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【 CASE REPORT 】

Esophageal Leiomyosarcoma Diagnosed by Endoscopic Ultrasound-guided Fine-needle Aspiration Biopsy and Cured with Surgical Resection

Masahide Ebi, Kazumasa Sakamoto, Satoshi Inoue, Tomonori Ozeki, Mikitoshi Kimura, Riki Kondo, Tomoya Sugiyama, Kazuhiro Yamamoto, Kazunori Adachi, Takashi Yoshimine, Yoshiharu Yamaguchi, Yasuhiro Tamura, Shinya Izawa, Yasutaka Hijikata, Yasushi Funaki, Naotaka Ogasawara, Makoto Sasaki and Kunio Kasugai

Abstract:
Esophageal leiomyosarcomas are rare. We herein present the case of an 82-year-old patient who underwent upper gastrointestinal endoscopy, which revealed a submucosal tumor of 30 mm in diameter that was in contact with the esophagus. Endoscopic ultrasound-guided fine needle aspiration biopsy was performed and the histopathological findings indicated esophageal leiomyosarcoma. Surgical resection was performed. On histopathological examination, the tumor was found to consist of spindle cells with deep chromatin nuclei. The tumor was finally diagnosed as esophageal leiomyosarcoma. We were able to diagnose early-stage esophageal leiomyosarcoma using endoscopic ultrasound-guided fine-needle aspiration biopsy (EUS-FNA). EUS-FNA is mostly recommended as a diagnostic tool for esophageal submucosal tumors.

Key words: endoscopic ultrasound-guided fine-needle aspiration biopsy, esophageal leiomyosarcoma

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Introduction

Leiomyosarcomas of the esophagus are rare, accounting for approximately 0.1-0.5% of malignant esophageal tumors (1, 2). A preoperative pathological diagnosis is difficult to make because leiomyosarcomas originate from the muscle layers. However, a few previous studies reported on leiomyosarcomas that were diagnosed before treatment. We herein report a case of leiomyosarcoma of the esophagus that was diagnosed by endoscopic ultrasound-guided fine-needle aspiration biopsy (EUS-FNA) and cured by surgical resection.

Case Report

An 82-year-old man visited our hospital due to high fever and epigastric pain. He had been treated for chronic renal failure due to nephrosclerosis. He was a smoker and had a habit of drinking alcohol every day. A laboratory examination revealed liver dysfunction, bilirubinemia, and an increased inflammatory response. The patient's white blood cell count (WBC) was 9,700/μL and his C-reactive protein (CRP) level was 11.58 (Table).

Abdominal CT revealed a central biliary duct (CBD) stone and a tumor located in the posterior mediastinum. The tumor was approximately 3 cm in size and was in contact with the esophagus (Fig. 1). He was diagnosed with cholangitis caused by a CBD stone and underwent endoscopic sphincterotomy (EST). No adverse events occurred during EST. Esophagoscopy revealed a submucosal tumor of approximately 30 mm in diameter on the posterior side of the lower esophagus, 35 cm from the incisors (Fig. 2). Endoscopic ultrasonography (EUS) revealed a mosaic-echo-patterned tumor of approximately 30 mm in diameter that originated from the muscularis propria (Fig. 3), and rich

Department of Gastroenterology, Aichi Medical University School of Medicine, Japan
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Correspondence to Dr. Masahide Ebi, mebi@aichi-med-u.ac.jp

Table. **Laboratory Examination Results on Admission.**

Urinalysis		Biochemistry		Immunology	
pH	6.0	TP	5.9 g/dL	HBs-Ag	<0.01 IU/mL
Protein	(2+)	Alb	3.1 g/dL	HBs-Ab	<0.1 mIU/mL
Glucose	(1+)	T-bil	2.94 mg/dL	HCV-Ab	0.13 S/CO
O.B.	(1+)	D-bil	1.85 mg/dL		
Bilirubin	(1+)	AST	423 U/L		
Ketone body	(-)	ALT	591 U/L		
		LD	275 U/L		
Complete blood count		γ-GTP	1,184 U/L		
WBC	9,700 /μL	CK	234 U/L		
Neu	82 %	BUN	27.8 mg/dL		
Lymph	9.2 %	Cr	1.74 mg/dL		
Eos	0.1 %	Na	137 mmol/L		
RBC	366×10^4 /μL	K	3.9 mmol/L		
Hb	12.4 g/dL	Cl	101 mmol/L		
HT	34.0 %	CRP	11.58 mg/dL		
Plt	17.6×10^4 /μL				

