CASE REPORTS

Endoscopy and computed tomography in the diagnosis and follow up of oesophageal leiomyoma

M A Cox, B T Cooper, G Sagar

Abstract

Leiomyoma of the oesophagus, although the commonest benign oesophageal tumour, is still rare compared with malignant tumours of the oesophagus. Leiomyomas of the oesophagus are usually diagnosed on barium swallow or histological examination after section. Five cases of leiomyoma of the oesophagus are reported where diagnosis was made by the combination of barium swallow, upper gastrointestinal endoscopy, and computed tomography and all but one patient were followed up for one to four years. The endoscopic biopsy specimens were non-specific in all five patients but none showed any evidence of malignancy. None of the five patients had a history of dysphagia. This paper describes a conservative approach to medical treatment in asymptomatic oesophageal leiomyoma rather than surgical excision as commonly published. It also emphasises the importance of negative endoscopic pinch biopsy specimens and the role of computed tomography in the diagnosis of oesophageal leiomyoma.

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Leiomyomas account for more than 50% of benign oesophageal tumours.¹ They are still rare, however, when compared with the incidence of oesophageal carcinoma. According to one report, leiomyoma constitutes 0.4% of oesophageal tumours.² Most patients with oesophageal leiomyoma are asymptomatic, however, symptoms most commonly reported are dysphagia, which tends to be intermittent, pain that occurs retrosternally or over the left chest, heartburn, and weight loss.³⁻⁵ To date patients with symptoms have tended to have had surgical excision of the tumour.

Published reports on the subject of oesophageal leiomyomas have been essentially from a surgical perspective and in this paper, we for the first time discuss the treatment of oesophageal leiomyomas from a medical viewpoint. The important features discussed are the distinctive computed tomography appearances, the role of endoscopic

biopsies, and the longterm endoscopic and computed tomography follow up of leiomyomas of the oesophagus.

Case reports

Case 1

A 48 year old Indian man was referred to the gastroenterology clinic with a 12 month history of constant upper abdominal pain precipitated by eating. There was no history of nausea, vomiting, chest pain, heartburn or dysphagia. An upper gastrointestinal endoscopy performed in May 1989 showed two mid-oesophageal submucosal masses; which were biopsied. The oesophagus was otherwise normal and the stomach and duodenum were normal. The biopsy specimens were non-specific histologically with no evidence of neoplasia. A barium swallow showed a 4.5 cm smooth impression on the wall of the mid-oesophagus compatible with a submucosal mass lesion. Before and after contrast, computed tomography showed a well defined enhancing dumb bell shaped mass arising from the wall of the oesophagus just above the level of the carina. There was no ring enhancement of this lesion. The diagnosis of leiomyoma of the oesophagus was made. The patient was followed up in the gastroenterology clinic and had a repeat upper gastrointestinal endoscopy and computed tomography of the oesophagus three years later, neither of which showed any change in the size or shape of the lesion. The endoscopic biopsy specimens were non-specific with no evidence of neoplasia.

Case 2

A 46 year old woman with a history of excess alcohol intake was investigated in February 1990 for folate deficiency. She had a history of nausea and vomiting but no chest pain, heartburn or dysphagia. An upper gastrointestinal endoscopy was performed to obtain a small bowel biopsy specimen. This showed a submucosal mass in the mid-oesophagus. The biopsy specimens of this lesion were non-specific with no evidence of neoplasia. A barium swallow showed a well defined filling defect arising from the left side of the oesophagus posteriorly about 5 cm below the carina. Pre and post contrast enhanced

Gastroenterology Unit
M A Cox
B T Cooper

and Department of Radiology
G Sagar

City Hospital, Birmingham

Correspondence to:
Dr B T Cooper,
Gastroenterology Unit, City
Hospital, Dudley Road,
Birmingham B18 7QH.

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Figure 1: Barium swallow shows well defined polypoid mass arising from the wall of the distal oesophagus (arrow) (case 3).

computed tomography of the mediastinum showed a 3 cm well defined soft tissue mass with homogeneous enhancement arising from the wall of the mid-oesophagus just below the carina and abutting the descending aorta. There was no ring enhancement. A diagnosis of leiomyoma of the oesophagus was made. No cause for her folate deficiency was found except for alcohol excess. She was treated with oral folic acid supplements. She was followed up in the gastroenterology clinic and two and a half years later had an upper gastrointestinal endoscopy and biopsy at which the appearances were unchanged and a computed tomography showed no change in the size or appearance of the oesophageal mass lesion.

