[症例報告]A pedunculated liposarcoma of the esophagus : A case report and literature review

A pedunculated liposarcoma of the esophagus: A case report and literature review

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ABSTRACT

Esophageal liposarcoma is extremely rare, and only 13 cases have been reported so far. We herein report a case of this disease which was successfully treated and also include a literature review. A 68-year-old man visited our hospital due to progressive dysphasia. An esophagography revealed an exophytic mass in the cervical esophagus. On endoscopy, the tumor showed subepithelial growth with a positive cushion sign. MRI revealed the tumor to be composed of two components separated by a septum. The tumor was completely resected through an esophagotomy. The glossy tumor was found to be a pedunculated submucosal tumor, which was histologically found to be a myxoid liposarcoma. The patient had an uneventful postoperative course with a normal esophageal function. Because of the pedunculated configuration and the low-grade malignant nature of these tumors, a tumor resection with a negative surgical margin through an esophagotomy may be the most appropriate treatment in these cases. *Ryukyu Med. J., 24(1) 27～32, 2005*

Key words: liposarcoma, esophagus, surgery

INTRODUCTION

Malignant mesenchymal tumors seldom develop in the esophagus. Among such tumors, esophageal liposarcoma (ELS) is so rare that little is known about its clinical and pathologic characteristics. An extensive literature search revealed that only thirteen cases of ELS have been so far reported[1-13]. We recently had a case of ELS which was successfully treated. We herein report the clinicopathologic features and clinical management of this case, and at the same time make a review literature of such cases. We also discuss the appropriate management of patients with ELS.

CASE REPORT

In October 2002, a 68-year-old man visited the Ryukyu University Hospital for an examination of his upper gastrointestinal tract because of progressive dysphagia on swallowing solid food. The patient first noticed this symptom in May 2001. A physical examination revealed an elastic mass palpable in the right lower neck, at the site of right thyroid. His laboratory data were all within normal limits, including the serum tumor marker concentrations and thyroid hormone levels. An upper gastrointestinal series revealed an exophytic mass measuring 5 cm in the proximal esophagus below the piriform fossa (Fig. 1). Esophageal endoscopy showed a protruding submucosal tumor narrowing the esophageal lumen at a level of 20 cm from the incisor. We could not determine whether or not the tumor had a stalk because the endoscopic view was not sufficiency clear. Endoscopically, the tumor was elastic soft with a positive cushion sign indicating that it was a lipomatous tumor[14]. Biopsy specimens taken from the tumor showed

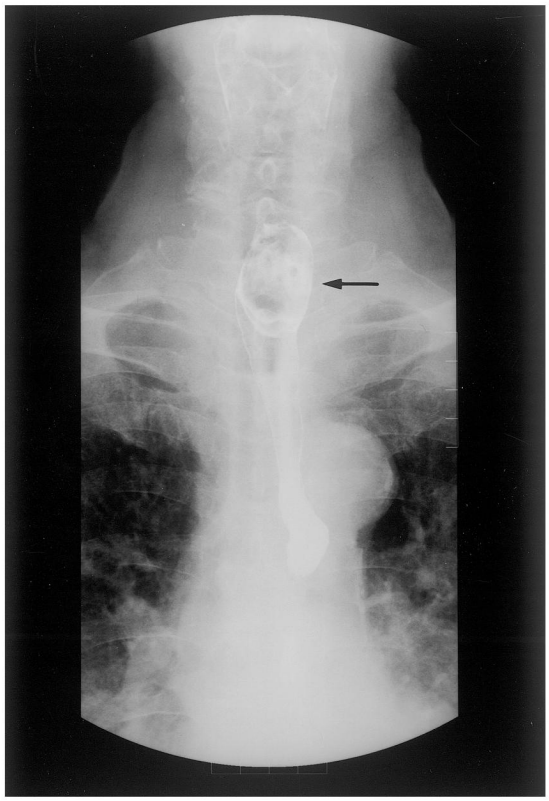

Fig. 1 Upper gastrointestinal series
An exophytic tumor occupying the cervical esophageal lumen was demonstrated (arrow).

