Boerhaave syndrome. A case report

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Abstract: Background: Spontaneous rupture of the esophagus (Boerhaave’s syndrome) is a rare, well defined clinical syndrome caused by a longitudinal perforation of the esophagus. This syndrome was first described by Boerhaave in 1724. This severe disease causes high mortality rates and it is difficult to diagnose it not only because it is rare but also because it is frequently confused with other severe clinical conditions. Case presentation: In this report we present a unique case of Boerhaave’s syndrome in a 49-year-old male patient. In the period October 16-21 2009 he was hospitalized with the diagnosis: septic shock, rupture of esophagus, ARDS. The patient died on October 21, 2009 by cardiac arrest. Conclusion: The rupture of the esophagus is a serious condition and its diagnosis is usually neglected during the initial assessment.

Key Words: esophageal disease, spontaneous rupture of esophagus, postoperative death

Boerhaave Syndrome, first described in 1724, is a life threatening of non-iatrogenic rupture of the esophagus, and rupture usually occurs in the left postero-lateral wall of the lower third of the esophagus. Herman Boerhaave (1668-1738) first described esophageal rupture and the subsequent mediastinal sepsis based upon his careful clinical and autopsy findings and hundreds of references have since been written about Boerhaave’s Syndrome. Boerhaave’s first report was the case of Baron Jan Gerrit van Wassenaer Heer van Rosenberg (1672 - 1723), Prefect of Rhineland and Grand Admiral of the Dutch Fleet. This figure was a nobleman and war hero at the peak of the Dutch Golden Age who played his role in steering the course of European history. Wassenaer had severe left-sided chest pain after vomiting. He died 18 hours later and autopsy showed a tear in the distal esophagus, emphysema and food in the mediastinum. Boerhaave undertook the autopsy and first described the appearance of the corpse, noting the strange accumulation of fluid in the flanks and the spongy crepitation from air under the skin. When he next opened the thoracic cavity he immediately felt more air and detected the strong odor of duck. With deeper dissection, Boerhaave found a ragged communication between the pleural cavities. Then, at the bottom of the chest, he found his answer: a hole in the baron’s esophagus, just above his diaphragm. He could detect no sign of chronic ulceration or inflammation, but a fresh injury resulting in massive contamination of the pleural cavity by food, saliva, and stomach contents. Boerhaave had thus uncovered the first recorded case of an esophageal rupture due to forceful vomiting [1].

The term spontaneous perforation of the esophagus, although commonly used in an inappropriate term, as the rupture is rarely spontaneous and almost invariably follows barotrauma from a sudden post-emetic rise in esophageal pressure. Although, the original report of Boerhaave describes a transverse tear in the lower left esophagus, most tears are longitudinal varying in length from 0.5 to 20 cm, located on the left postero-lateral wall of the esophagus 2 to 6 cm above the diaphragm in 80% of cases [2]. Furthermore, presentations of esophageal perforation can be distinguished as acute, subacute, and chronic. Acute perforation

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Presents with symptoms within twenty-four hours after rupture. In a subacute rupture, symptoms develop between twenty-four hours to two weeks following perforation. With chronic perforation, the onset of symptoms is more insidious, often delaying presentation and diagnosis for weeks to months after rupture [3,4].

A history of forceful emesis, subxiphoid chest pain, and subcutaneous emphysema (termed the Mackler Triad) is common triad of symptoms that should suggest acute esophageal rupture [5,6]. However, atypical presentations in which esophageal rupture mimics pneumonia, myocardial infarction, or aortic aneurysm are sometimes seen as well. Chest films may reveal pneumomediastinum, unilateral effusion, pneumothorax, hydropneumothorax, subcutaneous emphysema, or mediastinal widening. Additionally, X-rays may show radiolucent streaks of air which dissect through fascial planes behind the heart in the shape of the letter “V”, the so-called “V-sign” of Nacleiro [7,8]. Other imaging techniques to consider in diagnosing esophageal rupture are barium esophagram and CT chest [9]. In both techniques, there will be evidence of extravasation of food particles or bile from the esophageal lumen into the pleural space or mediastinum [3]. Finally, endoscopy can identify the location of the esophageal defect and confirm the existence of extraluminal disease or to rule out the diagnosis altogether [10].