Case 3

A 61 year old schizophrenic man was referred to the gastroenterology clinic for an upper gastrointestinal endoscopy in August 1992 with a short history of epigastric discomfort and vomiting, but no chest pain, heartburn or dysphagia. The endoscopy showed mild reflux oesophagitis and a bluish submucosal mass in the distal oesophagus. Biopsies showed no evidence of neoplasia. A barium swallow showed a 3 cm well defined mass arising from the distal oesophagus. Computed tomography

of the mediastinum confirmed the presence of a well defined mass arising from the wall of the distal oesophagus. There was homogeneous but no ring enhancement of the lesion with intravenous contrast. The appearances were compatible with an oesophageal leiomyoma. The patient is being followed up and is well 18 months later.

Case 4

A 59 year old man presented to the gastroenterology clinic in April 1993 with a four year history of shortness of breath and epigastric discomfort. There was no history of chest pain, heartburn or dysphagia. Examination and investigation showed no evidence of cardiopulmonary disease but an upper gastrointestinal endoscopy showed two well defined submucosal masses arising in the distal oesophagus. Histological assessment showed no evidence of neoplasia. A barium swallow also showed the lesions. Computed tomography of the mediastinum showed a small well defined dumb bell lesion arising from the lower oesophagus and displacing the lumen laterally. There was homogeneous but no ring enhancement with intravenous contrast. The patient is well 12 months later.

Case 5

A 59 year old Indian man was admitted under a non-gastroenterological team in November 1993 with a three day history of chest pain radiating to the epigastrium. He also complained of retrosternal burning and regurgitation. There was no past history of ischaemic heart disease. Examination and investigations showed no evidence of cardiopulmonary disease. A barium swallow showed a 3 cm well defined intramural lesion with an intact mucosa in the upper third of the oesophagus. Computed tomography of the mediastinum before and after contrast showed a well defined solid lesion with homogeneous enhancement confined to the wall of the oesophagus. There was no ring enhancement. A diagnosis of leiomyoma was made and an upper gastrointestinal endoscopy was performed, which confirmed the radiological findings; biopsies of the lesion were non-specific with no evidence of neoplasia. The patient's doctors advised surgery at which a 3 cm spherical lesion was removed and histological examination confirmed the diagnosis.

Radiology

In each patient, the barium swallow examination showed the typical appearance of an oesophageal leiomyoma; a smooth eccentric well defined submucosal lesion with no evidence of mucosal distortion or damage (Fig 1).

Computed tomography appearances were similar in each patient. A well defined eccentric intramucosal mass was seen, which had homogeneous enhancement after administration of intravenous contrast. There was no ring enhancement, which strongly suggested that

Figure 2: Computed tomography shows well defined mass arising from the wall of oesophagus displacing the lumen laterally (arrows). No involvement of surrounding structures (case 4).

the lesion was not arising from the mucosal layers. The lumen of the oesophagus was displaced laterally rather than showing concentric narrowing or obliteration, which would be characteristic of a malignant tumour. There was also preservation of the mediastinal fat planes. Two typical computed tomograms are shown (Figs 2 and 3).

Discussion

We have reported five cases of leiomyoma of the oesophagus; four patients were male and one was female. Their age ranged from 46–61 years (mean 55). Four presented with epigastric pain and one with nausea and vomiting. None has dysphagia. In four the diagnosis was suspected at upper gastrointestinal endoscopy and confirmed on barium swallow and computed tomography. In the other case the diagnosis was suspected on barium swallow and confirmed at computed tomography and upper gastrointestinal endoscopy. All the patients had endoscopic biopsies and none were diagnostic of leiomyoma. One patient had surgical excision and

Figure 3: Post contrast computed tomography shows well defined enhancing eccentric mass arising from wall of oesophagus, displacing lumen laterally (arrows) (case 5).

histological examination confirmed the diagnosis. Four patients have been followed up for one to three years; all are well and two have been followed up with repeat upper gastrointestinal endoscopy and computed tomography at 2.5 and three years. The appearances at endoscopy and computed tomography did not change.

Leiomyoma of the oesophagus is an unusual condition that arises from the smooth muscle in the wall of the oesophagus, the muscularis mucosae or the smooth muscle in the blood vessels contained in the oesophageal wall.⁶ Over 90% of oesophageal leiomyomas are reported to occur in the lower or middle third and only occasionally in the upper third.^{3,4} There has been reported a male preponderance in this condition³ and our study supports this. The age range of patients reported in published works is between 12 and 80 years, with a mean of 44.³ Most of the lesions are intramural or submucosal and solitary, but multiple lesions are seen in 3–4% of patients.^{3,7,8} Tumours are very variable in size, the largest reported weighed 1000 g but most are 2–8 cm in diameter.⁹ The tumours are thought to be slow growing and only when they are strategically positioned do they cause significant symptoms. It is estimated that roughly half of patients with oesophageal leiomyoma are asymptomatic.³ The most frequent symptoms in order of frequency are dysphagia, which is intermittent and gradually worsening, pain or discomfort, which is mainly retrosternal but sometimes over the left chest, heartburn, and weight loss.^{3–5} The symptoms are mostly of long duration, over two years in 70% of cases.³ Calcification in oesophageal leiomyoma is rare and has been seen in an estimated 1.8% of all cases reported.³

