A Case of Pedunculated Esophageal Leiomyoma Successfully Treated by Endoscopic Mucosal Resection

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(Received August 2, 2016; Accepted June 19, 2017)

Leiomyoma is one of the most commonly observed esophageal submucosal tumors, often appearing as a smooth-surfaced and semicircular protruded lesion. It sometimes grows toward the esophageal lumen and may be pedunculated in rare cases. We encountered a case of a pedunculated esophageal submucosal tumor diagnosed before treatment as a leiomyoma originating in the muscularis mucosae of a 68-year-old man. As the tumor arose in the muscularis mucosae, it could be safely resected via an endoscopic procedure. Only one case of pedunculated leiomyoma has been reported to date, and we herein report the second case, which was successfully treated by a minimally invasive endoscopic technique.

Key words: esophagus, submucosal tumor, leiomyoma, endoscopic mucosal resection, pedunculated tumor

INTRODUCTION

Esophageal submucosal tumors indicated for surgical treatment are usually symptomatic, exceed 2 cm in diameter, or suspected to be malignant [1, 2]. Most originate in or adhere to the muscularis propria, and therefore, unlike mucosal lesions, cannot be treated by endoscopy without a high risk of perforation. However, if submucosal tumors morphologically appear as pedunculated lesions and originate in the muscularis mucosae, they can be safely resected by endoscopy, thus avoiding surgery. We herein report a rare case of a pedunculated submucosal tumor originating in the muscularis mucosae preoperatively evaluated by endoscopic ultrasonography and successfully treated by endoscopic mucosal resection.

CASE REPORT

A 68-year-old man had severe solid food dysphagia for 6 months. A submucosal tumor of esophagus was suspected in an upper gastrointestinal contrast examination conducted during his annual health check-up, and he was referred to our hospital. He had previously undergone endoscopic mucosal resection for colonic polyps at ages 64 and 65 years.

Blood tests performed at our hospital did not show any abnormal results, including those for tumor markers, squamous cell carcinoma antigen, and cytokeratin 19 fragment. The upper gastrointestinal contrast study revealed a pedunculated mass in the upper thoracic esophagus. The stalk of the mass measured 25 mm in length, and the head measured 28 x 20 mm in size. The mass was smooth and regular with no ulceration (Fig. 1). Upper gastrointestinal endoscopy also revealed a smooth pedunculated mass lesion with a stalk originating 22 cm from the incisors and a head covered by normal mucosae. The surface of both the tumor stalk and head showed no irregularity, and all areas could be stained with Lugol's solution. The stalk of the mass was soft and flexible when touched with forceps, whereas its head was hard and elastic (Fig. 2). The biopsy specimens obtained from the tumor head and stalk contained only normal esophageal mucosa. Therefore, the mass lesion was assumed to be a pedunculated submucosal tumor, an uncommon morphological type of esophageal submucosal tumor.

The tumor was further examined by an endoscopic ultrasonography (EUS). It appeared as a homogeneous hypoechoic mass with no obvious blood flow as determined by using color Doppler ultrasonography, which suggested a solid, homogeneous tumor with rather poor blood flow. The hypoechoic area of the mass continued to the second of the five layers of the esophageal wall (Fig. 3), and continuity of the third layer was maintained under the tumor base, suggesting that the tumor originated in the muscularis mucosae. These findings indicated that the submucosal tumor was probably a leiomyoma or gastrointestinal stromal tumor (GIST) originating in the muscularis mucosae. Contrast-enhanced abdominal computed tomography (CT) showed that the tumor was relatively homogeneous with mild enhancement, and no proximal lymphadenopathy or metastasis to other organs was observed (Fig. 4). Magnetic resonance imaging (MRI) depicted the tumor as slightly hyperintense on T2-weighted images with a homogenous interior, without

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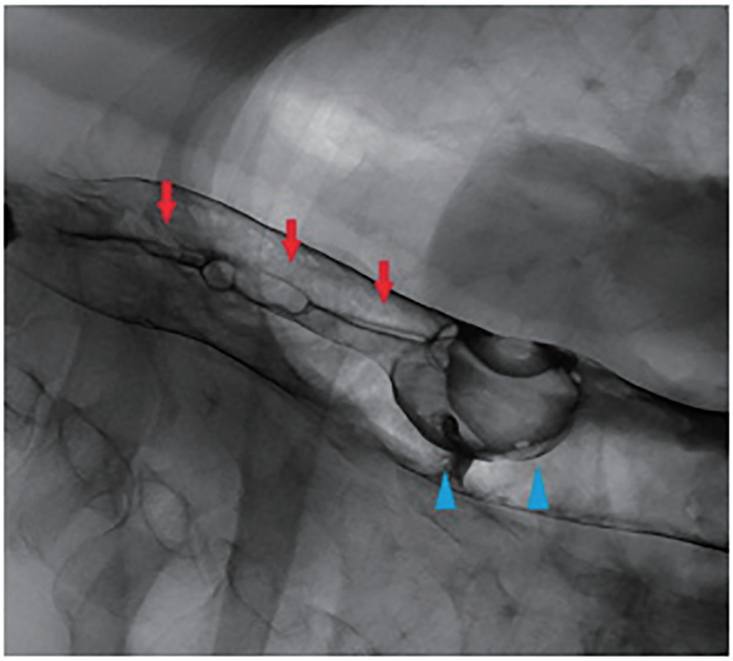