Alb: albumin, ALT: alanine aminotransferase, AST: aspartate aminotransferase, CK: creatinine kinase, Cl: chloride, CRP: c-reactive protein, Eos: eosinophil, γ-GTP: gamma-glutamyl transpeptidase, Hb: hemoglobin, HBs-Ag: hepatitis B surface-antigen, HBs-Ab: hepatitis B surface-antibody, HCV-Ab: hepatitis C virus-antibody, HT: hematoclit, K: potassium, LD: lactate dehydrogenase, lymph: lymphocyte, Na: sodium, Neu: neutrophil, O.B.: occult blood, Plt: platelet, T-bil: total bilirubin, TP: total protein, RBC: red blood cell count, WBC: white blood cell count

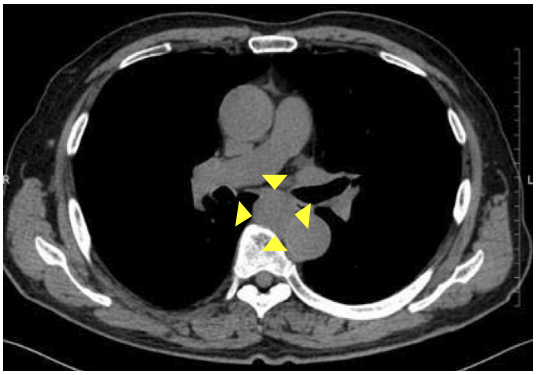

Figure 1. **Computed tomography showed wall thickening of the esophagus (arrowheads).**

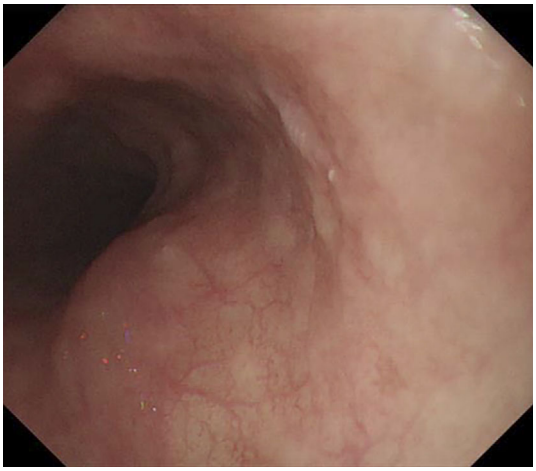

Figure 2. **Endoscopy showed a submucosal tumor in the esophagus, with no mucosal change.**

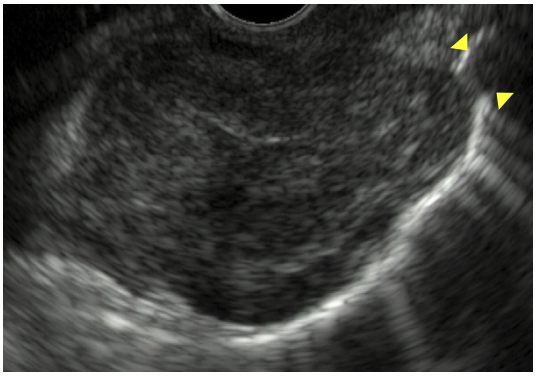

Figure 3. **Endoscopic ultrasonography (frequency, 7.5 MHz) showed that the submucosal tumor located in the muscularis propria (arrowheads) had a heterogeneous appearance and rich vascularity.**

vascularity in the tumor was detected in color Doppler mode. The differential diagnoses of submucosal tumors include gastrointestinal stromal tumor (GIST), leiomyoma, and leiomyosarcoma. The EUS findings of leiomyosarcoma include cystic spaces and irregular tumor margins, while the EUS findings of GIST include an inhomogeneous and anechoic or high echo pattern. On the other hand, leiomyomas show a homogenous echoic pattern on EUS (3, 4). Thus, the tumor in our case was considered to be leiomyosarcoma or GIST. EUS-FNA was subsequently performed. A histopathological examination revealed spindle cells with irregular nuclei (Fig. 4), and immunohistochemical staining showed

that the tumor cells were positive for caldesmon, desmin, and α-smooth muscle actin (α-SMA), but negative for c-kit, CD34, S-100, and anti-pan cytokeratin antibody (AE1/AE3). The Ki-67 index was 50-60% (Fig. 5). The histopathological findings indicated an esophageal leiomyosarcoma. Surgical resection revealed a tumor of 35×30 mm in size that was white and light pink in color (Fig. 6). A histopathological examination revealed that the tumor consisted of spindle cells with deep chromatin nuclei. The immunohistochemistry findings of the surgically resected tumor were in line with the EUS-FNA findings. The patient was finally diagnosed with an esophageal leiomyosarcoma.