Boerhaave Syndrome produces an extremely severe clinical profile resulting from mediastinitis and sepsis that develop rapidly due to extravasation of digestive sections and food into the mediastinum and pleural space [11]. Diagnosis is often delayed, since it is a rare disease and is usually confused with other equally serious but more prevalent pathologies such as acute myocardial infarction, perforated peptic ulcer and acute pancreatitis [12]. These factors contribute to its high mortality rate, roughly 39%, despite current medical resources [13].

Case presentation

A 49-year-old male eas referred to the Emergency Clinical County Hospital Tg. Mureș (October 16, 2009) with the diagnosis: septic shock, pyopneumothorax with rupture of the esophagus, resuscitated cardiac arrest. During hospitalization he was treated in order to reestablish his hydro-electrolyte balance, he received intravenous antibiotics, inotropic medication, gastric protectors, corticosteroids for acute kidney insufficiency (probably of pre-renal type), ARDS (acute respiratory distress syndrome), coagulopathy. On October 17, 2009 a surgical intervention was performed consisting of left posterior-lateral thoracotomy, lavage, suture the esophageal perforation, left pleural drainage. The patient dies on October 21, 2009 by cardiac arrest.

At the internal examination of the body we noticed multiple pathological lesions. Pleura – without shine, zonal yellowish purulent deposits bilaterally, without adherences. In the pleural cavities we found bilateral each 100 ml of purulent yellowish content with a bad smell. Trachea and bronchia had grayish-wet mucosa and in the lumen a small quantity of mucous-purulent content. The lungs fill the thoracic cavity, on the surface they are violet-grey, crepitations are absent bilaterally, mottled aspect on section surface, grey zones alternate with violet and reddish zones, after pressure air filled reddish blood pours out in big quantity while in the bronchioles in some areas whitish/yellowish purulent corks are visible. The esophagus has a hyperemic mucosa, longitudinal

Figure 1. Esophageal inflammation (Hpt.nr. 23443/2009; col. HE x 100)

Figure 2. Esophageal hyperaemia(Hpt.nr. 23442/2009; col. HE x 100)
folds washed out, on the posterior part in the lower third a surgical suture with continuous thread on a solution of continuity with anfractuous margins in the form of letter “V”, with one side of 5 cm and the other of 4 cm; the lumen of the esophagus is free of any content. Histopathology examination established inflammation in esophagus (fig.1), and hyperemia of the esophagus (fig.2), liver dysplasia – focal necrosis (fig.3), and severe pneumonia (fig.4). The medico-legal conclusion was that the death of the 49-year-old male was non-violent.

The death was caused by cardio-respiratory arrest due to the sepsis status as a result of mediastinitis, bilateral purulent pleuresy, pneumonia and obstructive purulent bronchiolitis, complication that appeared in the evolution of a Boerhaave Syndrome (spontaneous rupture of the esophagus) in the person with chronic alcoholism. On the head, torso and limbs there were no visible traumatic lesions noticed.

Discussion

Esophageal rupture unassociated with external trauma, operation, instrumentation, foreign body or overt esophageal disease had been inappropriately described as “spontaneous perforation of the esophagus” [14].

Most cases (more than 95 percent) are caused by vomiting and the causative term emetogenic may be appropriately applied [15]. Excessive food and alcohol ingestion is usual in such cases of esophageal rupture. Nonemetogenic Boerhaave syndrome, occurring in less than 5 percent of cases, has been described in association with asthma, epilepsy, compressed-air accidents, forceful swallowing, abdominal blows, and straining during parturition or defecation. Lifting is conspicuously absent from many report [16,17,18] that list various other causes and probably accounts for less than 1 percent of cases of Boerhaave syndrome.

