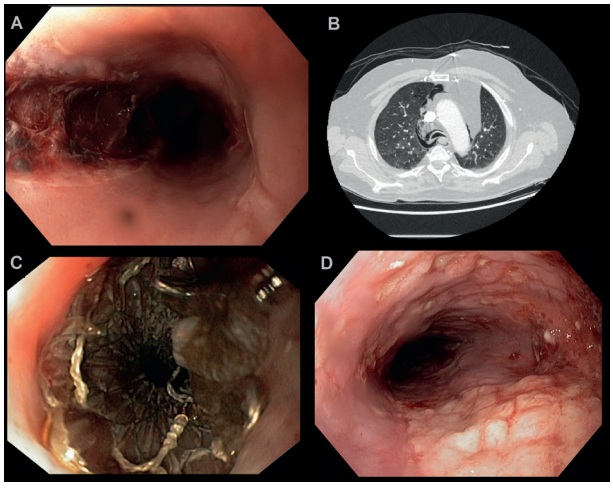

Figure 2. — A. Multiple lacerations in the esophagus during endoscopy on admission. B. CT scan on admission : pneumomediastinum. C. An endoscopic esophageal stent was placed. D. Upper endoscopy after stent removal, a pressure ulcer in the distal esophagus was seen.

Laboratory results showed an elevated white blood cell count with left shift, with normal CRP and hemoglobin level. Urgent upper endoscopy revealed multiple lacerations of the esophagus (Figure 2A), necrotic patches at the distal esophagus with adherent blood clots and active bleeding from two sites in the distal esophagus, which appeared as small fistulas.

After endoscopic injection of diluted adrenaline at the active bleeding sites, the patient underwent an urgent chest and mediastinum CT, which showed a pneumomediastinum, compatible with a full thickness perforation of the esophagus (Figure 2B). There was no free abdominal air, nor the presence of any fistulas or abscess collections. Because of the extent of the esophageal injuries, a fully covered (metallic) esophageal stent was placed endoscopically, on top of classic treatment consisting of broad spectrum antibiotics piperacillin-tazobactam and IV fluids, as well as a fasting regimen and PPI's (Figure 2C). He remained hemodynamically stable. Chest X-ray with gastrografin 3 days after stent placing showed no leakage of contrast, after which oral fluids and nutrition were gradually increased. The patient was discharged after 7 days of hospitalization.

Follow-up gastroscopy after 2 weeks showed good positioning of the stent (minor proximal migration) with healing of the distal esophagus. Because of local irritation and painful regurgitation the stent was extracted 3 weeks after initial placement. Underlying mucosa was bleeding slightly after removal and also showed a pressure ulcer in the distal esophagus (Figure 2D).

Discussion

Early diagnosis of Boerhaave's syndrome is essential for optimal treatment. The classic Mackler triad consists of vomiting, lower chest pain and subcutaneous emphy-

sema, which is present in 50% of cases, although it is not always recognized. Other symptoms can vary from vomiting, fever, coughing, dysphagia, chest or epigastric pain to (mild) hematemesis (1,3,4,6,7). In our patients the initial clinical suspicion was rather low because of the apparent mild symptoms and signs : there were only a few episodes of vomiting with minor hematemesis. This stresses the need for a high clinical index of suspicion, even in cases with an unlikely presentation. The post-emetic rupture is usually situated in the lower third of the esophagus, in the left lateral position, probably because of an anatomic weakness at this point. The length is on average 2.2 cm (1,6,7). In Boerhaave's syndrome gastric and oral secretions can leak into the mediastinum or the pleural cavities, with the possibility to cause local inflammation and a high risk of progression to sepsis (1,2,4). Diagnosis is made based on the history, clinical examination and chest X-ray, which can show signs such as subcutaneous emphysema, pneumomediastinum or mediastinal air-fluid levels. If the diagnosis is suspected, an additional contrast esophagography can be performed. Currently, CT is being advocated as a useful diagnostic tool to evaluate the extensiveness. In both our cases however, an upper endoscopy was the first exam performed. After visualization of the esophageal tears, endoscopy was quickly terminated to prevent progression of the rupture because of carbon dioxide insufflation. Further diagnostic workup included an additional contrast-enhanced CT in both cases (4,8,9).

The initial phase of therapy in Boerhaave's syndrome is based on resuscitation, cessation of oral intake and initiation of PPI's, to limit mediastinal contamination. Broad-spectrum antibiotic therapy is recommended, covering both aerobic and anaerobic bacteria to avoid polymicrobial contamination of the mediastinum (2,5,10). There are no uniform guidelines on the further treatment of Boerhaave's syndrome and the majority of cases are treated surgically. A more novel treatment strategy is endoscopic stenting, which shows promising results (11). The literature on conservative management is scarce, it can only be sufficient in highly selected cases (1,5), for which the following criteria are proposed : minimal symptoms, a well-contained rupture, no pleural contamination nor systemic inflammation. In practice those criteria can usually only be established in patients with a relatively late diagnosis (after 48 hours). If there are no negative prognostic factors at that time, conservative treatment is a valid option (1,2,6,7,12,13).

In both cases, the lacerations were quite extensive and accompanied with mediastinal emphysema, however the pleural cavities were not involved and there was no evolution to septic shock. Therefore, a non-surgical strategy was adopted in both patients, with endoscopic stenting in the second case.

After healing of the lacerations, a work-up for esophageal abnormalities can be considered to exclude an underlying esophageal disorder. In both cases, the follow-up gastroscopy showed good healing, an additional manometry to exclude motility disorders was also normal in the first patient (13). In the second case, no additional manometry was performed.

Conclusion

The Boerhaave syndrome is a rare but life-threatening condition, which is generally treated surgically. However, in highly selected cases a conservative (medical – endoscopic) approach can be considered.

Conflict of interest

The authors of this paper report no conflicts of interest.

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