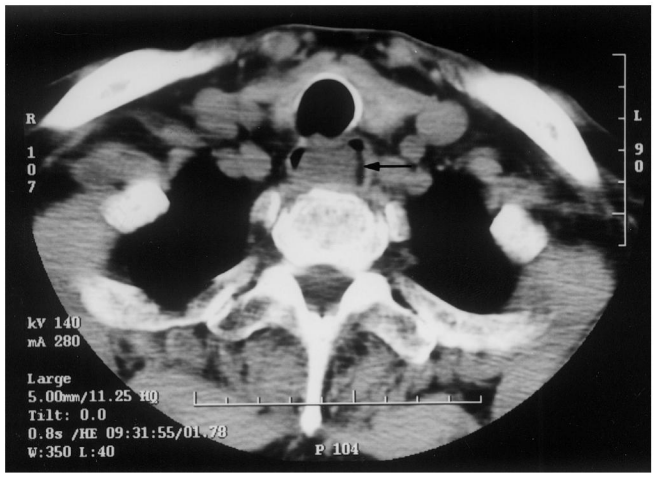

Fig. 2 Plain CT findings
A homogeneous low density mass was revealed in the cervical esophagus (arrow).

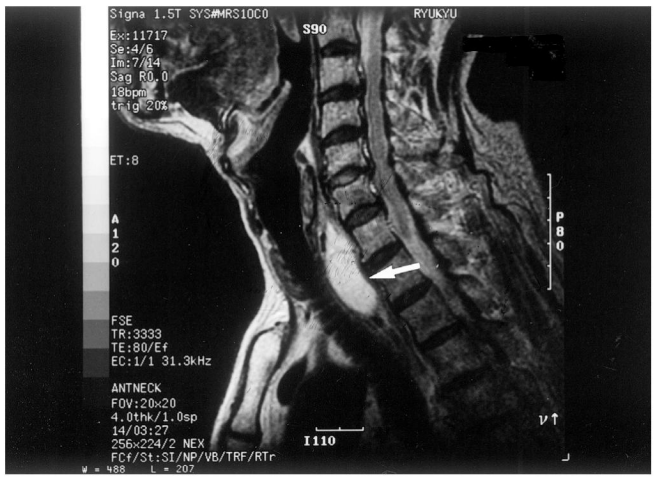

Fig. 3 MRI findings
MRI showed the tumor to have a septum and an increased signal on the T2-weighted images (arrow).

normal esophageal squamous epithelium with mild inflammatory changes. Plain computed tomography (CT) demonstrated a homogenous mass lesion occupying the cervical esophageal lumen (Fig. 2), and a calcified tumor in the right lobe of the thyroid gland. Percutaneous ultrasonography, but not endoscopical ultrasonography, showed the esophageal tumor to be cystic tumor having low echo with a septum formation. A Doppler echogram showed that the tumor had low vascularity. The calcified tumor in the right lobe of the thyroid gland was confirmed by ultrasonography. The esophageal tumor had a septum inside it and had an increased signal on T2-weighted magnetic resonance imaging (MRI), and a decreased signal on T1-weighted imaging (Fig. 3).

With a preliminary diagnosis of symptomatic myxomatous change or cystic tumor of the cervical esophagus and a right thyroid tumor, the patient underwent an operation in December 2002. During the operation, two hard tumors were found in the thyroid gland: one was located in the upper pole of the right lobe, and the other in the lower pole of the left lobe. A subtotal thyroidectomy was performed because frozen sections taken from the right upper tumor revealed an adenomatous tumor with a papillary structure. After removing the thyroid tumor, a longitudinal esophagotomy was performed on the right side of the cervical esophagus, and a pedunculated tumor prolapsed outside through the esophagotomy wound. On inspection, the tumor was completely covered with a normal esophageal epithelium and it was attached to the left wall of the cervical esophagus with a small stalk consisting of the normal mucosal and submucosal tissue of the esophagus. On palpation, the tumor was composed of two components: an elastic soft component

