

Rare cancer of the oesophagus - leiomyosarcoma

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Introduction

Leiomyosarcoma of the oesophagus is a rare, malignant tumour of smooth muscle origin. The reported incidence is less than 1% of all oesophageal malignancies [1].

Case presentation

A 62-year-old female presented with progressive dysphagia for solid diet for the four-month duration. Her clinical examination was unremarkable. Upper GI endoscopy revealed an extra mucosal bulge with normal overlying mucosa in the oesophagus at 29cm from incisor teeth (figure-1) and benign-looking small pre-pyloric peptic ulcer. Endoscopic ultrasound scan (EUS) was done and it shows a hyper-echoic lesion arising from the muscularis propria of the oesophagus with no surrounding invasion, most likely to be a leiomyoma. Contrast-enhanced tomography of chest (Figure-2) reveals a mid-oesophageal, well-defined lesion, further in favour of a benign leiomyoma.

An oesophageal gastrointestinal stromal tumour (GIST), neurofibroma and fibrovascular polyp were the other differential diagnosis. With the background of a benign tumour, the patient underwent a thoracoscopic enucleation of the tumour. The patient had an uneventful recovery postoperatively. Histology reveals a leiomyosarcoma of the oesophagus (Smooth muscle actin & Desmin positive immunohistochemistry with high mitotic count) and after discussing at a multi-disciplinary oncology meeting, the patient underwent thoraco-laparoscopic oesophagectomy. Early postoperative period was complicated with gram-negative septicaemia and patient develop an enterocutaneous fistula after two weeks of surgery, which is confirmed by contrast studies.

Subsequent upper GI endoscopy revealed a perforated peptic ulcer along the lesser curve of the stomach tube, with healed, intact gastro-oesophageal anastomosis. Several attempts of



Figure 1. Endoscopy picture

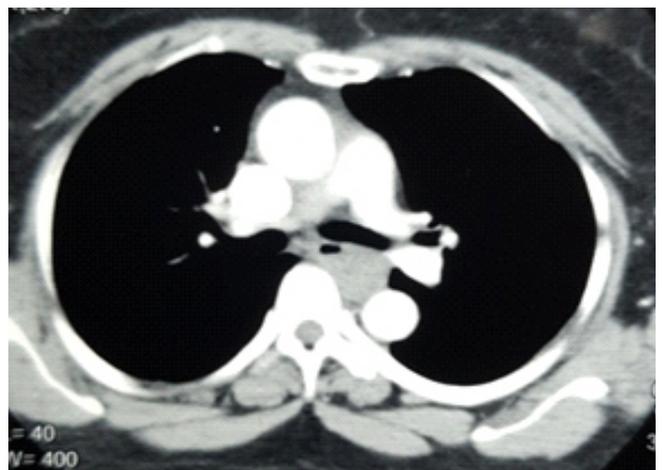


Figure 2. CT image of the tumour

'over the scope clipping' of the perforated site failed and the patient died due to myocardial infarction and multi-organ failure by the third week from surgery.

Discussion

Dysphagia is the common clinical presentation in patients suffering from an oesophageal leiomyosarcoma [1]. Upper gastrointestinal endoscopy (UGIE) is the investigation of

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choice to evaluate dysphagia, which might show an extra mucosal bulge as in this case. An endoscopic luminal biopsy may give a high false-negative rate especially when overlying mucosa remain intact [5]. Preoperatively, imaging studies should be done with an endoscopic ultrasound scan (EUS) with or without MRI to see the tissue plane of origin, depth of invasion and possible diagnosis. Contrast-enhanced CT chest and abdomen is indicated if any suspected malignant lesion. Even though with the imaging, it's very difficult to differentiate between leiomyoma and leiomyosarcoma of the oesophagus [2]. Preoperative misdiagnosis rates are as high as 82% in some case series (of the reported literature). Most of the cases were ultimately diagnosed by an oesophageal open biopsy and immunohistochemical analysis [4].

Recently endoscopic ultrasound-guided fine-needle biopsy (EUS-FNB), using 19 G Franseen tip needle and immunohistochemistry looking for smooth muscle actin (SMA), desmin is an accurate method for diagnosing leiomyosarcoma from other oesophageal tumours pre-operatively and may be used to guide treatment precisely [4]. The current treatment of choice for leiomyosarcoma of the oesophagus is radical oesophagectomy [5]. Prognosis of leiomyosarcoma is comparatively superior to squamous cell carcinoma of the oesophagus if resected completely at surgery [3]. The place for adjuvant radiotherapy and chemotherapy is controversial [5]. Even though leiomyosarcoma has poor sensitivity to radiation, the tumour can be effectively controlled by increasing the radiation dose appropriately, which provide acceptable survival rates in patients who are unfit for surgery and metastatic disease [4].

In this case report the patient had dysphagia with extra mucosal tumour of oesophagus found in endoscopy, she was subjected to imaging studies with EUS, which revealed a lesion in muscularis propria, but was not subjected to EUS-FNB to confirm the diagnosis at the early stage and because of

that she was underwent two major surgeries subsequently with lethal postoperative complications, which has lead into end of her life.

Conclusion

Leiomyosarcoma of the oesophagus is a rare malignant tumour. EUS-FNB is a gold standard investigation, which helps to diagnose at the initial stage. At current practice, radical oesophagectomy is the treatment of choice. High dose radiotherapy can be reserved for patients with advanced cancer and who are unfit for surgery.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Early diagnosis of leiomyosarcoma of oesophagus can be achieved by EUS-FNB, which is the gold standard investigation in current practice.
- Radical oesophagectomy is the treatment of choice with good prognostic outcome than other common oesophageal malignancies.
- There is a place for high dose radiotherapy in patients with advanced tumor and who are unfit for surgery.