Spontaneous Intramural Haematoma Of The Oesophagus: An Uncommon Cause Of Chest Pain

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Citation

Abstract
Spontaneous intramural haematoma of the oesophagus (SIHO) is an important cause of chest pain. It presents as sudden onset of chest pain, typically accompanied by dysphagia or odynophagia and haematemesis. Gastroscopy is safe and diagnostic, although endoscopic findings change rapidly and may be misinterpreted. Conservative management leads to excellent prognosis. Hence, accurate diagnosis of SIHO is vital for appropriate management. We report a case of SIHO in a 67 year old lady with no apparent precipitating factors.

CASE REPORT
A 67 year old women presented with an acute onset central chest pain which occurred while bending down. The pain radiated across her chest and back. This was accompanied by acute odynophagia and bouts of haematemesis. She has a background of rheumatoid arthritis with no previous history of ischaemic heart disease, clotting disorders, peptic ulcer disease or gastro-oesophageal reflux disease. She was not on any regular medications, aspirin or nonsteroidal anti-inflammatory drugs. She did not have a history of vomiting, trauma or ingestion of toxic substance. Her family history included both her father and her son affected by ulcerative colitis.

On examination, she was well and not clinically anaemic. She was apyrexial and had no surgical emphysema. The pulse was 80 beats/min and blood pressure of 140/80 mmHg being equal in both arms. Respiratory system was unremarkable. There was no abdominal tenderness and no melaena. Investigation revealed a normal full blood count (Hb 130g/l, platelets 269 x 109/l). Clotting screen was normal. Biochemical parameters were normal. The chest radiograph (CXR) and electrocardiogram were normal.

The patient was kept nil by mouth and started on intravenous fluids. An urgent endoscopy was performed 48 hours after admission. At 20 cm, just below cricopharyngeus, the mucosa on the posterior wall was elevated. A large bluish submucosal haematoma which extended from this point to within a few centimeters above the gastro-oesophageal junction. (Fig 1a and 1b). The endoscopic features were consistent with spontaneous haematoma of the oesophagus (SIHO). Computed tomography (CT) of the thorax was normal.

Figure 1
Figure 1a and 1b: Intramural oesophageal haematoma. The dark haematoma projects into the lumen with paler mucosa seen lifted over the surface, showing the haematoma is submucosal. (Figure 1b)
The patient responded well to conservative management. She was treated with antibiotics because of a low grade fever and Lansoprazole 30mg daily for gastric acid suppression to prevent acid reflux. Oral fluids were gradually introduced and two days later she was managing a normal diet. She remained well and was discharged 10 days later. She had a repeat gastroscopy six weeks later which revealed a normal oesophagus.

DISCUSSION
SIHO has been considered a rare condition which occurs more commonly in women over 50 years old. This leads to varying degrees of submucosal dissection of the oesophageal wall. Oesophageal apoplexy was the term used to describe the development of spontaneous intramural haematoma of the oesophagus. Marks and Keet first reported a case of SIHO in 1968. The commonest presentation is acute central chest pain. Diagnosis is aided by the common co-existence of haemetemesis and acute dysphagia or odynophagia. Majority of patients present with haemetemesis, usually of mild severity which rarely requires blood tranfusions.

The aetiology of SIHO is uncertain. Submucosal haemorrhage that dissects the submucosal plane and ruptures through the mucosa is one possible theory. This was hypothesised in a patient with caverno-capillary haemangiomatosis; a rare vascular malformation which may have caused the submucosal bleeding that was observed. Extensive intramural haematoma formation has been documented in patients who have haemophilia, thrombocytopenia and in patients treated with anticoagulant. However, haemorrhagic dissection of the submucosa has been shown in patients without coagulopathy as in our case report. The other possibility is whether SIHO is an intermediate injury between Mallory-Weiss tear and oesophageal perforation. This is unlikely as both the latter conditions have profuse vomiting as a major differentiating factor; while vomiting is often minor or absent in SIHO. In our patient, the intraoesophageal pressure may have been increased while bending down, similar to vomiting, resulting in mucosal tear. An abnormal swallowing mechanism may be another aetiology as there has been an association with symptoms occurring after eating and drinking but manometry performed in these patients has been reported as normal.

Endoscopy is a safe procedure and diagnostic for SIHO. However, some authors would argue that with evidence of oesophageal injury, radiological contrast studies should be done first to exclude perforation. In the early stages, the endoscopic appearance of the large haematoma is pathognomonic. The haematoma is usually posterior and can be seen to lie submucosally. Endoscopic features change rapidly over time and may be misinterpreted by endoscopist who are unfamiliar with the condition making it an important training point. The smooth surface of the haematoma breaks down to a longitudinal ulcer over subsequent days before complete resolution. Other appropriate modalities of investigation includes CXR, barium swallow and CT of the thorax. CXR helps to exclude perforation. The barium contrast studies may show ‘double-barelled’ oesophagus and ‘mucosal stripe’ sign; both describe the appearances produced by intramural dissection. CT of the thorax is useful in facilitating early diagnosis of other mediastinal masses including dissecting thoracic aneurysm.

A systematic analysis of similar cases reported in the worldwide literature reveals that it is a benign condition with good prognosis. Management is conservative. Patients are kept nil by mouth initially when there is pain or dysphagia and gradually re-introduced to oral fluids, followed by soft diet. Gastric acid suppression with a proton-pump inhibitor may be reasonable to prevent acid reflux. The role of antibiotic treatment is uncertain and is only clinically indicated in the presence of fever.

CONCLUSION
In conclusion, SIHO may be more common than previously thought and under-recognised. In our case, the clinical significance of the patient's symptoms was not appreciated initially. Prompt gastroscopy led to an accurate diagnosis and management. This case illustrates a rare case of SIHO.
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occurring in a lady with no apparent precipitating factors. It may mimic spontaneous rupture of the oesophagus (Boerhaave syndrome), dissection of the thoracic aorta or myocardial infarction. Confirmation of SIHO is vital for the appropriate management because erroneous thrombolysis or surgical intervention may prove disastrous in a condition with an excellent prognosis on purely conservative management. Recurrence has only been documented in patients with achalasia3,9.

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