Boerhaave's syndrome initially presented with sore throat and cough complicated by a bilateral pneumothorax

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Abstract

A 14-year-old male was admitted to the Emergency Department with sore throat and cough. One hour after his admission, he presented a hemodynamic compromise with a respiratory failure. The thoracic tomodensitometry highlighted a tension bilateral pneumothorax and mediastinum consecutive to an esophageal rupture in the left posterolateral wall also known as Boerhaave's syndrome which was treated successfully with a non-operative management. To avoid a recurrence of bilateral pneumothorax, a left pleuroscopy with talc pleurodesis was performed. (Acta gastroenterol. belg., **2020**, 83, **322-324**).

Key words : Boerhaave Syndrome, tension bilateral pneumothorax, hemodynamic compromise.

Introduction

The Boerhaave's Syndrome is typically described as a combination of symptoms represented by lower chest pain, forceful vomiting and subcutaneous emphysema also called Mackler's Triad (1). The clinical features in Boerhaave's Syndrome are often atypical and unspecific. We report a case of Boerhaave's Syndrome whose initially presenting with sore throat and cough was complicated by tension bilateral pneumothorax and pneumomediastinum which was treated successfully with a non-operative management.

This was a spontaneous oesophageal rupture with potentially life-threatening consequences if the diagnosis had be missed or delayed. If not for the uncommon clinical story, which then led to further imaging, this diagnosis could have been overlooked. To our knowledge, this is the only case report where Boerhaave's Syndrome has presented in this way.

Case report

A 14-year-old male was admitted to the Department of Pneumology for spontaneous bilateral pneumothorax. He complained about sore throat with cough. One hour after his admission, he presented a hemodynamic compromise with a respiratory failure.

Chest X-ray was performed after the patient was stabilized with fluid intake and oxygenotherapy. The radiologist objectified a bilateral pneumothorax (Figure 1). At the Emergency Department, two chest tubes were implemented with aspiration in the second intercostal space on medioclavicular line.

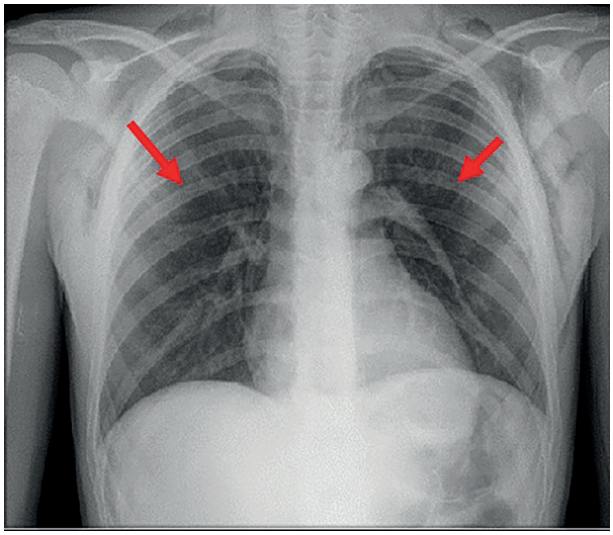

Figure 1. — Chest X-ray showing bilateral pneumothorax.

The patient and his family didn't report any physical traumatism or difficulty in swallowing. He has no relevant past medical history, was not taking any medications and has no allergies. One brother and the sister suffered from gastroesophageal reflux in the childhood. Therefore, the patient remembered he occasionally complained about it too and he frequently underwent hiccup attacks.

Thoracic tomodensitometry centered on the esophageal region was performed three days after his admission in the Department of Pneumology and a disruption in the distal posterolateral wall of the oesophagus extending into the mediastinal cavity (Figure 2A) was highlighted with the abnormal air within the soft tissues and in the mediastinal and pericardic cavities (Figure 2B). The diagnosis of Boerhaave's syndrome was retained.