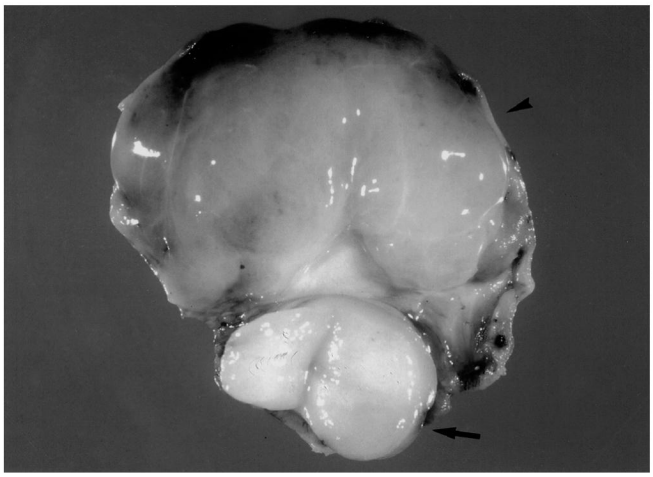

Fig. 4 Macroscopic findings of the resected specimen
The glossy tumor consisted of two components: a myxomatous component forming most of the projecting part (arrowhead), and the solid component forming the basal part of the tumor (arrow).

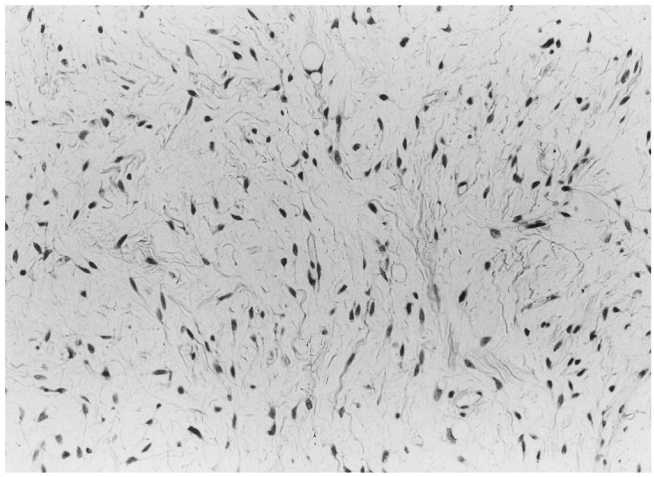

Fig. 5 Histological findings of the superficial portion
The superficial portion of the tumor showed a sparse distribution of spindle cells in an abundant amount of myxomatous tissue (Hematoxylin-Eosin staining, ×50).

forming most of the projected portion of the tumor, and an elastic hard component forming the basal part. The tumor was resected from the esophageal wall together with the esophageal mucosa lining, while obtaining sufficient surgical margin under direct vision. The patient had an uneventful postoperative course with normal swallowing function, and was discharged in January 2003.

Pathology: A cut surface examination of the tumor glossy showed a myxomatous component measuring 5 cm in diameter, separated by a fibrous septum from a yellowish white solid component measuring 1 cm in diameter (Fig. 4). The former component formed the superficial portion and the latter formed the basal portion of the tumor. Histologically, both components consisted of spindle cells with pleomorphism and fibrotic tissue intervening between the mature adipose cells. The superficial portion showed a sparse distribution of spindle cells in an abundant amount of myxomatous tissue (Fig. 5), whereas the basal portion had a dense population of these cells with fibrous tissue (Fig. 6). Based on these findings, the esophageal tumor was diagnosed to be a myxoid liposarcoma. There were no malignant cells at the resected margin of the cut specimen. The thyroid tumors were histologically determined to be follicular adenomas.

