A leiomyosarcoma of the oesophagus presenting incidentally without dysphagia

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Abstract

Leiomyosarcoma is a rare tumour that accounts for 0.5% oesophageal sarcomas. The most common presenting symptom is dysphagia. This report presents a case of oesophageal leiomyosarcoma in a 56-year-old Caucasian man found incidentally while being investigated for refractory cough. There was no history of dysphagia in spite of tumour mass occupying most of the oesophageal lumen. The leiomyosarcoma was managed successfully with surgical resection and adjuvant radiotherapy. The patient remains disease free after 15 months after surgical intervention. The unusual case presentation is discussed and the surgical management of this rare condition reviewed. © 2002 Published by Elsevier Science B.V.

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1. Case report

A 56-year-old Caucasian man presented to his general practitioner with a 4-week history of a non-productive cough. He had been a non-smoker for over 40 years. No relief was gained with proprietary treatments and he was referred to a respiratory physician for further assessment. Chest radiograph revealed a right superior para-mediastinal lesion ‘not typically bronchogenic in nature’ (Fig. 1). On presentation he was slightly obese with no lymphadenopathy, clubbing or stridor. There was no history of gastrointestinal symptoms including dysphagia. Examination of his chest and abdomen was unremarkable. Lateral chest radiograph showed the mass to lie in the posterior mediastinum. Computed tomography revealed a lesion relating to the mid-oesophagus at the level of the aortic arch and carina. The mass was described as being ‘slightly endophytic and encroaching into the lumen and measuring 6.5 cm in diameter’. At oesophagoscopy an exophytic and macroscopically malignant looking tumour was confirmed in the mid-oesophagus at 23–29 cm. Multiple biopsies were taken which later showed evidence of malignant cells but of no specific type. Bronchoscopy during the same anaesthetic was normal while abdominal ultrasound scan and radioisotope bone scan showed no evidence of metastatic disease.

The patient proceeded to an Ivor-Lewis esophagectomy where intra-operatively tumour was confirmed with no evidence of local invasion. Bleeding from the oesophageal bed requiring re-exploration complicated post-operative recovery but he subsequently made a full recovery and was discharged on the 11th post-operative day. Pathology revealed a 36 × 20 × 20 mm tumour with the entire oesophageal wall replaced by malignancy. The tumour also contained a few small areas of necrosis. Microscopy demonstrated an epithelioid smooth muscle tumour forming polypoid projections into the oesophageal lumen as well as infiltrating the oesophageal wall and peri-oesophageal tissues. Immunocytochemistry revealed the specimen to be strongly positive for smooth muscle actin (Fig. 2). The specimen was negative for desmin and cytokeratin antibodies. The surgical resection margins were clear and all biopsied lymph nodes were free of metastases (Stage T1N0). A diagnosis of leiomyosarcoma was therefore made. In light of these findings the original biopsy specimens were reviewed and found to confirm the diagnosis of the malignancy. The tumour was classified as low grade with a mitotic count of less than five/ten per high power field. Surgical resection was followed by adjuvant radiotherapy to the oesophageal bed. At the time of submission of this paper...
the patient remains recurrence free at 15 months having required dilatation of an anastomotic stricture for dysphagia on two occasions.

2. Discussion

Leiomyosarcomas of the oesophagus are rare tumours first described almost 100 years ago [1]. They are malignant tumours that originate from smooth muscle cells that may be difficult to differentiate from the benign tumours leiomyomas, especially if they are well-differentiated. The tumours typically have a slow rate of growth that may further lure the surgeon into a false diagnosis. Leiomyosarcomas account for 5% of gastrointestinal and 0.5% of all oesophageal sarcomas. In most instances dysphagia and odynophagia are the commonest presenting symptoms [2]. We know of only one case in medical literature that presented without dysphagia [3]. Furthermore, there are no reports in the English literature of such tumours presenting as incidental findings on chest radiographs during routine investigation.

In our case history the absence of weight loss and other constitutional symptoms did not favour a diagnosis of malignancy. Although recurrent cough is usually suggestive of tracheal invasion in oesophageal malignancy, we excluded this in our case with pre-operative bronchoscopy and CT scanning. In the absence of local invasion on histopathology of the resected specimen the patient was considered to have had curative surgery. Since the first case of surgical resection by Harrington in 1945 [4] the results for surgery of oesophageal leiomyosarcomas have been encouraging such that most groups now recommend oesophagectomy as the standard treatment for most patients [5,6]. The Mayo Clinic has demonstrated 5-year survival rates of almost 80% with no significant long-term advantage offered by adjuvant chemotherapy and, or radiotherapy. The study also showed that the tumour usually spreads to the liver or lung, which shortens survival rate. However, the presence of metastases at presentation should not preclude surgical intervention for palliation as there have been reports of success [7]. Polypoid lesions also tend to have a more favorable prognosis than infiltrating or intramural lesions by almost 60% at 5 years [6–8]. Similarly, as would be expected, patients with well-differentiated tumours had improved survival rates than poorly differentiated sarcomas [6–9].

In conclusion, we report a case of leiomyosarcoma of the oesophagus presenting unusually without dysphagia but revealed incidentally on chest radiograph. As this rare tumour of the oesophagus is potentially curable, early diagnosis and proper staging procedures are mandatory as respectable survival rates with low morbidity and mortality are possible with tumour resection, especially if the tumour is well-differentiated and polypoidal.

References


