A rare oesophageal inflammatory myofibroblastic tumour treated by endoscopy

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ABSTRACT

An inflammatory myofibroblastic tumour is a mesenchymal neoplasm that mostly involves the lung and rarely involves the oesophagus. Surgery has been most commonly used for the treatment of oesophageal inflammatory myofibroblastic tumours but there are no definite guidelines for their diagnosis and treatment. We describe the case of a 60-year-old woman presenting with dysphagia and poor appetite who was diagnosed with a submucosal oesophageal tumour by contrast enhanced computed tomography and ultrasonography endoscopy. She was treated successfully by endoscopic submucosal dissection with no complications. The final diagnosis was confirmed by pathological examination.

KEYWORDS

Dysphagia – Endoscopic submucosal dissection – Endoscopy – Oesophageal disease – Inflammatory myofibroblastic tumour

Accepted 3 April 2017

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An inflammatory myofibroblastic tumour (IMT) is a mesenchymal neoplasm composed of myofibroblastic spindle cells with an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils.¹ Although the lung is the most common site involved, IMTs can occur in diverse extrapulmonary locations. Coffin *et al* reported 84 extrapulmonary cases; 49 of these IMTs were located in the abdomen (with 36 in the mesentery and omentum followed by 13 in the liver and porta hepatis, stomach, ileum, rectum and spleen).² However, none were in the oesophagus. IMTs typically appear to be benign. Nevertheless, recurrences and aggressive behaviour have been discovered before.¹ We report the case of a female patient who underwent successful endoscopic resection of an oesophageal IMT.

Case history

A 60-year-old female farmer without hypertension, diabetes or smoking history presented to us with the chief complaint of gradual onset progressive dysphagia for solid or dry food for one month. She had reduced appetite but no recorded weight loss. She had had no significant medical or surgical illness or intervention before coming to our hospital for treatment. General and systemic clinical examinations as well as haematological and biochemical investigations were normal.

Oesophageal enhanced computed tomography showed a high density, sharp edged, oval shadow at the middle of the oesophagus, with mild to moderate heterogeneous enhancement (Fig 1). Ultrasonography gastroscopy revealed

submucosal swelling, with a distinct border, allowing the possibility of angioma but not excluding carcinosarcoma (Fig 2). The patient underwent endoscopic submucosal dissection (ESD) to clarify the diagnosis. During the operation, it became clear that the mass was adhered extensively to the muscularis propria. Part of the mass (3.5cm × 1.5cm in size) was resected for pathological examination. This revealed an oesophageal IMT with only a few cells immunopositive for smooth muscle actin and a Ki-67 index of 10%. A multidisciplinary team meeting attended by thoracic surgeons concluded that oesophagectomy was required but this was refused by the patient based on the surgical risk and postoperative morbidity. She was offered organ preserving endoscopic therapy and accepted.

Consequently, 36 days after the first attempt at ESD, the patient reattended for further ESD and subtotal resection of the residual oesophageal tumour. After submucosal injection of a mixture of glycerine fructose, epinephrine and indigo carmine into each border of the lesion, a DualKnife™ (Olympus, Tokyo, Japan) was used for the proximal mucosal precut. Partial separation was performed using an ITknife nano™ (Olympus) alternating with snare electric coagulation excision. The mass was resected and pulled out using a net bag. It was 4.0cm × 3.0cm × 3.0cm in size and was sent for pathological examination (Fig 3). Hot biopsy forceps and an argon plasma coagulator were used for haemostasis and wound management. There were no complications during the procedure.

Postoperatively, the patient was kept in the ward with an indwelling gastric tube, intravenous cefoperazone/

Figure 1 Oesophageal enhanced computed tomography of the mass (arrow). The mass was 2.3cm × 3.2cm in size, with a mostly uniform density and a clear border.

Figure 2 Ultrasonography gastroscopy showing the mass. The mass had a low and uniform echo, a clear border and originated from the submucosa. The cross-sectional area measured 2.57cm × 3.10cm.

sulbactam, esomeprazole, carbazochrome sodium sulphonate and intravenous fluid infusion. She was started on a liquid diet initially, and could later swallow both solid and liquid food without dysphagia. Microscopic examination of the mass revealed spindle tumour cells arranged like bundles in the inflammatory background (Fig 4). The cells were immunonegative for anaplastic lymphoma kinase (ALK),

Figure 3 The resected mass.

Figure 4 Microscopic examination of the resected mass. The nucleus of the tumour cells was vesicular and oval in shape, with a definite nucleolus.

S100, desmin, CD34, CD117, DOG1 and pancytokeratin. Only a few cells were immunopositive for Ki-67 and most cells were immunopositive for vimentin.

The patient has not returned to our hospital for further consultation. She was contacted by telephone two months after the resection and she stated that she was without discomfort. No further follow-up has been possible.

Discussion

IMTs in the oesophagus are extremely rare. According to a MEDLINE® search from 2001 to the present, only been 12 cases have been reported. Our patient presented clinically

with progressive dysphagia and poor appetite. There were no 'red flag' symptoms to suggest malignancy, nor were there specific symptoms that are pathognomonic for IMT. The imaging features of IMT resemble those of leiomyoma of the oesophagus. Endoscopic examination showed the lesion at the middle of the thoracic portion of the oesophagus. There was no evidence of infiltration or extension around the lesion.

Histologically, IMTs are characterised by spindle cell proliferation in a myxoid to collagenous stroma with prominent plasma cell and lymphocyte infiltration.⁵ More than 50% of IMTs are immunopositive for ALK protein. Although ALK reactivity is not specific to IMTs, it appears to be a factor associated with metastasis and recurrence.¹ Our patient may therefore have a better prognosis than most. Surgery has been the most common therapeutic option used for oesophageal IMTs. However, only one case has been reported previously for endoscopic resection, using the conventional technique for polypectomy at the mucosal level.¹ In contrast, in our case, the resection was performed at the level of the submucosa by ESD.

The major complications of ESD in the oesophagus are bleeding, perforation and stricture.⁴ In our patient, none of these were observed. Our measures to prevent complications included: a) intraoperative use of DualKnife™, ITknife nano™, hot biopsy forceps coagulation and argon plasma coagulation for prophylactic electrocautery as well as post-operative use of a proton pump inhibitor with a coagulant to prevent bleeding; b) ultrasonography gastroendoscopy to

clarify the infiltrated layer of the lesion and the use of intraoperative indigo carmine to identify the correct plane of dissection for a lower perforation rate; and c) endoscopic balloon dilation and administration of steroids considered if stricture were to occur following ESD.

Conclusions

Although oesophageal IMT is extremely rare, it should be considered as a differential diagnosis in patients with dysphagia and an endoscopic finding of a submucosal oesophageal tumour. The diagnosis is usually made postoperatively, after pathological examination of the resected mass. Surgery is the most common treatment for oesophageal IMTs but endoscopic resection is another possible approach given the high surgical risk. Owing to the potential for recurrence, long-term follow-up review should be undertaken.

References

1. Surabhi VR, Chua S, Patel RP *et al.* Inflammatory myofibroblastic tumors: current update. *Radiol Clin North Am* 2016; **54**: 553–563.
2. Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. *Am J Surg Pathol* 1995; **19**: 859–872.
3. Gleason BC, Hornick JL. Inflammatory myofibroblastic tumours: where are we now? *J Clin Pathol* 2008; **61**: 428–437.
4. Fernández-Esparrach G, Calderón A, de la Peña J *et al.* Endoscopic submucosal dissection. *Endoscopy* 2014; **46**: 361–370.