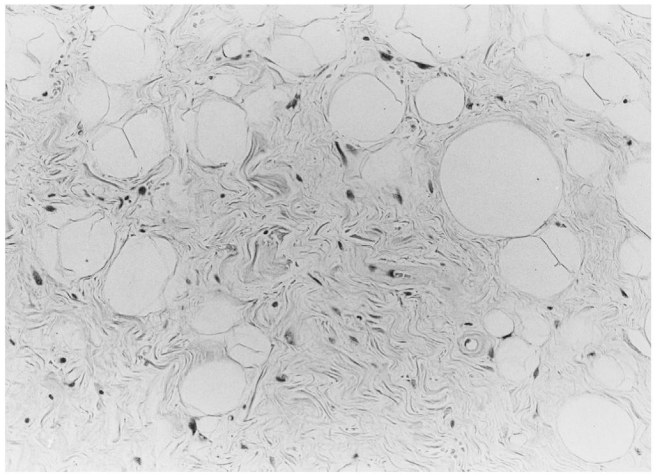

Fig. 6 Histological findings of the basal portion
The basal portion of the tumor consisted of pleomorphic spindle cells and mature adipose cells with fibrous tissue (Hematoxylin-Eosin staining, ×50).

DISCUSSION

The most common malignant mesenchymal tumor of the esophagus is leiomyosarcoma, although only 82 reported cases were found in a massive literature review[15]. Esophageal liposarcoma (ELS) is extremely rare, and there have been only thirteen cases reported so far in the English literature[1-13]. The present case is therefore the fourteenth reported case of ELS. The clinicopathological features of these cases are summarized in Table 1.

Table 1 Clinicopathological features of patients with esophageal liposarcoma

author	age	sex	chief complaint	site	macroscopic findings	size(cm)	histology	Tx	recurrence	TTR	follow-up
Mansour	53	M	dysphagia/sensation of a lump in the throat/cyanosis/cough/choking	CE	P	4	MX	TCE	N	−	NED 1y
Bak	49	F	dysphagia/weight loss/anemia	CE	P	20	WD	TE	N	−	NED 7m
Yates	49	M	dyaphagia/sensation of a lump in the throat/weight loss	CE	P	−	MX	TTE	Y	6.5y	DWD 8.5y
Baca	66	F	dysphagia/weight loss	CE	P	12	MX	TCE	N	−	NED 30m
Cooper	68	M	dysphagia	ThE	P	7	MX	SE	N	−	NED 1y
Boggi	50	M	dysphagia/sensation of a lump in the throat/anemia	CE	P	23	MX	TG/TO	N	−	NED 1y
Mandell	62	F	dysphagia	CE	P	7	WD	TCE	N	−	NED 4m
Temes	69	M	dysphagia/foreign body sensation	CE	P	12	WD	ER	N	−	NED 1m
Sails	73	M	dysphagia/dyspnea	CE	P	15	WD	TCE	N	−	NED 10m
Ruppert	72	F	mild dysphagia/weight loss/fatigue	CE	P	23	WD	ER	N	−	−
Beaudoin	68	F	dysphagia	CE	P	8.5	WD	TOR	N	−	−
Chung	56	M	dyaphagia/throat discomfort	CE	P	21	WD	TLE	N	−	−
Brehant	70	M	dysphagia/weight loss	ThE	P	20	WD	ER/TAE	N	−	NED 16m
Present	68	M	dysphagia	CE	P	6	MX	Ex	N	−	NED 13m

Tx: treatment, TTR: time to first recurrence, CE: cervical esophagus, P: pedunculated type, ThE: thoracic esophagus, WD: well-differentiated liposarcoma, MX: myxoid liposarcoma, TCE: trans-cervical esophagotomy, TTE: trans-thoracic esophagotomy, TE: total esophagotomy, SE: subtotal esophagotomy, TG/TO: combined transgastric/transoral resection, ER: endoscopic resection, TOR: trans-oral excision, TLE: total laryngopharyngoesophagectomy, TAE: trans-abdominal esophagotomy, Ex: tumor extirpation, Y: yes, N: no, NED: alive with no evidence of disease, DWD: dead of other causes with probable disease, y: year, m: month.