Published works report a very high rate of surgical resection. It used to be thought that the presence of the leiomyoma was indication for surgical removal especially as there is a risk of sarcomatous transformation.^{3,10,11} As it is now realised, however, that most cases are asymptomatic and the risk of malignant transformation is very low, probably less than 1%,³ surgical intervention is less frequently indicated. However, the medical treatment of this condition has not been reported in published works. In this study for the first time it has been shown that conservative treatment of oesophageal leiomyoma is safe and does not seem to affect outcome in cases that do not have significant symptoms directly related to the tumour. Surgical excision should be reserved for those with significant symptoms directly related to their tumour; troublesome dysphagia would be the most common indication for surgery. None of our cases had dysphagia. Our fifth case, who was not assessed by a gastroenterological physician almost certainly did not require surgery as his symptoms were of three days duration and the appearance of his leiomyoma at endoscopy and radiology was no different to the others. Review of our other cases illustrate the comparative lack of symptoms experienced by the patients and highlights the slow growth of oesophageal leiomyoma. These points confirm the view that surgery is only necessary in a few patients.

The diagnosis is usually first suspected at routine upper gastrointestinal endoscopy. The appearance of a submucosal mass lesion raises the possibility of leiomyoma.¹² The mucosa is almost always intact, which contrasts with gastric leiomyomas in which the mucosa often ulcerates in the acid environment and can cause significant upper gastrointestinal blood loss.⁵ The fact that the mucosa is intact accounts for the failure of endoscopic pinch biopsy specimens to achieve a conclusive histological diagnosis.⁶

The diagnosis is further supported by the characteristic barium swallow appearances of a well defined submucosal lesion arising from the wall of the oesophagus with a smooth margin and intact overlying mucosa.^{14 12 13} The appearances in our cases are compatible with the descriptions in published reports.

Computed tomography of the mediastinum with and without intravenous contrast allows the oesophagus and surrounding structures to be examined accurately. The presence of the tumour can be confirmed and the absence of infiltration of the mediastinal fat planes can be seen helping in the differentiation from oesophageal malignancies. The use of computed tomography in the diagnosis of oesophageal leiomyoma has been suggested for difficult cases but has not been reported previously and computed tomograms in all our five cases had the same characteristics. We believe that the computed tomogram appearance of a well defined eccentric intramural mass, which shows homogeneous enhancement after administration of intravenous contrast with no ring enhancement suggesting that the lesion is not arising from the mucosal layers of the oesophagus, strongly suggests the diagnosis of oesophageal leiomyoma. The displacement of the oesophageal lumen to one side rather than causing concentric narrowing or obliteration of the oesophageal lumen with preservation of the mediastinal fat plains help to differentiate benign leiomyoma from malignant oesophageal neoplasms.

The new technique of endoscopic ultrasound seems to have potential value in the diagnosis of oesophageal leiomyoma and the exclusion of malignancy.¹⁴ However, this technique is not widely available at present.

We believe that the combination of direct vision via the endoscope and computed tomography allows the diagnosis of oesophageal leiomyoma to be made confidently. Endoscopic examination has two important roles in the diagnosis. The first is to exclude other oesophageal, gastric or duodenal disorders as

leiomyomas are so frequently asymptomatic. The second is that endoscopic biopsy specimens can be taken to exclude mucosal malignancy. The diagnosis in all but one of our cases was made without histological confirmation. In cases where histological confirmation is absent it seems prudent to follow up the tumour over a prolonged period time especially with the recognised risk of sarcomatous transformation. Our patients have been followed up for between one to four years. We have seen no change in the endoscopic or computed tomogram appearances of the leiomyomas over this period. Our policy is that patients are reviewed every two to three years with both endoscopy with biopsy and computed tomography. These investigations would be expedited if the patient was experiencing any symptoms that could be attributable to their leiomyoma.

We conclude that the combination of endoscopy with biopsy and computed tomography of the mediastinum with and without intravenous contrast is the ideal method of making the diagnosis of oesophageal leiomyoma and also useful in following up the course of this unusual tumour. We also suggest medical observation in most cases and would only advocate surgical excision in those patients experiencing dysphagia or other significant symptoms definitely attributable to their leiomyoma.

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