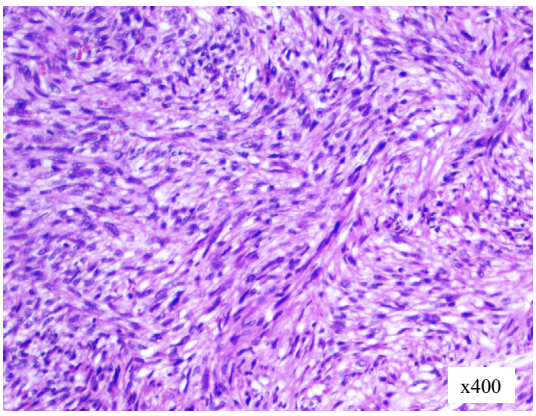

Figure 4. The histological examination of the tumor specimen obtained from endoscopic ultrasonography-fine needle aspiration. Hematoxylin and Eosin staining showed that the tumor consisted of spindle cells (original magnification, ×400).

Discussion

We reported a case in which esophageal sarcoma could be diagnosed by EUS-FNA. Malignant mesenchymal lesions are classified into eight pathologic diagnoses. Leiomyosarcomas are reported to account for up 8.4% of malignant mesenchymal lesions (5). Esophageal leiomyosarcomas are the most common malignant sarcomas. Suzuki et al. reported that esophageal leiomyosarcomas are extremely rare, with a reported incidence rate of 0.17% among all resected esophageal malignant tumors (6). The preoperative diagnosis of esophageal leiomyosarcomas is very rare; only 20-30% of esophageal leiomyosarcomas are diagnosed before surgery. This is a rare case of an esophageal leiomyosarcoma that was diagnosed by EUS-FNA before surgical resection. To date approximately 200 cases of leiomyosarcomas of the esophagus have been reported (2, 7-11). However, only two cases were diagnosed before treatment by EUS-FNA (12, 13). The tumor sizes in those cases were 14.5 cm and 9 cm, respectively. However, the tumor in our case was 3.5 cm in size, and was smaller than the tumors in the two other cases. Our case is the first report to describe the diagnosis of a leiomyosarcoma of the esophagus of <4 cm in size by EUS-FNA. EUS-FNA is a useful method for diagnosing gastrointestinal submucosal tumors (4) in general. The accuracy rate of EUS-FNA in the diagnosis of gastrointestinal submucosal tumors in the esophagus was reported to be 52.3-100% (14, 15), while that in the stomach was reported to be 92.2-92.7% (16, 17). EUS-FNA is considered a safe procedure, with an adverse event rate of 0.5%. There is only one reported case of patient who developed streptococcal sepsis with high fever and abdominal pain after EUS-

