Rare disease

Spontaneous intramural oesophageal haematoma: an unusual cause of chest pain

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Correspondence to Dr Elizabeth Sarah Concannon, lizconcannon@gmail.com**Summary**

The following case describes a 29-year-old previously well gentleman who presented with an acute onset of chest pain, dysphagia, odynophagia and vomiting without haematemesis. An oesophageal lesion was visualised on CT angiography and further investigation via oesophago-gastro-duodenoscopy (OGD) diagnosed a spontaneous intramural oesophageal haematoma as the cause of his symptomatology. Conservative medical management in the form of triple therapy and softened diet was well tolerated and a follow-up OGD at 6 weeks after discharge from hospital showed spontaneous resolution of the haematoma.

BACKGROUND

Intramural oesophageal haematoma (IOH) is an uncommon but potentially lethal cause of chest pain. It may present with associated symptoms of odynophagia, dysphagia and haematemesis. He had no associated diaphoresis, no shortness of breath, no palpitations, no cough or abdominal pain. Delay in the diagnosis of IOH may lead to inappropriate management with thrombolytic or anticoagulant therapy which could result in catastrophic oesophageal haemorrhage. Conservative medical management, as described in our case, is appropriate once IOH is confirmed with the majority of cases resolving spontaneously without adverse sequelae.

CASE PRESENTATION

A 29-year-old construction worker was referred to an accident and emergency department by his general practitioner presenting with a 2 day history of sudden onset central 'gripping' chest pain radiating directly through to his back associated with persistent nausea and two episodes of vomiting. This pain was non-pleuritic in nature. He also complained of acute dysphagia and odynophagia. He described a foreign body sensation 'like a golf ball' lodged in his chest and throat for 2 days, rendering him unable to eat or drink comfortably. He had two episodes of vomiting on the day of his admission to hospital without haematemesis or malena. His chest pain intensified on swallowing and was not exacerbated or relieved by either episode of vomiting. He had no associated diaphoresis, no shortness of breath, no palpitations, no cough or abdominal pain. There was no recent history of foreign body/toxin/excessive alcohol ingestion or of other trauma to the oesophagus. The patient had no personal or family history of haematological disorders such as haemophilia.

The patient was well prior to this admission except for a 1 year history intermittent acid reflux and waterbrash, he had no previous surgery and had never had an oesophago-gastro-duodenoscopy (OGD). He was not taking any regular medications. He did not suffer from hypertension, did not

smoke and consumed approximately 10 units of alcohol per week. He had a strong family history of ischaemic heart disease (father at the age of 40 suffered from acute myocardial infarction (MI), brother at the age of 38 suffered an acute MI).

On examination, he was afebrile with SpO₂ of 100% on room air, heart rate 68 bpm and blood pressure 166/91 in both arms and respiratory rate of 18/min at rest. Cardiovascular and respiratory examinations were unremarkable. Gastrointestinal (GI) examination revealed a soft non-tender abdomen with some discomfort in the epigastric area on deep palpation. Murphys sign was negative. Bowel sounds were present. No organomegaly or palpable masses identified and hernial orifices were intact.

INVESTIGATIONS

Serial ECGs were performed which showed T wave inversion in leads II and III and no acute ischaemic changes. Serial troponins and other cardiac markers were not supportive of an acute ischaemic event. Other blood parameters on admission including white cell count, platelet count, coagulation screen, liver screen, amylase, urea, creatinine and electrolytes were all within normal limits.

- ▶ Haemoglobin 14.3 (13–18) g/dl
- ▶ D-dimer 367.0 (0–255) ng/ml
- ▶ Troponin T <0.010 (0.0–0.1) ng/ml and <0.010 when repeated.

A chest x-ray was performed as part of his initial investigation on day 1 of his admission which showed normal heart and mediastinal contours and clear lung fields. A CT thorax with intravenous contrast was performed on day 2 of this admission which revealed an extensive intramural oesophageal mass extending from the distal oesophagus to the oesophagogastric junction. A crescentic rim of air (red arrows on CT images/figure 1A,B) could be seen posteriorly to this uniformly cystic lesion in the oesophageal wall which extends from the 3'o clock to the 9'o clock positions. The aorta and great vessels enhanced uniformly with no dissection or mediastinal haematoma.

