Oesophageal haemangioma: imaging characteristics of this rare condition

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Oesophageal haemangiomas are rare, benign, vascular tumours that have characteristic features on computed tomography and magnetic resonance imaging. This enables radiologists to suggest the correct diagnosis without the need for invasive biopsy. Diagnostic characteristics include phleboliths seen on computed tomography, and intense homogeneous enhancement of the lesion on magnetic resonance imaging. These tumours have life-threatening haemorrhagic potential and therefore require surgical resection.

Case report

A 59-year-old woman, who had a history of cervical cancer managed with both chemotherapy and radiotherapy, presented in December 2008 with a 1-week history of vomiting blood and passing melaena. Her haemoglobin level had dropped from 93 g/L to 75 g/L. The patient had been referred for endoscopic evaluation of her oesophagus and this examination revealed a mid-oesophageal, submucosal mass measuring 20 to 25 cm. There were no ulcerations or erosions. An endoscopic ultrasound (EUS) showed a heterogeneously echogenic lesion involving the submucosa and extending outside the muscularis propria. Multiple large vascular channels ranging in size from 7 to 15 mm were present (Fig 1a and b). Computed tomography (CT) of the thorax showed a mildly enhancing oesophageal mass and the presence of phleboliths (Fig 2a). Magnetic resonance imaging (MRI) of the thorax was performed to better delineate the longitudinal extent of the lesion. Efervescent granules were given to the patient before scanning to ensure adequate distension of her oesophagus. Magnetic resonance imaging showed a large, well-marginated submucosal mass arising from the left posterolateral wall of the upper third of the oesophagus, causing narrowing and displacement of the lumen. The longitudinal length of the lesion was about 6 cm. The mass was isointense on T1-weighted imaging, heterogeneously hyperintense on T2-weighted imaging and showed avid enhancement after intravenous contrast was administered. A few hypointense foci seen on T2-weighted imaging, suggestive of calcifications/flow voids, were evident within the oesophageal mass (Fig 2b, c, and d). These imaging features are classical characteristics of benign oesophageal haemangiomas. The patient underwent a robotic-assisted partial oesophagectomy (Fig 1c) after which oesophageal haemangioma was confirmed histologically.

Discussion

Gastro-intestinal haemangiomas are uncommon benign vascular tumours that usually present during infancy and childhood but may occur at any age and be found anywhere along the gastro-intestinal tract. They are found most frequently in the small intestine and account for 7 to 10% of all benign small intestinal tumours.1

Oesophageal haemangiomas are very rare, representing 3.3% of all benign oesophageal tumours.2 These tumours may occur at any level within the oesophagus but the most common location is the lower oesophagus, followed by the middle, then the upper oesophagus. They affect both genders equally3 and usually occur singly. Multiple lesions are associated with Osler-Weber-Rendu disease, Klippel-Trénaunay syndrome, or the congenital blue rubber bleb nevus syndrome. Oesophageal haemangiomas cause symptoms of obstruction and haemorrhage, including dysphagia, dyspnoea, haematemesis and melaena. Chest or epigastric pain is occasionally reported. Barium oesophagogram findings are non-specific, showing either a well-defined lobulated intramural mass, pedunculated intraluminal mass, or an infiltrating annular mass.4 Endoscopic ultrasound is the most sensitive and specific method for assessing the location, depth, and size of an oesophageal tumour.4 Use of Doppler ultrasonography with EUS enables vascular channels within the haemangioma to be demonstrated precisely, information that will help the surgeons to decide whether laparoscopic surgery is feasible or not. On CT scanning, an oesophageal haemangioma usually appears as a well-defined soft tissue mass within the oesophageal wall of the oesophagus. Phleboliths are apparent on CT and
considered a specific finding for this tumour. In the absence of phleboliths, the radiological diagnosis is less certain and the differential diagnosis should include leiomyomas, polyps, neurofibromas, and gastro-intestinal stromal tumours.

In summary, though they are rare, oesophageal haemangiomas can usually be identified by their characteristic imaging features, correlated with clinical symptoms and endoscopic findings. Endoscopic ultrasound is the most sensitive and specific investigative tool available for diagnosing and delineating the extent of a gastro-intestinal haemangioma. As EUS becomes more readily available, CT and MRI should be considered in areas where EUS is not accessible. Introduction of effervescent granules before CT/MR scanning permits a more accurate assessment of the extent of the lesion.

References