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Giant oesophageal hamartoma

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Benign tumours of the oesophagus are rare, representing less than 1% of all oesophageal neoplasms.[1] Most of them are intramural leiomyomas and other benign tumours encountered only infrequently; among these pedunculated intraluminal hamartomas form a particularly rare group. Special difficulties are associated with their diagnosis, but early detection is important because despite their benign histological nature life threatening sequelae may ensue. We present the case history of a patient with a protracted illness due to a large oesophageal hamartoma, which had remained undiagnosed despite multiple endoscopies and biopsy.

Case report

A 52 year old non-smoker presented in late 1979 with a dry cough and mild but progressive dysphagia. Barium studies showed a megaoesophagus with large, lobulated filling defects. At oesophagoscopy grossly dilated mucosal folds were visualised within the dilated oesophagus, biopsy of which showed only chronic inflammatory changes. The differential diagnosis included achalasia, oesophagitis with polyp formation, and giant mucosal folds of the oesophagus. The patient was treated symptomatically until November 1982, when he presented again with rapidly progressive dysphagia. The results of the investigations were as before; the condition was thought to be inflammatory and the patient was treated with a brief course of prednisolone, which produced a transient but substantial symptomatic improvement.

The patient was readmitted five months later with an episode of complete dysphagia. On admission he was malnourished, hypoalbuminaemic, and anaemic with an iron deficiency picture. Nasoenteric feeding was started and he transferred to the Brompton Hospital. A plain posteroanterior chest radiograph showed a grossly dilated oesophagus with a shelf like filling defect in the upper third. Barium studies (fig 1) showed a megaoesophagus with large, lobulated filling defects. The findings at oesophagoscopy were unchanged. The patient, requiring urgent relief of his complete dysphagia, proceeded to thoracotomy. The approach was made initially through the left chest and a long oesophagotomy was made at the level of the inferior pulmonary vein before the true nature of the lesion was appreciated. This allowed delivery of the apex of a single large polypoid lesion. We decided at this stage to proceed to formal oesophagectomy as primary closure of the compressed and dilated oesophagus was likely to be hazardous. The stomach was mobilised and the thoracotomy closed, the patient was turned, and a right thoracotomy was performed. A mobile mass could be felt

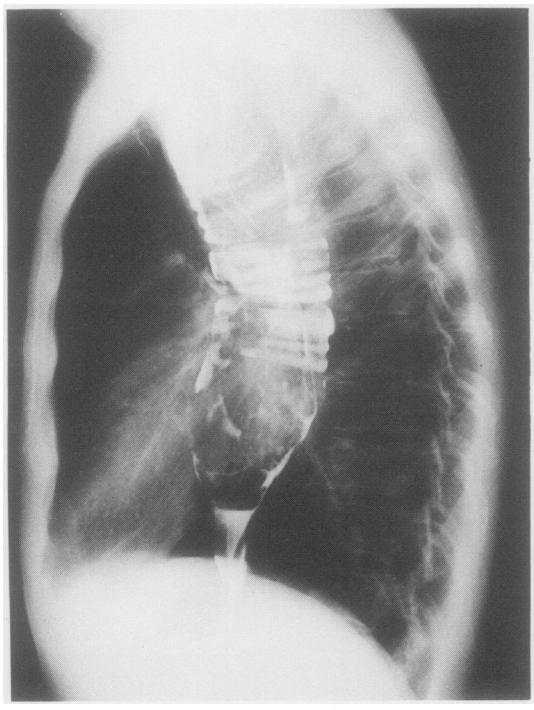

Fig 1 *Lateral view of barium swallow showing an extensive lobulated mass lying within the dilated oesophagus.*

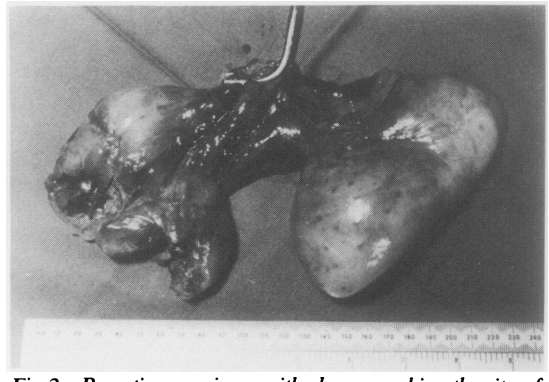

Fig 2 *Resection specimen with clamp marking the site of the tumour pedicle.*

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in the oesophagus with a discrete pedicle at the level of the thoracic inlet. The oesophagus was opened at the apex of the pleura, the mass delivered, and the pedicle divided. Gastro-oesophageal anastomosis was performed. After closure of the thoracotomy the cervical oesophagus was explored through the left neck. The pedicle of the tumour was found to originate at the level of the cricopharyngeus anteriorly. The base of the pedicle was excised and oversewn. The patient made an uneventful recovery; he now has no dysphagia and has gained 19 kg in weight. Histological examination showed a single polypoid oesophageal hamartoma 20 × 14 × 7 cm (fig 2).

Discussion

Pedunculated oesophageal hamartomas are not true neoplasms, but represent developmental lesions whose origin is obscure—possibly they derive from rests of embryonic tissue associated with development of the foregut and the tracheobronchial groove. Typically they are pedunculated, and the vast majority arise in the upper third of the oesophagus at the level of the cricoid cartilage.[2] The presentation is extremely varied, occurring both during infancy[3 4] and in adult life.[5 6] The slow growing, benign nature of the lesion often allows them to reach very considerable sizes before presentation and masses of up to 25 cm have been described.[5] In adult life typically there is progressive dysphagia with concomitant weight loss associated with a hypochromic, microcytic anaemia resulting from chronic blood loss from the ulcerated mucosal surface. The episodic sloughing of this epithelium may be responsible for the episodes of spontaneous remission that are sometimes seen. Occasionally, however, presentation is dramatic and life threatening with acute airway obstruction from regurgitation of the mass.[6]

Establishing the correct diagnosis is frequently very difficult as the lesion is covered by inflamed, ulcerated oesophageal mucosa, biopsy of which serves to disguise the histological nature of the underlying tissue. Typically, as in our own case, multiple endoscopies and biopsy are performed before the correct diagnosis is established, which is often at the time of histological examination of the resected specimen. In our case this difficulty was compounded as the site of the pedicle was not apparent, as this was compressed by the underlying mass and the "folded" lobes of the mass gave the appearance of mucosal folds.

In contrast to the difficulty of establishing the diagnosis, surgery for these lesions is relatively simple and is followed by dramatic and permanent relief of symptoms. The typical site of the stalk at the level of the cricoid cartilage allows removal of all but the largest tumours via a cervical oesophagotomy[4 6] and occasionally, if the tumour is small, by snare excision at oesophagoscopy. Very large masses may require thoracotomy for excision, as in our own patient. Oesophageal hamartomas represent a rare, benign, but potentially fatal condition. An increased clinical awareness of these lesions will allow earlier diagnosis and facilitate curative surgery.

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