Figure 1 (A, B) CT images: Crescentic shaped mass of intramural origin in the distal oesophagus extending from approximately 29 to 40 cm. A crescentic rim of air can be seen posteriorly (see red arrow) to the lesion which extends from the 3'o clock to the 9'o clock positions to the level of the oesophagogastric junction. The lesion has a uniformly cystic attenuation. Appearances are consistent with a submucosal haematoma/leiomyoma/duplication or simple cyst of oesophagus.

An OGD was performed on day 3 of this admission (figure 2) to further define the nature of the oesophageal lesion seen on CT. A lesion with the characteristic appearance of an oesophageal haematoma was seen extending from 29 cm to the oesophago-gastric junction (OGJ) at 40 cm (figure 2). The stomach was normal apart from fullness and oedema of the aspect of OGJ and there was no blood within the stomach. The first and second part of duodenum were normal. An antral biopsy was taken for Clo Testing, which was positive for *Helicobacter pylori* colonisation. The haematoma appeared to be spontaneous with no associated comorbidity.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis: (Pre CT)

- Gastrooesophageal reflux disease/gastritis
- ACS
- Pulmonary embolism (PE)

- Aortic aneurysm
 - Thoracic aortic dissection
 - Boerrhaves syndrome
 - Mallory Weiss tear
 - Retrosternal goitre
 - Oesophageal/gastric/bronchial carcinoma
- Differential diagnosis: (based on CT appearance)
- Spontaneous intramural oesophageal haematoma (SIOH).
 - IOH secondary to trauma/ingestion of toxic substances or foreign bodies/iatrogenic from interventions such as oesophagoduodenoscopy, ng tube insertion, cardioversion/haematologic disorder etc.
 - Leiomyoma. Statistically commonest benign submucosal oesophageal tumour.
 - Simple oesophageal cyst/oesophageal duplication cyst—More commonly diagnosed in childhood but may be similar in presentation to IOH.

TREATMENT

The management plan post OGD was of conservative medical management:

- 1) Soft diet.
- 2) Triple therapy: proton pump inhibitor (PPI) with clarithromycin and amoxicillin for 14 days.
- 3) PO/IM antiemetic as required.
- 4) Regular analgesia with regular paracetamol and opiate based analgesia as required.
- 5) Advised not to return to construction work and to avoid exertion for at least 6 weeks.

OUTCOME AND FOLLOW-UP

The patient responded well to the above management and was discharged home after 5 days in hospital with complete resolution of chest pain, odynophagia and nausea. Upon discharge his vital signs and haemoglobin remained stable, he was tolerating a soft diet well and had no further episodes of vomiting or haematemesis.

He underwent repeat OGD 2 weeks post discharge whereby a resolving haematoma was seen at 30–38 cm with no obstruction. OGD was repeated once more at 6 weeks post discharge. This showed slightly erythematous mucosa at 30–38 cm and together with a repeat CT thorax the impression was of a resolved spontaneous oesophageal haematoma without underlying oesophageal disease or identifiable cause.

He was followed up in the surgical outpatient clinic 1 month later. He has returned to work without incident and has been asymptomatic and well since discharge on a once daily dose of PPI which he was advised to continue for 6 more weeks.

DISCUSSION

Intramural or submucosal oesophageal haematoma (IOH) is a rare and commonly overlooked cause of chest pain whereby blood dissects between the mucosal and muscular layers of the oesophagus. Spontaneous IOH was first described in 1968 by Marks and Keet¹ and has been well described in literature since then in the form of various case reports and case studies. IOH occurs with greater frequency in middle aged and older female patients, particularly in those with known aetiology/risk factors,^{1 2} making the case described above relatively atypical.

Aetiology

Oesophageal haematoma may occur secondary to a variety of aetiologies; trauma, ingestion of toxic substances, ingestion of pills and foreign bodies, iatrogenic from interventions such as nasogastric tube insertion, endoscopic interventions and even electrical cardioversion.³ It may also occur secondary to coagulopathy⁴ or secondary to anticoagulant/antiplatelet agents. A recent case report described haemodynamic compromise occurring secondary to thrombolysis for treatment of acute MI in a gentleman with a coexisting oesophageal haematoma⁵ and the potential hazards involved in mis-diagnosing SIOH as ACS have been previously described.⁶ Mallory Weiss tear has been suggested as another possible aetiology, although the evidence to support this hypothesis is circumstantial.⁷ It also occurs spontaneously as in the case described above, without any identifiable cause although

