CASE REPORT

A RARE OESOPHAGEAL TUMOUR

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ABSTRACT: Inflammatory myofibroblastic tumours are regarded as intermediate-grade tumours with a potential for recurrence. Although these lesions have been found in nearly every anatomic location, there are few documented cases of oesophageal localization. Here reporting this rare case concerns a 28-year-old male with an extremely large inflammatory myofibroblastic tumour of the upper & middle oesophagus; the tumour was 18 cm in length and 6 cm in largest diameter. Surgical excision with subtotal oesophagectomy was performed. Histopathological and immunohistochemical analyses confirmed the diagnosis of an inflammatory myofibroblastic tumour.

KEY WORDS: inflammatory myofibroblastic tumour ; esophagus

INTRODUCTION: Inflammatory myofibroblastic tumours (IMT) were classified as intermediate-grade tumours by the World Health Organization in 2002. These tumours have a potential for local recurrence and a risk of metastasis. IMT is described as a distinctive lesion composed of myofibroblastic spindle cells accompanied by inflammatory infiltration of plasma cells, lymphocytes, and eosinophils. It is most commonly found in lung, abdomen, retroperitoneum, and extremities, but is extremely rare in the oesophagus. Complete surgical resection is essential for treatment, and there is no need for postoperative radiation or chemotherapy or corticosteroid treatment. Periodic follow-up should be undertaken for early detection in case of recurrence or metastasis.

CASE REPORT: A 28-year-old male experienced dysphagia to hard solids for 1 year, loss of weight during the course of last 6 months and experienced chest discomfort in substernal region and an intermittent fever. Other than these symptoms, his medical history was unremarkable.

Laboratory analysis upon admission to the hospital was: white-cell count, $8.43 \times 10^9/L$; neutrophil count, $7.38 \times 10^9/L$; lymphocyte count, $0.61 \times 10^9/L$; erythrocyte count, $2.99 \times 10^{12}/L$, and the haemoglobin level was 8.1 g/dL, which suggested moderate anaemia. The hemoculture was negative, and the erythrocyte sedimentation rate (ESR) was 40 mm/h.

Upper GI endoscopy revealed oesophageal mucosa normal with extrinsic compression in upper 1/3rd and middle 1/3rd of oesophagus (fig 1).

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A computed tomography (CT) scan of thorax demonstrated that -Non enhancing soft tissue attenuation lesion involving the left lateral wall of oesophagus forming a mass causing deviation of oesophagus to the right. No local invasion(fig 2).

Open biopsy from Neck: F/S/O Inflammatory Myofibroblastic Pseudotumour.

Surgical exploration was undertaken through a Rt Posterolateral Thoracotomy. After opening, there was a huge dissociated columned mass locally attached to the posterolateral wall of the middle third of the oesophagus(fig 3). Lower normal oesophagus was just seen above the hiatus, Otherwise the oesophagus was involved in the tumor. We enucleated the mass from the oesophagus except about two inches of oesophagus was very much involved in the tumor so we landed up in doing subtotal oesophagectomy & gastric tube with gastroesophageal anastomosis in the neck on left side.

The postoperative pathology revealed that the lesion was composed of spindle-shaped cells, and the inflammatory infiltration was composed predominantly of lymphocytes and plasma cells, with few eosinophils. Immunohistochemistry revealed smooth muscle actin and vimentin but no CD117, CD34, S100, anaplastic lymphoma kinase (ALK), or desmin. From these findings, the mass was diagnosed as an IMT.

The patient's fever subsided 3 days after the operation. At 4 days, his white-cell count was $8.82 \times 10^9/L$, the neutrophil count was $6.91 \times 10^9/L$, lymphocytes were $1.11 \times 10^9/L$, the erythrocyte count was $3.94 \times 10^{12}/L$, and haemoglobin was elevated to 10.8 g/dL. No blood transfusion had been given. No recurrence or tumor metastasis was observed at the examination 6 months after the operation.

COMMENT: IMTs are regarded as intermediate-grade tumours with potential for recurrence. These tumours are most commonly found in the lung, abdomen, retroperitoneum, and extremities, whereas occurrence in the oesophagus is extremely rare. The most frequent symptoms of IMTs of the oesophagus are dysphagia and substernal pain, which are usually caused by the tumor invasion of adjacent structures. Unlike other reported cases of oesophageal IMTs, this patient had irregular high fever, anemia, weight loss, and an elevated ESR. Blood count analysis and hemoculture provided no clues to the cause of these symptoms; in fact, the fever was suggestive of infection. The fever disappeared 3 days after the tumor resection, which suggests that the tumor may have secreted cytokines that caused the patient's nonspecific symptoms.

IMTs of the oesophagus generally appear as small nodules or circumscribed masses and are frequently associated with mucosal ulceration. The lesion in the patient in study was different: it was a huge, dissociated, columned mass covered with normal mucosa and locally attached with a stem to the posterior wall of the superior segment of the oesophagus. Although IMTs come in various shapes and sizes, such a huge mass in oesophagus is highly unusual.

Evaluation of tumor pathology is important for the correct diagnosis. The IMT was described as a distinctive lesion composed of myofibroblastic spindle cells accompanied by inflammatory infiltration of plasma cells, lymphocytes, and eosinophils. Immunohistochemistry shows cells are positive for vimentin, smooth muscle actin, and muscle-specific actin and are negative for myogenin, myoglobin, CD34, CD117 (cKit), and S100. More than half of IMTs are also immunoreactive for ALK protein. Although ALK reactivity is not specific to IMT, it appears to be a factor associated with metastasis and recurrence.

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This patient's lesion was composed of spindle-shaped cells, and the inflammatory infiltration was composed predominantly of lymphocytes with a few neutrophil, plasma cells, and eosinophils. Some spindle-shaped cells had prominent clumped chromatin and nucleoli with occasional mitotic figures. The tumor cells were positive for smooth muscle actin and vimentin, but not CD117, CD34, S100, ALK, or desmin. IMTs that do not express ALK are usually associated with older patients. ALK expression is also associated with subtle histologic differences, local recurrence, and death from disease. A follow-up of this patient should provide additional information about the relationship between the ALK and prognosis of patients with IMT.

IMTs had been regarded as benign neoplasm; however, a retrospective study by Fabre and colleagues showed some pulmonary IMTs resulted in local invasion, distant metastasis, local recurrence, and sarcomatous degeneration. Oesophageal IMTs have not been associated with metastasis, however. Surgical resection is generally regarded as the most effective treatment for oesophageal IMTs. Radical resection and adjuvant therapy (radiotherapy or chemotherapy) are reserved for IMTs with an aggressive biologic behaviour. Periodic follow-up is very important for early detection of recurrence or metastasis.

In conclusion, oesophageal IMT is extremely rare and varies greatly in terms of clinical features. Pathology combined with immunohistochemistry is exceeding valuable in diagnosis. Complete surgical resection and periodic follow-up are essential for the treatment of tumor.

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Fig 1 showing endoscopy photograph showing external compression

Fig 2 CECT thorax showing large esophageal tumour

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Fig 3 intra-operative photograph showing the esophageal tumour and white arrow showing collapsed lung