Fig. 1 A barium contrast image of the esophagus
The image reveals a pedunculated smooth mass originating in the upper thoracic esophagus. The red arrows and the blue arrowheads indicate the long stalk and the head of the polypoid lesion, respectively.

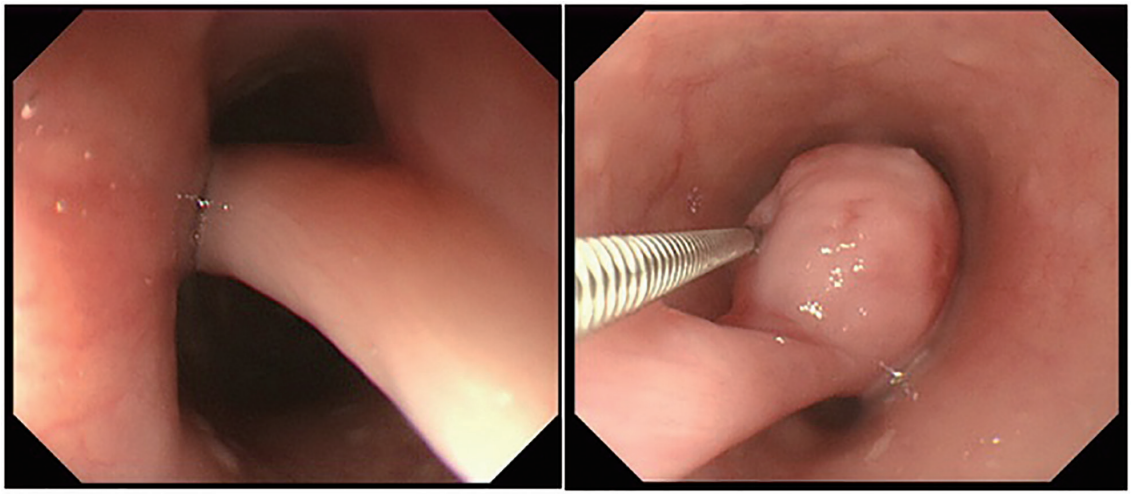

Fig. 2 Endoscopic views of the esophageal tumor
The image shows that the tumor was a similar color as the cortical layer and was pedunculated, easily movable, and elastic and hard when compressed with forceps.

any lipid component. Based on the CT and MRI findings, the tumor was solid, composed of homogeneous tissue, and none of the findings suggested malignancy. Because the tumor was assumed to be probably a benign but symptomatic submucosal tumor originating in the muscularis mucosae, endoscopic resection was our therapeutic choice. After a detachable snare was placed at the stalk, the head was entrapped with the snare, and the tumor was resected electrically using coagulation waves. No complications were observed during or after the procedure, and the patient was discharged after 5 days. Discomfort upon swallowing was resolved after the endoscopic surgery.

Histopathology of the resected tissue revealed a submucosal tumor composed of bundle-like structure on hematoxylin-eosin staining as well as proliferating spindle cells (Fig. 5). None of the findings were suggestive of malignancy, such as highly atypical cells, increasing karyokinesis, or necrosis. Immunostaining showed expression of smooth muscle actin, while expression of either c-kit (CD117) or S-100 protein was not detected (data not shown). The tumor continued to the layer of muscularis mucosae as depicted in Fig. 6. The tumor was therefore finally diagnosed as a leiomy-

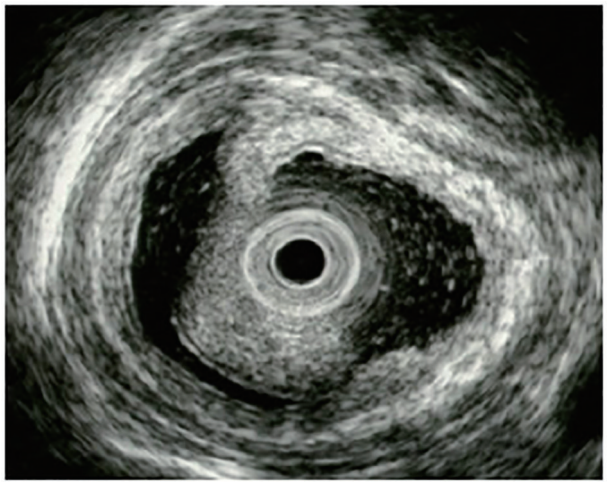

Fig. 3 Endoscopic ultrasonography
The endoscopic ultrasonogram shows that the main body of the tumor continued to the second layer, and that continuity of the third layer, including the base of the tumor, was maintained.