Figure 2 Oesophagogastroduodenoscopy image: A characteristic dark blue discoloration of oesophageal mucosa is seen here extending from 29 to 40 cm, consistent with oesophageal haematoma, partially occluding the lumen in an otherwise normal oesophagus with no active bleeding or Mallory Weiss tear. The haematoma appears spontaneous with no associated comorbidity or identifiable aetiology on either radiological/endoscopic investigation or in the patients history. Picture taken at 35 cm.

an underlying aetiology is found in over 60% of cases of IOH.⁸

Signs and symptoms

The cardinal presenting feature of IOH includes severe retrosternal chest pain usually sudden in onset and sharp in character which can be associated with haematemesis or vomiting and acute dysphagia or odynophagia. At least two of these symptoms are present in up to 50% of cases of IOH.⁹ Obstructive symptoms may occur when a large IOH progressively obstructs the oesophageal lumen. Careful history, examination and timely investigation is required to diagnose IOH and to differentiate it from other causes of chest pain and vomiting as listed in the differentials above.

Investigation

CT, upper endoscopy and endoscopic ultrasound allow definitive diagnosis of IOH. CT thorax imaging in the case of IOH commonly demonstrates an oesophageal mass with a density similar to that of blood. Clinical correlation and further investigation is often required to differentiate between IOH and other types of oesophageal mass such as malignant lesions/benign simple or duplication cyst/leiomyoma. CT is useful in the early diagnosis of other mediastinal masses, such as dissecting thoracic aneurysm.

Endoscopy reveals a pathognomonic smooth swelling underlying an area of blue-violet mucosal discoloration with or without extrinsic compression. The mucosa may be friable and in some cases an aetiological factor may be seen at endoscopy such as a Mallory weiss tear indicating

excessive vomiting/retching as a cause of IOH. It is worth noting that full thickness perforation of the oesophagus has been reported during endoscopy of IOH.¹⁰ However, it remains a valuable tool in the diagnosis and management of this condition.

Endoscopic ultrasound may be required in certain circumstances to identify the depth/mural level invaded by an oesophageal wall mass. It is also a useful tool to exclude aortic dissection/upper GI malignancies and also allows histological diagnosis with fine needle aspiration cytology as appropriate. Oesophageal wall biopsy would not be advisable however, where there is a suspicion of IOH due to the potential for catastrophic haemorrhage.

Radiological contrast studies are useful initial investigations to exclude oesophageal perforation in cases where there is evidence of oesophageal injury. In cases of IOH, barium swallow studies may show a luminal filling defect (the so called 'double barrel sign' which represents intramural dissection of the IOH), although this technique has inferior diagnostic potential compared to CT/endoscopy due to its lack of specificity.

It is particularly crucial to differentiate large IOH from aorto-oesophageal fistula (AOF), another less common but more life threatening cause of upper GI bleeding.¹¹ AOF usually occurs in association with thoracic aortic aneurysm, oesophageal malignancy, foreign bodies, following trauma or prosthetic aortic grafting. A pulsatile esophageal mass may be seen at endoscopy in the case of AOF and comparison of pre and post contrast CT studies with or without endoscopic ultrasound could identify an intimal flap or communication between the esophagus and the aorta.

Treatment

Once a definitive diagnosis of IOH has been made conservative medical management (as described in this particular case) is usually appropriate. This involves: soft diet/NPO order initially followed by gradual introduction of soft diet as appropriate to patients clinical condition, antiemetics as needed, acid suppression with PPI to reduce the risk of oesophageal ulceration and correction of coagulation abnormalities as indicated. The majority of cases resolve spontaneously within 2–3 weeks via luminal haematoma ulceration/rupture followed by mucosal healing without long-term sequelae and recurrence is extremely rare.¹² In two separate case series of IOH, 80% of cases resolved with conservative management¹³ with the remainder of cases requiring emergency surgical intervention. Sclerotherapy may also be employed at endoscopy to manage extensive oesophageal haematoma or IOH with haematemesis¹⁴ in suitable cases. However, endoscopic sclerotherapy also

carries a risk of both haematoma rupture and indications for its use as therapeutic management of IOH have not yet been clearly defined.

Learning points

- ▶ A rare cause of chest pain which should be considered as a differential diagnosis.
- ▶ Treatment of ACS/PE with thrombolysis/antiplatelet agents could cause massive GI bleed in presence of oesophageal haematoma.
- ▶ CT and endoscopic appearance of IOH may be mistaken for oesophageal tumours/varices which could lead to inappropriate surgical management.
- ▶ Conservative medical management of SIOH is appropriate in the majority of cases.

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Competing interests None.

Patient consent Obtained.

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