All cases of this syndrome have in common increased intraluminal pressure, which may be either primary (such as compressed-air accidents or forceful swallowing), or secondary (such as increased intra-abdominal and intragastric pressure transmitted across the gastroesophageal junction). Risk of rupture is increased if esophagogastric competence is compromised while the upper esophagus is closed by contraction of wall muscle or the inferior pharyngeal constrictor. The mechanism is described by the terms pressure, pneumatic and barogenic rupture. Experiments by Mackler [19] and other with pneumatic distention of the cadaver esophagus have shown as predilection for distal (lower) leftsided rupture. This was true both in situ and after removal of the organ. Weakness is evidently related to the distribution of esophageal muscle. Perforation of the thoracic esophagus is on the most serious digestive tract injuries in regard to mortality and morbidity [11]. It causes a serious condition involving mediastinitis and sepsis, due to the extravasation of digestive secretions and food particles into the mediastinum and pleural space. The prognosis mainly depends on the length of the delay between diagnosis and treatment, and, according to literature, mortality can be as high as 92% if the condition goes untreated and 60% if such treatment is delayed [20].

Postemetic rupture of the esophagus accounts for 10% to 15% of all cases of thoracic esophageal perforation and is the third most common cause of this condition [13]. Diagnosis is particularly difficult and is usually delayed since it is a rare disease that can be confused with other serious clinical entities that are
more common in emergency situation [11]. Clinical suspicion is relation to this entity is of utmost importance for proper diagnosis. Vomiting, particularly when repetitive, could lead to uncoordinated esophageal sphincter reflex opening. This dysfunction could result in a sudden, high magnitude increase in the intraluminal esophageal pressure, leading to rupture in the weakest region, which is the left lateral wall [21].

Treatment of spontaneous esophageal rupture can be either non-operative or operative [10]. Nonoperative treatment is best for patients with a contained perforation and the absence of clinical mediastinitis [22]. Such therapy usually includes targeted drainage, intravenous antibiotics, nasogastric decompression, and enteral nutrition. Patients who are either unstable, have clinically significant mediastinitis, or a non-contained rupture, generally require surgery [23].

When the rupture us larger than 1 cm with considerable mediastinal contamination, the recommended treatment is T-tube controlled esophagocutaneous fistula, which allows esophageal drainage and promotes healing of the surrounding structures. With amore severe case of rupture, thoracotomy with direct repair may by necessary [24,25]. The recommended treatment for special condition, when possible, is as follows: a thoracotomy with primary closure of the defect, with or without local reinforcement; mediastinal debridement and pleural drainage; gastrostomy to divert gastric secretions; and, occasionally, a jejunostomy for nutritional support [26]. Other recommened treatment options include the following: esophagectomy with or without immediate reconstruction of the pharyngogastric tract [27]; esophageal occlusion using cervical esophagogastrostomy, cerclage and gastrostomy; and closure of the defect in conjunction with the use of a T-tube, similar to that used to drain bile ducts. These last procedures are usually performed in very serious situations with significant damage to the local tissue, in cases of late diagnosis or when primary repair is not possible [12,13,28].

In conclusion, postemetic rupture of the esophagus is a serious condition whose diagnosis is usually neglected during the initial assessment. Vomiting, pleural effusion (left, right), thoracocentesis with aspiration therapy usually includes targeted drainage, intravenous antibiotics, nasogastric decompression, and enteral nutrition. Patients who are either unstable, have clinically significant mediastinitis, or a non-contained rupture, generally require surgery [23].

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In conclusion, postemetic rupture of the esophagus is a serious condition whose diagnosis is usually neglected during the initial assessment. Vomiting, pleural effusion (left, right), thoracocentesis with aspiration of fluid suspected to the digestive secretions and elevated levels of amylase were the important factors in making the diagnosis and recommending the surgical treatment [11,29].

References