Management of Boerhaave’s Syndrome: Report of Three Cases

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Abstract

From 2000 to 2005, three patients with Boerhaave’s syndrome were successfully managed in our Department. Two of them received the appropriate treatment belatedly, with primary closure and bolstering tissue wrap. One of them required further intervention with a cervical esophagostomy and exclusion of the perforated esophagus. The third patient with an esophageal perforation related disorder, was managed with surgical exploration and drainage alone. Primary suturing of the esophagus should be performed only in patients with an early perforation. In cases of prolonged delay between rupture and diagnosis, esophageal resection with cervical esophagostomy and gastrostomy is advocated as the safest therapy.

Key words

Primary esophageal perforation - Boerhaave’s syndrome - current treatment

Introduction

Boerhaave’s syndrome represents the most sinister cause of esophageal perforation [1]. It remains a potentially lethal complication, and is still associated with a high mortality rate [1-6]. Classical presentation of this syndrome is vomiting, chest pain, and subcutaneous emphysema. However, this triad, first described by Mackler [3] is seldom found. Delay in diagnosis is common, resulting in substantial mortality. Some authors report mortality figures more than 65% after 24 hours and 75-89% after 48 hours [1,2,4].

The general consensus, concerning the management of this syndrome, is early diagnosis and timely inhibition of the fatal esophageal injury-induced inflammatory process[1,7-10]. Nevertheless, the optimal therapeutic approach remains controversial. Indeed, divergent therapeutic approaches have been reported with significant variance in outcome and complications, with mortality rates ranging from 3% to more than 50% [1]. The aim of this study is to present our experience on diagnosis and management of spontaneous esophageal rupture and assess the current management status according to recent literature data.

Case report

From 2000 to 2005, three consecutive spontaneous esophageal perforations were managed in our Department. All were men, aged between 32 and 47 years. Clinical presentation, radiographic findings, localization of rupture, degree of contamination in the thorax, days from admission till operation, treatment method, complications, the period of admission and ICU stay are listed in Table I.

Patient 1

A 42-year-old man, with a history of organic psychosis and pyloric channel stricture secondary to chronic duodenal ulcer, was transferred to our Department after a spontaneous esophageal rupture that had been diagnosed elsewhere. The original diagnosis was delayed because mid epigastric and lower thoracic pain after forcible coffee-ground emesis was misinterpreted as an upper gastrointestinal (GI) tract hemorrhage. On the fifth day of hospitalisation, after a hemorrhage relapse along with hemodynamic instability and systemic septic clinical signs, the patient underwent a complementary evaluation, with upright plain x-ray, contrast enhanced CT scan and esophagogram, revealing a right sided, distal esophageal rupture, with ipsilateral hydro pneumothorax and pleural empyema (Fig.1). A chest tube was placed for empyema drainage and the patient was transferred to our Hospital. Resuscitation required intravenous fluids, packed red blood cells, and antibiotic coverage, but despite this his septic profile failed to improve and was further aggravated by concomitant GI bleeding. The patient underwent emergency operation, where ligation of
<table>
<thead>
<tr>
<th>Pt</th>
<th>Sex/Age</th>
<th>Chief complaints</th>
<th>Time from onset till surgery</th>
<th>Imaging studies</th>
<th>Esophagography</th>
<th>Location of rupture</th>
<th>Surgical procedure</th>
<th>Subsequent procedures</th>
<th>ICU stay</th>
<th>Time of discharge</th>
</tr>
</thead>
<tbody>
<tr>
<td>1M 42</td>
<td>Postemetic, mid-epigastric, lower thoracic pain, dyspnea</td>
<td>5 days</td>
<td>Right hydrothorax</td>
<td>Right pleural empyema</td>
<td>Leakage into right pleural cavity</td>
<td>Right side wall of LTE</td>
<td>Right thoracotomy-drainage, laparotomy-bleeding duodenal ulcer oversewing-pyloroplasty</td>
<td>Esophageal SECMS</td>
<td>10 days</td>
<td>15 weeks</td>
</tr>
<tr>
<td>2M 32</td>
<td>Postemetic lower thoracic pain, dyspnea, subcutaneous emphysema</td>
<td>&gt;25 hours</td>
<td>Bilateral pleural effusion</td>
<td>Bilateral pleural effusion</td>
<td>Leakage into left pleural cavity</td>
<td>Left side wall of LTE</td>
<td>Bilateral thoracotomies-drainage, primary closure</td>
<td>Cervical esophagostomy, esophageal exclusion, gastrostomy, feeding jejunostomy, continuous pleural irrigation, Delayed esophageal reconstruction with left colic flexure</td>
<td>35 days</td>
<td>9 weeks</td>
</tr>
<tr>
<td>3M 47</td>
<td>Postemetic mid-epigastric and back pain, dyspnea</td>
<td>12 hours</td>
<td>Pneumomediastinum</td>
<td>Left pleural effusion</td>
<td>Leakage into left pleural cavity</td>
<td>Left side wall of LTE</td>
<td>Left thoracotomy-drainage, laparotomy-primary closure+foundoplication</td>
<td>None</td>
<td>2 days</td>
<td>5 weeks</td>
</tr>
</tbody>
</table>