Grossly, all tumors show intraesophageal exophytic growth as a pedunculated submucosal tumor. Twelve (85.7%) of the 14 ELSs developed in the cervical esophagus.

ELSs have a tendency of not being detected until they grow into considerably large tumors thus causing disturbances in swallowing. The reported tumor sizes range from 4 to 23 cm in size (Table 1). These findings indicate that ELS may be difficult to detect at an early stage of tumor growth in which the patients have no symptoms. Furthermore, the anatomic characteristics of the cervical esophagus close to the upper esophageal sphincter where ELS commonly occurs may preclude an early detection on either endoscopy or esophagography. Generally, ELS is believed to be a slow growing tumor with a low-grade malignancy. However, Chung *et al* reported a case of ELS showing rapid increase in size from 9 to 21 cm in only 7 months[12]. The symptoms caused by ELS include dysphagia and the feeling of having a lump in the throat associated with weight loss and anemia (Table 1). However, some life-threatening symptoms such as cyanosis and asphyxia can also occur due to an acute airway obstruction by their movable pedunculated configurations and proximity to the larynx[1], thus suggesting the necessity of an urgent tumor resection.

Both CT and MRI have been reported to be useful in making a diagnosis of ELS[16], because tumors composed of adipose tissue usually appear as a low-density mass on CT and a high intensity area on the T2-weighted images of MRI. These characteristics of ELS were observed in the present case also. An endoscopic biopsy may be of no use in diagnosing ELS because ELS is usually covered with a normal esophageal epithelium. However, an assessment of the presence or absence of a cushion sign using biopsy forceps during endoscopy may be of adjunctive use for the diagnosis of ELS because the characteristic softness of lipomatous tumors can be demonstrated by this procedure.

Hajdu reported the occurrence of second primary neoplasms in 12% of liposarcoma patients[17]. In the present case, follicular adenoma of the thyroid gland was concomitantly associated with ELS. These findings may suggest the necessity of looking for a possible second primary tumor in patients with ELS.

According to the World Health Organization Classification of soft tissue tumors, liposarcomas are histologically classified into 4 subtypes: well-

differentiated, myxoid, pleomorphic and dedifferentiated liposarcoma[18]. Peterson JJ *et al* reported that well-differentiated, myxoid, pleomorphic and dedifferentiated types were found in 47.5%, 20.5%, 6.2%, and 8.9% of patients with liposarcoma, respectively[16]. The histological appearance of the present case was compatible with the diagnostic criteria of myxoid liposarcoma because of the presence of mature lipocytes, lipoblasts, and pleomorphic spindle cells with myxomatous substance.

Most ELS are considered to have low-grade malignancy and they rarely metastasize to distant organs. However, local recurrence after an incomplete resection has been reported in a case described by Yates *et al*[3]. Therefore, a tumor resection with negative margins is important for a complete cure. Endoscopic resection has been reported in three cases of ELS[8,10,13]. Among these cases, Olivier *et al* have reported a case of local recurrence after an endoscopic resection requiring an additional esophagectomy[13]. Accordingly, the endoscopic removal of ELS may result in an increased risk of an incomplete resection of tumor tissue because the esophageal lumen is too narrow for sufficient manipulation through an endoscope, particularly in cases of large ELSs. Therefore, endoscopic removal is not recommended for large ELSs. In the absence of lymph node metastasis, radical esophagectomy with an extensive lymphadenectomy, which is important for curing esophageal carcinoma, may be unnecessary. The removal of the tumor with sufficient surgical margins under direct vision through an esophagotomy may be the most appropriate surgical modality because of the simplicity of this procedure, the excellent preservation of the native esophageal function after surgery, and the pedunculated configurations of ELSs with small stalks which allows for a minimal esophageal resection.

In conclusion, ELS is an extremely rare malignant mesenchymal tumor, which is mostly considered to have a low-grade malignancy. Despite that, a complete surgical resection is needed due to the possible occurrence of an acute airway obstruction and the risk of local recurrence after an incomplete removal.

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