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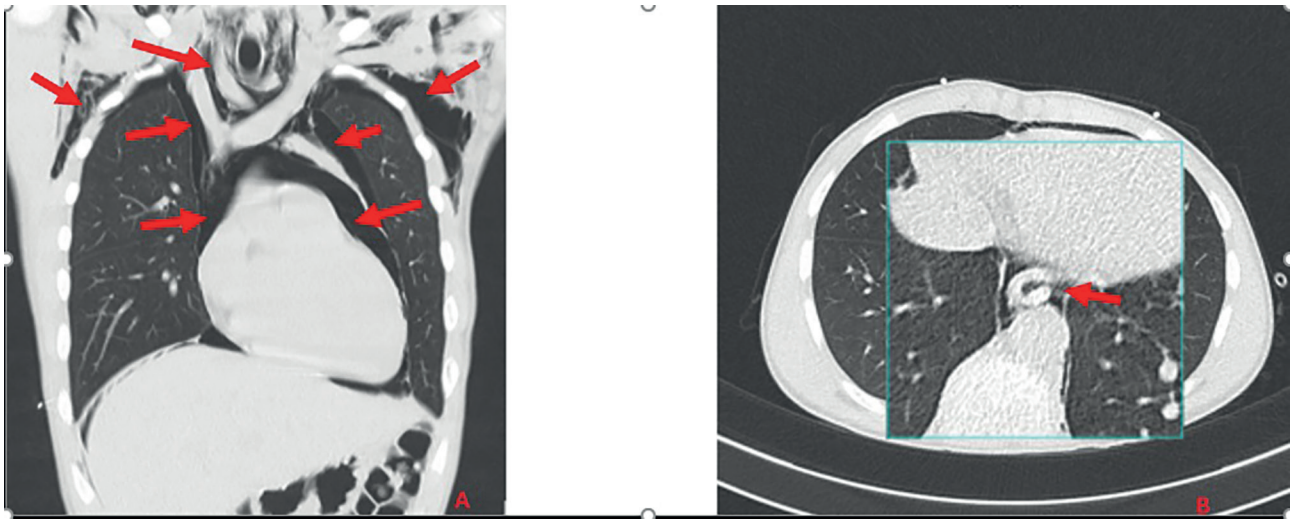

Figure 2. — A. Tomodensitometry of the chest. Multiple pockets of gas in the mediastinal and pericardic cavities with air within the soft tissues. B. Tomodensitometry of the chest (transverse section) shows the disruption in the oesophagus extended to the mediastinal cavity.

A treatment with amoxicillin/clavulanic acid and parenteral nutrition to avoid mediastinitis and pantoprazol to reduce the gastric secretions was started.

The radiological outcome was favourable and we postponed two weeks later the gastroscopy and bronchoscopy. We ensured there was no further leak from the oesophagus with barium swallow, suggesting that the point of rupture had already closed over. There was no leak in the esophagus highlighted with barium follow-through. Those both exams were negatives and we decided with the surgical team to carry out a left pleuroscopy with talc pleurodesis to prevent a recurrence of bilateral pneumothorax.

Discussion

The Boerhaave's Syndrome is also known as spontaneous transmural perforation of the oesophagus. In 1724, Hermann Boerhaave was the first to describe this condition, caused by a sudden rise in intraluminal oesophageal pressure or barogenic trauma following forceful vomiting with failure of the cricopharyngeus muscle to relax (2,3). It is a rare (representing only 15% of case of oesophageal traumatic perforation) but life-threatening condition and requires immediate medical attention (4).

The full-thickness rupture typically occurs on the left posterolateral wall of the lower third of the oesophagus, which is the weak point of oesophageal wall because of the thinning of the wall and normal anatomic deficiency of adjacent support (5,6).

Boerhaave's Syndrome classically presents as Mackler's Triad including : lower chest pain, vomiting and subcutaneous emphysema (4). Symptoms of complete disruption of the oesophagus are often unspecific such as lower pleuritic chest pain, sometimes aggravated by swallowing, associated with vomiting, haematemesis, subcutaneous emphysema, signs of systemic inflammatory response and cardiorespiratory compromisssion (3,7,13). The complete Mackler's Triad is uncommon and present in less than 50% of cases, which can make establishing the diagnosis difficult. Spontaneous oesophageal perforation is often unsuspected or misdiagnosed, closely mimicking more common diseases, including myocardial infarction, pneumonitis, acute pancreatitis, aortic dissection, perforated ulcer and many others (8). Delayed diagnosis may result in serious complications as mediastinitis, pneumonitis, pericarditis, empyema, and death. It carries a high mortality of 20% to 30% and this rises proportionally with the delay of diagnosis (9).