M=Male, LTE=Lower Thoracic Esophagus, SECMS=Self-Expandable Covered Metallic Stent, CT=Computed Tomography
Boerhaave's syndrome

A bleeding vessel in the duodenal ulceration, pyloroplasty, draining gastrostomy and feeding jejunostomy were fashioned via an upper midline laparotomy. Additionally, a right thoracotomy for better drainage and lavage of the right thoracic cavity was performed and the chest was closed with two large thoracostomy tubes in situ. A hypaque swallow study, on the 4th postoperative week demonstrated closure of the esophageal rent but revealed the development of a stricture along the lower third of the esophagus, which proved to be refractory to multiple endoscopic dilatations performed between the 9th and 11th postoperative weeks. The patient was considered to be unfit for esophageal replacement and in addition he was reluctant to undergo any major intervention. Hence, a covered self-expanding metallic stent (Ultraflex, Boston Scientific) was inserted on the 12th postoperative week, endoscopically. During the follow-up period he was able to eat normally and maintain his weight. He survived for 3 years and finally died of a severe depression disorder.

Patient 2
A 32-year-old man, with no history of previous illness but with an ambiguous reference to recent food and drink overindulgence, was presented to a peripheral Hospital Emergency department complaining of the sudden onset of gradually increasing, lower thorax post emetic pain and subcutaneous emphysema. The suspicion of spontaneous esophageal rupture was confirmed on a subsequent erect x-ray film, CT scan and esophagogram by the presentation of bilateral pleural effusion together with left sided contrast extravasation from the lower third of the esophagus (Fig.2).

Bilateral thoracotomies, closure of the esophageal perforation in two layers, buttress of the defect with viable regional pleural flap and placement of chest tubes were performed. Subsequently, the patient was transferred to our department because of lack of ICU. On postoperative day 8, the patient demonstrated clinical signs of a recurrent leak, with recrudescing systemic sepsis, dyspnea, and hemodynamic instability. The subsequent CT scan study demonstrated contrast medium extravasation accompanied with large pleural effusion bilaterally. Re-operation was considered inevitable, during which mediastinum and pleural debridement, forcible irrigation of pleural spaces, cervical esophagostomy in combination with closure of the distal esophagus, feeding jejunostomy and draining gastrostomy, were performed. Postoperatively, thoracotomies were left opened and continuous irrigation systems were applied to our patient's thoracic wounds [6]. The patient had a prolonged ICU stay, mainly because of the need for ventilation, as a result of severe pneumonitis, along with relapsing incidences of systemic sepsis. Eventually, he was returned to the main ward on the 5th postoperative week. After a total period of 14 weeks, the patient was discharged, continuing total enteral alimentation. Five months later, he underwent reconstruction by joining the cervical esophagus to the stomach using retrosternal left colic flexure based on the left branch of the middle colic vessels. He can now eat normally and maintain his weight and is leading a normal existence.

Patient 3
A 47-year-old man, with no history of previous illness, and with reference to a preceding overindulgence in a large meal, presented to our Department, complaining of
the sudden onset of a sharp, post emetic, mid epigastric pain, radiating to the left shoulder. On the basis of classic esophageal rupture history, the diagnosis was readily made by means of erect chest x-ray and CT (Fig.3). A confirmative subsequent hypaque swallow study demonstrated small extravasation of contrast medium from a distal esophageal perforation. After a short resuscitation period, the patient was transferred to the Operating Theatres, where the rupture site, located on the left esophageal wall next to the gastroesophageal junction, was approached with midline laparotomy. A primary esophageal closure in two layers with additional fundoplication was performed. Furthermore, a small left thoracotomy was performed and the thoracic cavity was copiously irrigated. Finally the chest was closed with a large (32-gauge) thoracostomy tube in situ. On the third postoperative day the patient was returned to the main ward where repeated esophageal contrast medium studies verified a successful outcome. On postoperative day 15 the patient began oral feeding. Five weeks after surgery, the patient was discharged in good general condition. At the time of writing, the patient continues to do well.