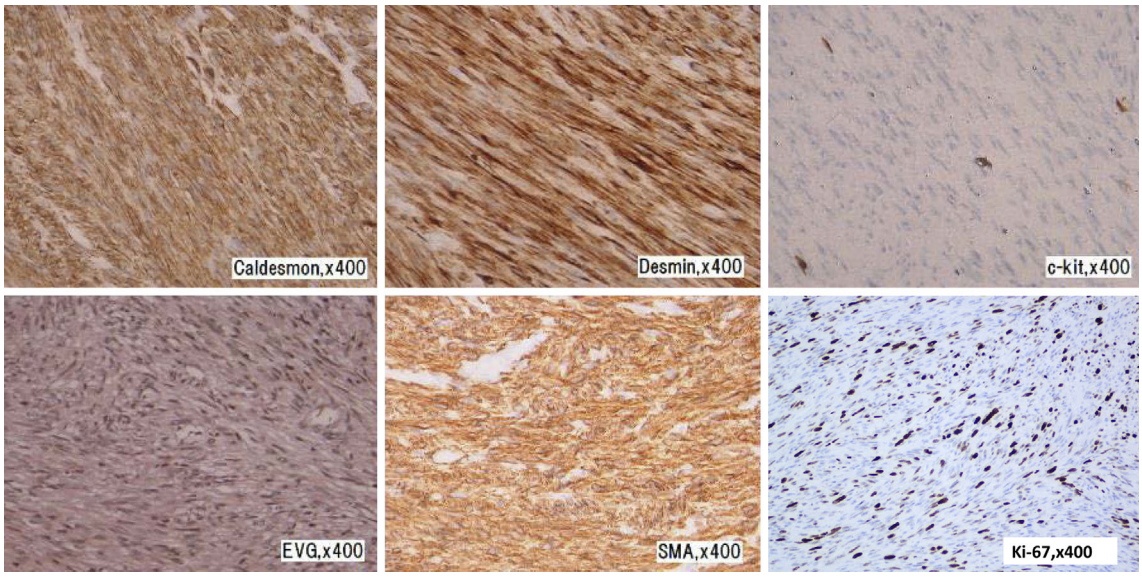

Figure 5. The histological examination of the resected tumor. Immunohistochemical staining revealed that the tumor cells were positive for caldesmon, desmin, and α-smooth muscle actin (SMA), and negative for c-kit and Elastica van Gieson staining (EVG). 50-60% of the tumor cells were Ki-67-positive (original magnification, ×400).

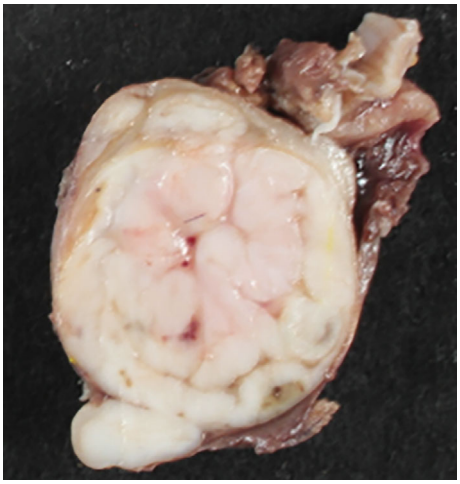

Figure 6. Macroscopic findings of the resected tumor.

FNA as a severe adverse event (16, 18, 19). Another modality by which submucosal tumors of the esophagus can be diagnosed is deep biopsy using endoscopic submucosal dissection (ESD). The diagnostic yield for upper gastrointestinal submucosal tumors was reported to be 90-96.15% (20, 21). On the other hand, the procedure requires ESD technique, and the severe adverse events such as a pneumomediastinum and bleeding were reported (21).

The principal symptoms of esophageal leiomyosarcomas include dysphagia, body weight loss, difficulty in swallowing food, cough, upper gastrointestinal hemorrhage, emesis, epigastric pain, and back pain (8, 11, 22, 23). The mean tumor size of esophageal leiomyosarcomas was previously reported to be 86.5 mm (11, 24, 25) and the development of symptoms was considered to be associated with an enlarged tumor that occupying the esophageal lumen. Although the patient had epigastric pain due to a CBD stone, no symptoms related to the esophageal tumor were recognized due to the small size of the tumor. The rate of recurrence in patients with leiomyosarcoma is reported to be 39-64% (2, 10, 11), and the sites of recurrence are reported to include the liver, lungs, and brain (2). The 5-year overall survival rate is reported to be 35-58% (2, 11, 26). The 5-year overall survival rate of patients with stage I and II diseases was reported to be 71.4%, which is significantly better than that of patients with stage III and IV (14.3%) disease (2). The Ki-67 index of the leiomyosarcoma in our case was 50-60%; however the risk of recurrence is not correlated with the grade of the Ki-67 expression. The growth type was reported to be associated with survival, with polypoid or intramural growth types having a better prognosis than the infiltrating type. Our case was an intramural growth type leiomyosarcoma. This case was completely resected by surgery, and no recurrence was detected in one year of follow-up.

In conclusion, the prognosis of early-stage esophageal leiomyosarcoma is relatively good, and EUS-FNA is mostly recommended as a diagnostic tool for esophageal submucosal tumors.

The authors state that they have no Conflict of Interest (COI).

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