Boerhaave Syndrome—over 290 years of surgical experiences. Can the disorder recur?

Marek Rokicki, Wojciech Rokicki, Małgorzata Mój, Tamer Bsoul, Mateusz Rydel

Department and Clinic of Thoracic Surgery in Zabrze, Medical University of Silesia, Zabrze, Poland

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ABSTRACT: Spontaneous perforation of the esophagus (Boerhaave syndrome) is a rare disease that poses a serious surgical challenge. The analysis of literature concerning the Boerhaave syndrome revealed cases of recurrent spontaneous perforation of the esophagus. The incidental nature of this condition calls for more accurate assessment of all such cases. The authors made a detailed evaluation of the data obtained from eight reports of recurrent Boerhaave syndrome. The data is presented as a summary table comparing the clinical course and outcomes of patients with the primary Boerhaave syndrome as well as recurrent Boerhaave syndrome.

KEYWORDS: esophageal perforation, Boerhaave syndrome, recurrence

Spontaneous perforation of the esophagus, also referred to as Boerhaave syndrome (BS) consists in longitudinal rupture of previously unremarkable esophagus [1]. The most common cause for the spontaneous perforation is a sudden increase of pressure within the esophageal lumen due to disordinated esophageal motility during forceful vomiting. In the course of a literature review focused on spontaneous esophageal perforation [2, 3], we encountered some publications reporting recurrence of the disorder [4–11]. Four reports originated from the US [4, 5, 6, 7], three others originated from Japan [8, 10, 11], and one report originated from England [9]. We carried out a detailed clinical analysis of data from these reports which we are presenting in the overall table.

The age of patients at BS recurrence ranged from 17 to 74 years, with the mean of slightly above 50 years and similar to that calculated by Brauer et al. for primary BS (52.4 years). Moreover, the male to female patient ratio in the study material was 8:1, similar to literature reports [12]. When comparing time intervals between primary perforation and its recurrence, wide diversity is observed with the shortest interval amounting to 24 days as compared to as much as 30 years for the longest one. In most cases, however, BS recurrence was observed several months to several years after the first episode. In the reported cases of BS recurrence, the main symptom consisted in pain within the chest (six patients) or epigastrium (three patients). The second most common symptom was vomiting observed in 6 patients [6, 7, 9, 10, 11], with hematemesis observed in four of these patients [6, 7, 11].

Only one of the nine patients included in the reports [7] presented with Mackler triad of symptoms consisting of chest pain, vomiting (including hematemesis) and subcutaneous emphysema of neck and chest [13]. Of these three, subcutaneous emphysema is the most pathognomonic sign; it is diagnosed in about 60% of patients with primary BS [14]. The proposed explanation for the fact that it occurred in as few as just one patient in the reported study group consists in the presence of pulmonary-pleural and esophago-pleural adhesions following the primary intervention which might have efficiently blocked the development of pneumothorax and subcutaneous emphysema [10]. Three out of nine patients had a history of long-term, severe alcohol abuse [5, 7, 10]. Other, isolated complaints included swallowing difficulties [4] and respiratory insufficiency [5].

In all the presented cases, diagnostic procedures started with endoscopic examination of the esophagus; however, detection of radiopaque contrast administered by mouth leaking outside the esophageal lumen in chest radiography was decisive for the diagnosis of spontaneous perforation [4, 5, 6, 7, 8, 9, 11]. Only in one patient, the diagnosis was made on the basis of emergency endoscopic examination due to intense bleeding from the intestinal tract requiring surgical intervention [10].

The predominant location of primary perforation was the posterior wall of the lower levolateral segment of thoracic esophagus (five patients) [5, 6, 7, 9, 10]. In other two patients, lesions were observed contralaterally at the same level [9, 11]. In another two patients, perforation was located within the central segment of thoracic esophagus [4, 8]. The predominance of levolateral locations of the primary lesions is explained by its relative weakening due to the esophagus being angled in that location and its wall being penetrated by vessels and nerves [15]. With regard to the location of the disease recurrence, only two patients experienced the recurrence at the original location [9, 10]. In other five patients, rupture was located contralaterally [5, 7, 9] or next to the primary perforation [6, 8]. Ieta et al., suggested that this noticeable trend towards recurrent perforation being located on the contralateral side of the esophagus may be due to esophageal wall being strengthened by pleural adhesions and scar formed as a result of previous treatment [10]. In the case of primary BS, the lengths of the ruptures were in the range of 10–30 mm with the mean of 25 mm while the lengths of recurrent ruptures ranged from 5 to 60 mm with the range of 31 mm. As seen from these results, recurrent perforations were somewhat longer.

When planning the treatment strategies for both primary and recurrent BS, the authors of the studies [4–11] qualified their patients for individual modalities by taking into account the time between the actual perforation taking place and the countermeasures being taken, local lesions within the esophageal wall, mediastinal and pleural infection status and overall health of patients. With regard to primary BS treatment, surgical treatment
was slightly more frequent [5, 6, 7, 9, 10] than the conservative approach [4, 8, 9, 11]. Similar situation was observed for recurrent perforations. Only three patients were subjected to conservative treatment due to severe overall condition, mediastinum and pleural cavities being massively affected by purulent processes inflammation, and, in one case, small perforation size with no septic symptoms. Surgical procedures used in the treatment of spontaneously recurrent perforations included primary repair with [9] or without [5, 6, 10] suture lines being strengthened by means of surrounding tissue patches as well as temporary “excision” of esophageal passage [7]. One out of nine patients presented died after the surgical procedure while the remaining cases ended up with patient survival. The patient who passed away had been admitted to hospital in extremely severe condition and with signs of respiratory insufficiency while his esophageal perforation was the longest of all those observed. According to the author, the death was due to postsurgical complications [5].