Discussion

The presentation of Boerhaave’s syndrome is usually non-specific and may mimic many other clinical disorders [11]. Our diagnostic tools were chest X-ray, CT, esophagography, and esophagoscopy, but principally a high index of suspicion is required for timely diagnosis. In our cases, CT scans were done immediately after oral contrast administration, to detect the level and size of perforation and define the surrounding tissue inflammation, which helped as in deciding on the most appropriate therapy [12,13].

The management of the syndrome remains controversial since treatment can be surgical or non-surgical, and indications vary according to the functional state of the esophagus, the presence of associated lesions and the habits of the different medical teams [1,9,14]. Today, it is accepted that the method of treatment plays an important role in the mortality rate and although surgery has been the most common approach, the selection criteria for conservative treatment reported by Altorjay et al [15] (intramural perforation, benign defects, and the absence of sepsis) in 1977, and Cameron et al [16] in 1979 (disruption contained in the mediastinum; the cavity draining back into the esophagus; minimal symptoms; and minimal signs of sepsis), are still valid and should be taken into account. With reference to the above, perforations and pleural contamination, once controlled by adequate drainage, simply become an esophagocutaneous fistula and will heal the same as most gastrointestinal fistulas [17].

Recently, some authors claimed that rapid closure of the esophageal leak and drainage, could also be achieved by the minimal invasive endoscopic approach by inserting an endoprosthesis, followed by interventional drainage and/or thoracoscopic irrigation of the contaminated thoracic cavity [18-20]. Nevertheless, we believe that this approach can be applied only for iatrogenic and early detected perforations.

A self-expandable covered metallic esophageal stent was placed in one of our patients in order to treat a distal esophageal post perforation stricture. Esophageal stenting for non-malignant strictures is controversial. The covered self-expanding metallic stent was originally used for the stricture or fistula caused by malignant diseases [18]. On the other hand, many authors investigated the application of this alternative method to the management of benign conditions [21]. In our case, stent placement was used mainly because of the patient’s refusal for further invasive procedure, as well as considering his moderate medical condition.

There is still controversy about the most appropriate type of surgery for patients with esophageal perforation. Some surgeons performed primary repair, regardless of the interval between the perforation and intervention, resulting in a diverse outcome [8,10]. Our opinion on primary repair is that it is better to be avoided in delayed patients. Conversely, if the inflammatory process is limited, primary repair is a reasonable option and may result in an excellent outcome [10]. Some others suggest that esophagectomy may be better from primary repair for patients with delayed perforation, because of the high risk of leakage [1,22]. We believe that primary reconstruction must be the first treatment option in stable, nonseptic patients. Debridement and drainage with or without continuous lavage [23, 24] is another option, especially if the patient’s general condition is impaired or progressive sepsis is apparent. We applied this method in one of our patients with satisfactory results. If the interval between injury and intervention exceeds 24h or CT shows signs of progressive periesophageal inflammation, reinforcement of the esophageal repair by viable regional tissue is recommended [11].

If delayed reconstruction is being considered, it is possible to bring up the stomach, the small intestine or the colon to join the cervical esophagus. The timing of reconstruction must be based on the patient’s condition or/and recovery. Reconstruction of the esophagus can be performed simultaneously if there is no severe systemic inflammatory response. Otherwise, delayed reconstruction (2-4 months) is possibly the best option [1]. The new esophagus can be
placed in the posterior mediastinum or in the retrosternal or subcutaneous positions. In patient 2 delayed esophageal reconstruction was performed with colonic graft positioned retrosternally in order to achieve a better cosmetic result.

All our patients were surgically treated after resuscitative measures such as fluids and antibiotics. Our intention was to prevent ongoing spoilage, to provide debridement of devitalized tissue and to perform wide drainage. Postoperatively, all patients received supportive management including nasogastric tube decompression of the stomach, broad-spectrum antibiotic administration, and nutrition.

In conclusion, without treatment, patients with esophageal rupture frequently die of mediastinitis. Because of the very small numbers of cases, no standard therapy has been established. Conservative treatment remains a controversial topic, although recent sporadic reports have documented the efficacy of nonoperative care, especially following perforations in nonseptic patients. Primary suturing of the esophagus should be performed only in patients with an early perforation. When ongoing mediastinitis and pleural contamination has occurred, only esophageal exclusion or resection can definitely eliminate the source of intrathoracic sepsis.

References