CASE REPORT

Giant oesophageal leiomyoma causing severe hypertension

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SUMMARY

Leiomyoma is the most common oesophageal tumour and is symptomatic in about half of the patients. Dysphagia is the most common symptom. A 41-year-old woman presented with recent onset of severe orthostatic hypertension. During the cardiological work-up, a mediastinal mass was found at transthoracic echocardiogram. Further investigation (upper gastrointestinal endoscopy, endoscopic ultrasonography, CT scan and cardiac MRI) confirmed the diagnosis of a large oesophageal submucosal mass compressing the supradiaphragmatic inferior vena cava. The mass was resected through a minimally invasive right thoracoscopic approach with complete relief of symptoms and compression on the vena cava.

BACKGROUND

Oesophageal leiomyoma typically located in the middle thoracic oesophagus, accounts for approximately two-thirds of all benign oesophageal tumours. The diagnosis is often incidental due to the non-specific clinical presentation or even the absence of symptoms. Dysphagia and/or retrosternal pain are the most common symptoms. Diagnosis is usually achieved by barium swallow study, upper gastrointestinal endoscopy, endoscopic ultrasonography and/or CT scan.^{1–3} Surgical enucleation through videothoracoscopy represents the gold standard treatment in selected patients and is associated with less morbidity and shorter hospital stay compared with thoracotomy.⁴ We present the case of a giant oesophageal leiomyoma incidentally discovered during work-up for severe systemic hypertension refractory to medical therapy.

CASE PRESENTATION

A 41-year-old woman was referred after incidental diagnosis of a mediastinal mass during echocardiography. Her body mass index was 20. Over the past few months she had been reporting of fatigue and palpitations. A severe hypertension with daily peaks up to 240/150 mm Hg was diagnosed by her general practitioner. Triple therapy with olmesartan, hydrochlorothiazide and nifedipine was ineffective. Patient's medical history was otherwise unremarkable, and there was no family history of hypertension.

INVESTIGATIONS

The physical examination was normal and the ECG showed normal sinus rhythm. Transthoracic echocardiography showed mild hypertrophy of the left ventricle and an isoechoic round mediastinal mass

with hyperechoic echoes and clear borders. The patient underwent an upper gastrointestinal endoscopy which revealed an extrinsic oesophageal compression between 23 and 34 cm from the incisors without any mucosal alteration. CT scan revealed the presence of an extramucosal oesophageal mass. Endoscopic ultrasonography confirmed that the mass originated from the fourth oesophageal muscle layer.

To further investigate the relationship of the mass with the heart and great vessels, a cardiac MRI was performed which showed a roundish mass, 88×59×28 mm in size, originating from the oesophagus and maintaining a cleavage plane with surrounding mediastinal structures, but compressing the supradiaphragmatic tract of the inferior vena cava (figure 1).

TREATMENT

The patient underwent surgical resection through a right thoracoscopy with a three-port access in the semiprone decubitus position and double-lung ventilation. Both vagal nerve trunks were identified and spared. On-table endoscopy was performed to confirm the location of the mass and to check the integrity of the oesophageal mucosa during and after surgical enucleation (figure 2). The surgical specimen was extracted by enlarging one of the port sites. Pathological examination confirmed the diagnosis of leiomyoma. Immunohistochemistry resulted negative for CD34 and c-Kit, and positive for actin, desmin and S100. Ki-67 index was low (figure 3).

OUTCOME AND FOLLOW-UP

A gastrographin swallow study on postoperative day 2 showed no leakage and a regular oesophago-gastric transit. The patient was allowed a soft diet

Figure 1 MRI showing the oesophageal leiomyoma compressing the inferior vena cava (L=leiomyoma, yellow arrow shows the inferior vena cava).

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Figure 2 Left: right thoroscopic approach showing a large roundish mass adherent to the oesophageal wall. Intraoperative endoscopy showed an extramucosal compression on the right wall of the oesophagus. Right: operative specimen.

Figure 3 Histology specimen of oesophageal leiomyoma. Left: the tumour is composed of irregularly oriented bundles of well-differentiated smooth-muscle cells. Right: immunohistochemical positivity for smooth-muscle actin is typical of this tumour.

Figure 4 Preoperative and postoperative cardiac MRI showing the compressed vena cava (left) and relief of compression after surgery (right).

and was discharged on the fifth postoperative day with a prescription of nebivolol 5 mg/day. At the 6-month follow-up visit she was asymptomatic and the hypertensive peaks had completely disappeared. An upper gastrointestinal endoscopy showed normal findings. The cardiac MRI confirmed that no residual mediastinal mass was present and, importantly, there was no evidence of compression on the inferior vena cava (figure 4).

DISCUSSION

To the best of our knowledge, this is the first reported case of a giant oesophageal leiomyoma presenting with severe hypertension uncontrolled by medical therapy. The pathophysiological mechanism explaining the orthostatic hypertensive peaks in our patient may involve the activation of the sympathetic nervous system with failure of the baroreflex pathway as a consequence

of subdiaphragmatic venous pooling and related decrease of cardiac output. Orthostatic hypertension immediately on standing has also been described in patients with pheochromocytoma and mast-cell activation disorder.⁵ We have previously described a similar clinical picture in a patient with postsurgical chylothorax who was completely relieved after reoperation and ligation of the thoracic duct.⁶

Oesophageal leiomyomas are asymptomatic in 50% of cases. Symptoms such as dysphagia and retrosternal pain are present in the other half of the patients, more frequently in those with a mass larger than 5 cm.⁷ Cardiorespiratory symptoms are uncommon. Cakar *et al*⁸ reported a patient with oesophageal leiomyoma presenting with progressive dyspnoea and fatigue; echocardiogram, CT scan and cardiac MRI demonstrated extrinsic left atrial compression and impaired left atrial filling that were relieved after enucleation of the mass via thoracotomy. Xu *et al*⁹ reported a patient

with a 2 cm oesophageal leiomyoma and swallowing-induced supraventricular tachycardia. The leiomyoma was resected endoscopically and immunohistochemistry was positive for S100. They speculated that there was a high density of sympathetic nerve fibres between the atrium and oesophagus and a sympathetic neural reflex may have triggered the arrhythmia.

Videothoroscopic enucleation is considered today the gold standard approach in patients with small-to-medium oesophageal leiomyomas.¹⁰ We have shown that even a giant leiomyoma can be safely resected through a minimally invasive approach in the semiprone position. Postoperative follow-up showed complete relief of symptoms and compression on the vena cava.

Learning points

- ▶ Oesophageal leiomyoma typically presents with dysphagia in symptomatic patients.
- ▶ There were no foregut symptoms in our patient, and the diagnosis was incidental at echocardiogram performed for severe, refractory hypertension.
- ▶ Cardiac MRI confirmed the compression of the supradiaphragmatic inferior vena cava, and the mass was resected through video-assisted thoracoscopy.
- ▶ Fatigue, palpitations and hypertension were relieved by surgery.
- ▶ The relationship between caval compression and hypertension remains unclear, but overcompensation by sympathetic pressor reflexes may explain the clinical picture.

Contributors AL, PM and EA researched and wrote the case report under the supervision of LB.

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