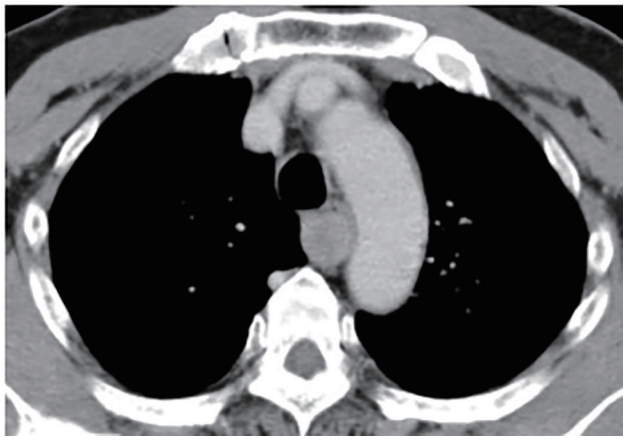

Fig. 4 Contrast-enhanced computed tomography
The image depicts the tumor as a less intensely enhanced mass compared to the great vessels with a homogeneous interior.

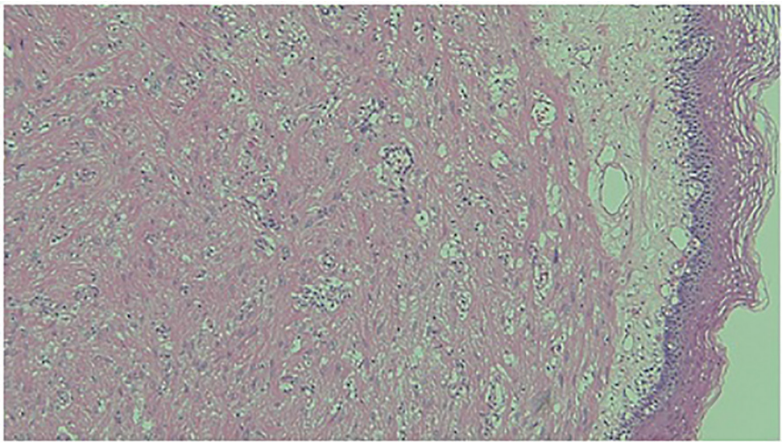

Fig. 5 Histopathology of the resected tumor on hematoxylin-eosin staining
Histopathology revealed a bundle-like structure as well as proliferating spindle cells. Highly atypical cells, increasing karyokinesis, or necrosis were not detected.

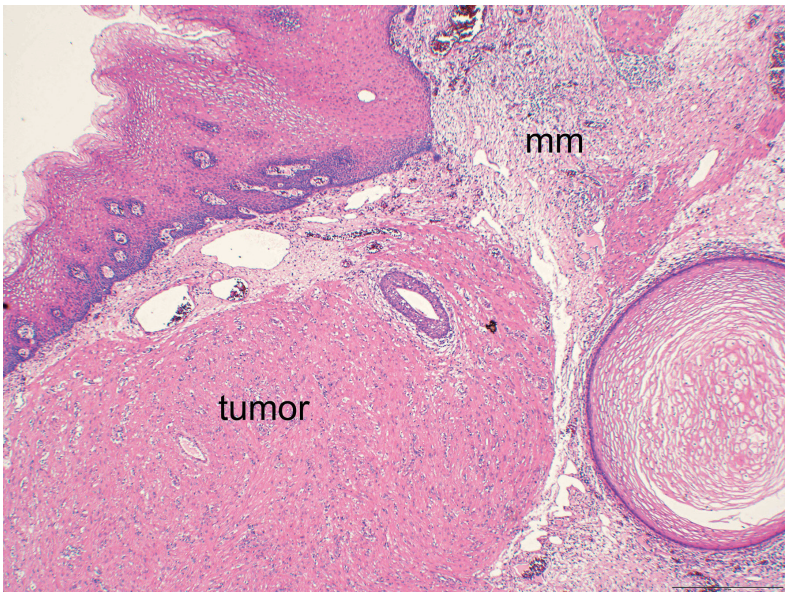

Fig.6 Histopathology of the resected tumor and the muscularis mucosae on hematoxylin-eosin staining
The submucosal tumor, indicated as "tumor" was very close or almost fused to the muscularis mucosae, indicated as "mm", suggesting that it originated in the muscularis mucosae.

