Missed Diagnosis

Spontaneous rupture of the oesophagus (Boerhaave’s syndrome)

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Summary: Five cases of spontaneous rupture of the oesophagus are reported. All cases had surgery within 12 hours, and all survived, illustrating the value of early diagnosis in this rare condition. None had their diagnosis made before admission to the hospital. Myocardial infarction is the commonest misdiagnosis and frequently results in delayed treatment. We believe that a simple direct question to enquire of the patient whether or not vomiting preceded the onset of the severe pain would significantly reduce the rate of misdiagnosis.

Introduction

Since Boerhaave’s original description of spontaneous oesophageal rupture in 1724 and Barrett’s report on the first successful surgical repair in 1947, more than 300 cases have been reported. Early diagnosis is the key to survival in these patients as delayed diagnosis carries a high mortality. The diagnosis is frequently made late because it is often not considered, and does not present with pathognomonic signs. The value of early diagnosis is illustrated in these five cases. None was diagnosed before hospital admission despite all giving a typical history, and only two were considered to have a surgical condition by the casualty officers.

Case reports

Case 1

A 42 year old man was admitted with severe epigastric and chest pain. Prior to this he had vomited following ingestion of alcohol and peanuts. On examination he was in pain and tachypnoeic. His abdomen was rigid and crepitus was present in the neck. A chest X-ray revealed a left-sided effusion. The patient was explored through a left thoracotomy. A 3 cm tear was found in the distal oesophagus just above the diaphragm. This was closed in two layers and chest drains inserted. He made an uneventful recovery.

Case 2

A 71 year old man presented in the accident and emergency department with severe substernal chest pain and dyspnoea after vomiting. However, the only physical sign elicited was tenderness in the right hypochondrium. Initially chest X-ray, electrocardiogram, serum amylase and full blood count were all normal. He deteriorated on conservative therapy and a subsequent chest X-ray, 2 hours after admission, revealed a pleural effusion on the right side. A rupture of the oesophagus was confirmed by a water-soluble contrast swallow and the patient was explored via a right thoracotomy. A long rent in the mid-oesophagus, 8 cm in length, was found starting just below the level of the azygos vein. There was gross pleural contamination with gastric contents. A satisfactory repair was performed in two layers and the patient made a full recovery.

Case 3

A 66 year old man presented with acute chest pain radiating to the back following the sudden onset of vomiting after consuming a sandwich and a large quantity of beer. On examination he was in shock and ischaemic changes on the electrocardiogram prompted his admission to the medical ward. Crepitus was later noticed in the neck by a nurse on the coronary care unit. A repeat chest X-ray revealed a left pleural effusion with pneumomediastinum. A water-soluble contrast study confirmed oesophageal rupture on the left side. The patient was explored through a left thoracotomy. There

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was gross pleural and mediastinal contamination with a 2.0 cm tear in the distal oesophagus, which was closed in two layers. This patient developed a leak which eventually healed on conservative management.

Case 4

This 67 year old man vomited after having eaten a large meal with alcohol and subsequently developed a sharp chest pain. On admission he was in severe pain with decreased air entry in the left lower chest. A chest X-ray showed a left pleural effusion. The correct diagnosis was made by the casualty officer, and confirmed with a water-soluble contrast swallow. The patient underwent primary repair of a 4 cm tear situated on the left lateral aspect of the lower oesophagus. He made an uneventful recovery.

Case 5

A 52 year old male patient was admitted to the medical ward with a history of epigastric pain radiating to the left shoulder. On examination he had a tachycardia with tenderness in the epigastrium and decreased air entry in the left side of his chest. A chest film revealed a left-sided pleural effusion. Presence of subcutaneous emphysema in the neck supported the diagnosis of a ruptured oesophagus. He admitted to having consumed alcohol and had vomited prior to the pain. A left thoracotomy was performed to repair a 5 cm tear in the lower oesophagus extending through the hiatus across the oesophago-gastric junction.