Sore throat with cough is an extremely rare inaugural symptom in Boerhaave's Syndrome. This fact explains why this case was a real diagnostic challenge. The diagnostic could be established thanks to the thoracic computed tomodensitometry performed in this case. This medical imaging technique is extremely helpful and actually considered as "one-stop-shop" with water-soluble swallow to help clinician to put the diagnosis of Boerhaave's Syndrome (10). The radiologist can find air surrounding the oesophagus and mediastinal gas and/or madiastinal fluid collections, oesophageal wall thickening, pleural effusion or hydrothorax (3). This technique provides results quickly and is largely available in Emergency Departments.

Water-soluble swallow can allow us to highlight the extra luminal contrast leak and confirm the diagnosis, but also delineate the anatomical site of the perforation and thus guide the surgeon to close the defect (5,10). However, there is 15-25% of false negative (5) and it's possible to engender a pulmonary oedema consecutive to aspiration of orally ingested contrast because of the patients are often "bed-ridden". That's the reason why we waited to perform this exam that proved no perforation.

In our case, the patient presented bilateral pneumothorax. The situation was under control with the use of chest tubes and an appropriate resuscitation. The tension pneumothorax is a rare complication observed in Intensive Care Unit (about 1-3% of patients). In most cases, we discover a pulmonary origin but a non-pulmonary etiology is sporadically highlighted taking into account a very rare association with Boerhaave' Syndrome (9,11). Our patient developed a hemodynamic compromise, a complication rarely reported, probably consecutively to the pneumomediastinum.

In Boerhaave's Syndrome, a communication with the left pleural cavity is observed in 80-90% of cases and less than 5% for the right pleural cavity (9). In the case we presented, the pneumothorax we observed in the first chest tomodensitometry was clearly dominant at the left side (Figure 2B). The picture observed at the left posterolateral wall is highly suggestive of an oesophageal perforation (Figure 2A). This rupture would explain the presence of gas surrounding the soft tissues. There was no abnormality at the bronchoscopy. Those three arguments helped us to put the diagnosis of Boerhaave's Syndrome despite the absence of perforation at the endoscopy performed two weeks after the admission at the hospital.

It's important to keep in mind oesophageal perforation is not a common condition (only 20 patients in a population of 900 000 during 14 years) and the mortality is significant (10). Following the previous studies, the 30-days mortality rate varies between 8 and 21 percent (12). The mortality is not influenced by the aetiology : the spontaneous oesophageal perforation occurred about half patients and the prognosis is similar to iatrogenic perforation. The mortality in operative and non-operative group is identical (7,12).

There are two parameters to decide between operative versus non-operative management : the size of mediastinal and pleural contamination and the level of systemic sepsis. The aetiology is not a deciding factor (12). The encroachment of the mediastinal space induces extending contamination of the pleural cavity. Therefore, the probability of successful non-operative management is considerably reduced. Non-operative strategies consist in fluid resuscitation, administration of intravenous antibiotics, intercostals tube drainage and acid suppression. The feeding has an important part in the treatment and nasojejunal feeding is mandatory (6).

Surgery was not chosen for our patient who was clinically stable with absence of liquid drainage outside of the oesophagus and none clinical evidence of sepsis.

Conclusion

This Case Report illustrates that unspecific presenting feature like sore throat and cough can occult a rare condition like the Boehaave's Syndrome. Furthermore, a tension pneumothorax can occur consecutively to a non-pulmonary aetiology : the Boerhaave Syndrome was mentioned five times in the medical literature and we reported the sixth case. Finally, the Boerhaave's Syndrome is an important diagnostic challenge because of his frequent unspecific presentation and his life-threatening complication.

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