Table Comparison of our case and five other cases of pedunculated esophageal submucosal tumors

Year/Author	Age/Sex	Symptom	Location	Morphological characteristics	Tumor size	EUS (head)	Layer of the origin	Pathological Diagnosis
2004/Kuo MJ [8]	61/M	None	Mt	smooth	30 mm	homogeneous, hypoechoic	2/5	Leiomyoma
2008/Best SR [9]	63/M	dysphagia	Ut	smooth	30 × 20	not evaluable	unevaluable	Lymphangioma
2008/Liu CH [10]	67/M	dysphagia	Ut	smooth and flexible	90 × 47 × 25	not evaluable	unevaluable	Lipoma
2014/Cuk [11]	29/M	Dysphagia fever	Ce	smooth	210 × 90× 60	heterogenic, hypoechoic-dominant	3/5	Fibrovascular polyp
2015/Qinying [12]	52/M	dysphagia	Ce	smooth and flexible	45 × 25 × 16	not done	---	Lipoma
2012/our case	68/M	dysphagia	Ut	smooth and flexible	28 × 20 mm	homogeneous hypoechoic	2/5	Leiomyoma

EUS, endoscopic ultrasonography
Ce, cervical esophagus
Ut, upper thoracic esophagus
Mt, middle thoracic esophagus

oma originating in muscularis mucosae.

DISCUSSION

Esophageal submucosal tumors are usually diagnosed by a conventional upper gastroenterological endoscopy. Further diagnosis of submucosal tumors, however, may be difficult because histological samples cannot be taken easily or safely, unlike mucosal lesions. Especially for the pedunculated ones, EUS-guided fine needle biopsy, a standard technique to get a sample from a submucosal tumor, is often avoided due to its high mobility meaning a high risk of perforation. Although modalities such as EUS, CT, and MRI are very useful, differentiating leiomyoma from leiomyosarcoma or GIST is challenging [3, 4]. Positron emission tomography may be useful for the differentiation, but the Japanese national insurance system has not covered such submucosal tumors without signs of highly suggesting malignancy and could not be performed in this case.

As for esophageal leiomyoma, most were located in the middle and lower parts of esophagus and rather rare in the upper part like this case and 62 % of the tumors originated in muscularis mucosae, according to Xu, *et al*. [5] Sato, *et al*. speculated that development of a pedunculated structure was created by the propulsive forces due to peristalsis combined with the traction of the passing stool, although for the colonic ones [6]. In a case of esophageal leiomyomatosis, a rare disorder in which proliferation of smooth muscle in both of the muscularis mucosae and the muscularis propria caused marked circumferential thickening and multiple SMT like protrusions in a large portion of esophagus, the only pedunculated one originated in the muscularis mucosae [7], suggesting that only one originating in the muscularis mucosae could be pedunculated and the incidence might be rare. Combining these reports [5–7], we speculated that an esophageal leiomyoma originated in the muscularis mucosae, thinner and weaker than the muscularis propria, might be pulled and change its appearance to be pedunculated by rather strong propulsive forces due to the food swallowing and peristalsis in relatively narrow upper esophagus.

Although quite rare, all the past cases of pedunculated leiomyoma originated in the muscularis mucosae, and the tumors can be treated by endoscopic resection without trouble. A search on PubMed using the keywords "esophagus," "submucosal tumor," and "pedunculated" yielded one report on leiomyoma [8], one on lymphangioma [9], one on fibrovascular polyp [10] and two on lipoma [11, 12] (Table). Another report on gastrointestinal submucosal lesions was also screened which contained 3 cases of esophageal submucosal tumors but those were all sessile ones [13]. In the 2 case of the lipoma, surgery was performed because of the undetermined layer of its origin by EUS [11] or the location of cervical esophagus and the large size of 45mm in diameter [12]. As for giant esophageal fibrovascular polyp, it was too large for endoscopic treatment and was treated by surgery [10]. However, in the case of leiomyoma, endoscopic resection was performed successfully, although the tumor size was thought to be possibly malignant based on its size of 30mm in diameter [8]. The standard treatment for leiomyosarcoma and GIST is surgery and leiomyoma is difficult to distinguish from leiomyosarcoma or GIST by imaging features, however the cases were all successfully treated by endoscopy, and thus, surgery could be avoided. If the safety of endoscopic surgery is ensured, diagnostic resection can be a useful choice for treatment of pedunculated submucosal tumors in the esophagus.

In conclusion, diagnostic endoscopic resection appears to be a viable option for treatment of pedunculated esophageal submucosal tumors, especially those that originate in the muscularis mucosae as confirmed through preoperative EUS.

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