There was no contamination of the peritoneal cavity. This patient needed prolonged ventilation postoperatively because of severe obstructive airway disease, but was eventually discharged after 5 weeks. He was readmitted a fortnight later with a meat bolus impacted in the lower oesophagus. This was successfully removed through a rigid scope.

Comment

Spontaneous rupture of the oesophagus is a well-documented surgical condition, but its presentation leads to a 'tradition' of misdiagnosis. Surgeons are the most likely to make the diagnosis, but patients are often considered to have a 'medical' problem, which may lead to delay. The best chance of survival follows an early diagnosis with surgical repair of the perforation within 12 hours. The treatment becomes complicated when diagnosis is delayed. Several procedures have been described to overcome difficulties in repair of the oesophageal defect. These include buttressing procedures using diaphragm or fundal patch and procedures that involve some form of oesophageal drainage (T-tube drainage). The more radical procedure of oesophagectomy has been performed for extensive rupture of the oesophagus or when the friable nature of the tissues precluded a simple repair. Most series report considerable morbidity and overall mortality ranging from 8% to 55%. Persistent leakage from the suture line is the main cause of morbidity and leak rates of 15–40% have been reported. In this series one patient developed a leak which eventually healed with conservative management. Sepsis and pulmonary complications also lead to substantial morbidity but modern intensive care units have considerably improved the outcome.

We present these cases mainly to raise the awareness of this condition and to emphasize that the diagnosis can be made from the history in nearly every case. The classical triad of vomiting, chest pain and presence of surgical emphysema was present in only two patients. The cardinal feature, often not volunteered by the patient, is that the severe pain is preceded by a vomit, as was the case in all our patients. A detailed analysis of the symptom complex in 47 personal cases Abbot et al. also confirmed this, except in those patients who had either associated active upper gastrointestinal disease or a neurological disorder. The clinical impression is frequently that of myocardial infarction. The symptoms may also mimic perforated ulcer, dissecting aneurysm, spontaneous pneumothorax, pancreatitis and pulmonary embolism. This condition has also been reported to occur in neonates. Approximately 10% of patients with perforations have pre-existing oesophageal diseases such as oesophagitis, hiatus hernia and diverticulosis. None of our patients had a pre-existing condition.

A chest radiograph may show a pleural effusion or pneumomediastinum though in three of our cases the initial X-rays were interpreted as 'normal'. The V sign of Naciero, a density behind the left cardiac border, has been described as a specific early sign. Pate et al. in their series found that the initial chest roentgenogram was abnormal in 97% of the patients but was interpreted as compatible with perforation of the oesophagus in only 27%. Contrast studies were helpful in establishing the diagnosis and an accuracy of 95% has been achieved by this method of investigation. Endoscopy is unnecessary and may even be contraindicated. Pleural aspiration, a simple though infrequently performed procedure, may reveal the presence of undigested food and a pleural fluid pH below 7.0.

Usually, the tear occurs on the left lateral aspect of the distal oesophagus as seen in four of our cases. Right-sided ruptures (Case 2) are rare. The mechanism of rupture is a build up of pressure
within the oesophagus due to incoordination during vomiting when the cricopharyngeus fails to relax at the appropriate moment. Consumption of alcohol, often with a heavy meal, may induce vomiting in an incoordinated manner.

Our own awareness of this condition has been raised by four cases presenting within 8 months. A similar high incidence of five cases in a 10 month period has been reported by Hendry et al.13

Only about one third of spontaneous oesophageal ruptures are diagnosed initially.4,8 In this report only in two cases was the diagnosis correct on admission. It is noteworthy that one case was spotted by a nurse on the coronary care unit on detecting surgical emphysema in the neck and one by the casualty officer on admission but none by the general practitioner. The casualty officer's awareness had been raised by the preceding cases which allowed her to ask the appropriate question about vomiting and make the diagnosis. This is a rare condition for which a high cost is paid for delay in diagnosis. We would recommend that the patient be asked the direct question of whether or not vomiting preceded the onset of pain.

References

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