Dissecting intramural haematoma of the oesophagus (DIHO) is a rare condition in which intramural haemorrhage leads to submucosal dissection of the oesophageal wall. It is usually associated with a rapid increase in intraoesophageal pressure, trauma or a coagulation disorder. The clinical presentation is with chest pain, haematemesis and dysphagia/odynophagia and an accurate history is vital to help distinguish it from other causes of acute chest pain, such as myocardial infarction, aortic dissection or oesophageal perforation. DIHO usually runs a benign course and has an excellent prognosis when managed conservatively. We present a rare case of spontaneous massive DIHO presenting with pericarditic electrocardiographic changes and causing left atrial compression. We emphasize the importance of an accurate history and document the utility of various imaging techniques, such as endoscopic ultrasound and computed tomography (CT), to make the diagnosis.

2. Case report

A 42-year-old male presented with a 48-h history of sudden onset severe central chest and interscapular pain associated with dysphagia and odynophagia. There was no history of vomiting, haematemesis or trauma. There was little previous medical history of note and he was not taking any regular medication. On examination, vital signs were: blood pressure, 104/49 mmHg with no differential between arms; pulse, 125 beats/min; respiratory rate, 24 breaths/min; and temperature, 39.3°C. The remainder of the examination was unremarkable. Blood analysis revealed Hb 15.5 g/dl, WBC 13.3×10⁹/l, Plt 284×10⁹/l, INR 1.2. Plain chest radiograph revealed the edge of a mediastinal soft tissue mass lateral to the right heart border. Twelve lead electrocardiogram revealed widespread saddle-shaped ST elevation across the precordial and limb leads consistent with pericarditis. Transthoracic echocardiogram revealed no evidence of pericardial effusion but severe compression of the left atrium. Upper GI endoscopy revealed extrinsic compression of the length of the oesophagus. A spiral chest CT with oral and intravenous contrast revealed a 10 cm retrocardiac paraoesophageal collection extending from the root of the neck down to the diaphragmatic hiatus with significant compression of the left atrium and the oesophageal lumen but no extravasation of contrast medium or air into the mediastinum; there were also bilateral pleural effusions with right basal atelectasis (Fig. 1). The appearances were consistent with a large dissecting intramural haematoma of the oesophagus. The patient was successfully managed conservatively with haemodynamic support in a high dependency unit setting. The symptoms gradually resolved and the patient was discharged after 7 days. Endoscopic ultrasound scanning 5 weeks later revealed a 7.5 cm×4.6 cm submucosal haematoma from 28 to 42 cm from the incisors (Fig. 2).

3. Discussion

The three different types of acute oesophageal injury are a mucosal tear (Mallory-Weiss syndrome), full-thickness...
rupture (Boerhaave’s syndrome) and dissecting intramural haematoma. Of the three, intramural haematoma is the least commonly encountered and the treatment and prognosis are the same as that for Mallory–Weiss syndrome [1]. Eighty percent of patients with DIHO have at least two of three typical presenting features of chest pain, haematemesis and dysphagia/odynophagia [2]. The differential diagnosis includes other causes of central chest pain and it is vital to obtain an accurate history of both gastrointestinal and cardiovascular symptoms. Analysis of the precipitating factors suggests that there are three main subgroups [2]. Firstly, a sudden pressure change in the oesophagus (e.g. swallowing, vomiting) perhaps associated with a temporary disruption in the normal co-ordinated opening mechanism of the upper and lower oesophageal sphincters. Secondly, direct injury following an endoscopic therapeutic intervention (e.g. oesophageal dilatation). Thirdly, about one fifth of patients appear to have a truly spontaneous origin although this may be associated with an underlying predisposition to abnormal pressure changes within the oesophagus (e.g. achalasia) or a bleeding disorder (e.g. due to anti-platelets, anti-coagulants or thrombolytics).

The pathophysiology is characterised by submucosal haemorrhage that dissects the submucosa and classically occurs in the distal oesophagus because this region is least supported by adjacent structures such as the trachea or heart [3]. The diagnosis of DIHO can be safely made with several complimentary investigations [1]. Firstly, contrast swallow reveals a 'double-barrelled oesophagus' (the 'mucosal stripe sign') or an elongated tubular filling defect [4]. Secondly, direct inspection of the oesophageal mucosa by endoscopy usually reveals a large fluctuant purplish haematoma compressing the lumen with the paler mucosa lifted over its surface indicating that the haematoma is submucosal in origin. Thirdly, cross-sectional imaging (e.g. CT/MRI) is invaluable and excludes other differential diagnoses, such as aortic dissection, and provides a non-invasive tool for follow-up [5,6]. Finally, endoscopic ultrasound scanning (EUS) clearly reveals that the haematoma lies in the submucosa and not in the extramural posterior mediastinal soft tissues [3].

Once the diagnosis has been reached, conservative treatment with correction of any coagulopathy is usually successful with resolution of symptoms occurring within 7-14 days [7]. Serial endoscopy reveals that the dissected mucosal layer sloughs away leaving a large longitudinal ulcer [8]. Occasionally severe bleeding or oesophageal perforation may occur necessitating urgent surgical intervention [2]. Follow-up investigations invariably show significant or complete resolution with no reports of late sequelae [7].

In conclusion, DIHO is a rare condition which should be considered in patients presenting with acute chest pain, haematemesis and dysphagia/odynophagia, particularly in the presence of a bleeding disorder or where there has been

Fig. 1. (A) Axial section obtained at the level of the inferior pulmonary veins showing a large soft tissue mass in the oesophageal wall compressing the left atrium and causing passive atelectasis of the adjacent lower lobes with small bilateral pleural effusions. (B) Coronal multi-planar reformat posterior to the left atrium demonstrating the large peri-oesophageal soft tissue mass causing lateral displacement of the descending aorta.

Fig. 2. Endoscopic ultrasound (EUS) of the oesophagus demonstrating a large (7.5×4.6 cm) submucosal hypoechoic area extending posteriorly and laterally to the right.
recent administration of antiplatelets, anticoagulants or thrombolytics. An accurate history eliciting both cardiovascular and gastrointestinal symptoms is vital to guide subsequent investigations and avoid inappropriate treatment. It carries an excellent prognosis when treated conservatively with surgical intervention reserved for those with severe haemorrhage or oesophageal perforation.

References