**FINDINGS**

We are aware that our analysis was based on very scant literature data. The presented very rare cases of BS recurring following earlier treatment provide a positive answer to the question posed in the title of this report. In addition, just like Ieta et al. [10], we observed an outline of potential correlation between the primary treatment modality and the time of disease recurrence. Following conservative treatment, the time to recurrence was up to six months [4, 8] while surgical treatment resulted in recurrence-free periods ranging from eight months to 30 years [5, 6, 7, 9, 10, 11].

Reports of patients being followed up after the treatment of spontaneous esophageal perforation are rarely encountered, and they cover a maximum period of 2–5 years following the first repair. In these follow-up periods, esophageal widening due to the stenosis of lower thoracic esophagus was required in 4 out of the 43 patients [16–19]. As shown by these data and by the possibility of BS recurrence, patient follow-up should be extended to as long as several decades after the treatment [2, 3, 4].

Recently, a report was published suggesting that besides alcohol abuse, asymptomatic gastroesophageal reflux disease, or hiatal hernia, antiphospholipid antibody syndrome may be another risk factor of BS recurrence [11].

In summary, the presented literature review shows that the diagnosis of recurrent spontaneous esophageal perforation is difficult due to the incidental nature of the disorder and its symptoms being frequently inconsistent and non-specific. In addition, the disorder may recur after one month as well as after 30 years after previous treatment. What is reassuring, however, is that the presented treatment modalities turned out to be successful.

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**Tab. I.** Clinical analysis of data obtained from literature reports [4–11].

<table>
<thead>
<tr>
<th>AUTHOR / COUNTRY</th>
<th>AGE</th>
<th>SEX</th>
<th>LOCATION OF PRIMARY PERFORATION</th>
<th>PERFORATION LENGTH (MM)</th>
<th>TREATMENT</th>
<th>TIME OF RECURRENT PERFORATION</th>
<th>PERFORATION LENGTH (MM)</th>
<th>LOCATION OF RECURRENT PERFORATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Saha S.P. [4] U.S.A.</td>
<td>17</td>
<td>F</td>
<td>Central</td>
<td>Not reported</td>
<td>Conservative</td>
<td>6 months</td>
<td>Chest pain</td>
<td>Central</td>
</tr>
<tr>
<td>Kish G.F. [5] U.S.A.</td>
<td>51</td>
<td>M</td>
<td>Lower left</td>
<td>30</td>
<td>Surgical</td>
<td>14 months</td>
<td>Chest pain, respiratory insufficiency</td>
<td>Lower right</td>
</tr>
<tr>
<td>Reeder L.B. [6] U.S.A.</td>
<td>66</td>
<td>M</td>
<td>Lower left</td>
<td>Not reported</td>
<td>Surgical</td>
<td>30 years</td>
<td>Epigastric pain, hematemesis</td>
<td>Lower left</td>
</tr>
<tr>
<td>Lujan K.J. [7] U.S.A.</td>
<td>45</td>
<td>M</td>
<td>Lower left</td>
<td>10</td>
<td>Surgical</td>
<td>8 months</td>
<td>Mackler triad</td>
<td>Lower right</td>
</tr>
<tr>
<td>Nokata Y. [8] Japan</td>
<td>74</td>
<td>M</td>
<td>Central left</td>
<td>25</td>
<td>Conservative</td>
<td>24 days</td>
<td>Chest pain upon swallowing</td>
<td>Central right</td>
</tr>
<tr>
<td>Khan D.A. [9] England</td>
<td>49</td>
<td>M</td>
<td>Lower left</td>
<td>30</td>
<td>Surgical</td>
<td>26 months</td>
<td>Epigastric pain, hematemesis</td>
<td>Lower right</td>
</tr>
<tr>
<td>Ieta K. [10] Japan</td>
<td>43</td>
<td>M</td>
<td>Lower left</td>
<td>Not reported</td>
<td>Surgical</td>
<td>6 years</td>
<td>Stomach pain, hematemesis</td>
<td>Lower left</td>
</tr>
<tr>
<td>Naitoh H. [11] Japan</td>
<td>52</td>
<td>M</td>
<td>Lower right</td>
<td>50</td>
<td>Surgical</td>
<td>8 years</td>
<td>Chest pain, shock</td>
<td>Lower right</td>
</tr>
</tbody>
</table>

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**Note:** The table includes only patients with spontaneous esophageal perforation and follows up after surgical treatment. The data were analyzed for primary and recurrent perforation location, treatment, time to recurrence, and treatment results.
REFERENCES:


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Corresponding author: Prof. dr hab. n. med. Marek Rokicki; Department and Clinic of Thoracic Surgery in Zabrze, Medical University of Silesia, Zabrze, Poland; E-mail: marekrokicki47@poczta.onet.pl

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