Case report

Giant leiomyosarcoma in the upper third of esophagus, a case report

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Abstract

Esophageal leiomyosarcoma (ELMS) is a rare tumor of mesenchymal origin that develops from the smooth muscles of the esophagus. ELMS is typically located in the lower third of the esophagus. We herein report a patient who underwent surgical resection for ELMS, 80 mm in diameter, in the upper third of the esophagus. A 58-year-old man experienced difficulty swallowing, and his swallowing function gradually deteriorated about 6 months before admission. McKeown operation, which is a kind of esophagectomy performed via the thoraco-laparo-cervicotomy approach, was conducted. The postoperative course was good, without complications, and he was discharged on postoperative day 13. A pathological examination revealed highly differentiated ELMS.

Keywords: esophageal leiomyoma, leiomyosarcoma, esophagectomy

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Introduction

Esophageal leiomyosarcoma (ELMS) is a rare tumor of mesenchymal origin that develops from the smooth muscles of the esophagus\(^1\). It accounts for <1% of all neoplasms of the esophagus\(^2\). It is mainly found in the lower to middle third of the esophagus, where the muscle layer of the organ consists only of smooth muscle fibers. Indeed, the largest case-series study of esophageal leiomyosarcoma showed that approximately 90% of ELMS cases developed from the lower or middle third of the esophagus\(^3\).

We herein report an extremely rare case of a large ELMS arising from the upper third of the esophagus.

Case presentation

The patient was a 58-year-old man who was admitted in 2019 to the Thoracic Oncology Department due to dysphagia. He had felt difficulty swallowing, and his swallowing function had gradually deteriorated over the six months before admission. He was able to eat soft food or liquid, and his weight loss was negligible. His body mass index was 24.5 (kg/m\(^2\)).

Examinations

Esophagoscopy revealed a submucosal protruding lesion with ulceration located 6 cm distal from the pharyngo-esophageal junction. Although a biopsy was taken at the surface of the lesion, the findings of squamous epithelium with hyperplasia were only revealed on a histological examination.

Figure 1 shows the findings of a barium meal examination. In the upper third of the thoracic esophagus, the lumen was locally narrowed, shifted to the right side and deformed.

Figure 2 shows the findings of thoracic computed tomography (CT). The oval-shaped volumetric mass in the upper mediastinum (55 × 65 × 80 mm) compressed the adjacent organs, including the trachea, descending thoracic aorta and upper lobe of the right lung. The lower part of the trachea was compressed by adjacent esophageal tumor but no findings of invasion were noted on bronchoscopy.
Figure 1  Esophagography with barium meal examination
The esophageal lumen narrowed and shifted to the right side in the upper third of the thoracic esophagus (white arrow).

Figure 2  The findings of thoracic CT scan
An oval-shaped volumetric mass in the upper mediastinum with dimensions of 55 × 65 × 80 mm.
The diagnosis and treatment

The potential diagnoses were gastrointestinal stromal tumor (GIST), leiomyoma or leiomyosarcoma of the upper thoracic esophagus. Through a multidisciplinary conference, McCeown’s operation, which is a kind of esophagectomy performed by the thoracolaparoservicotomy approach, with esophagogastrostomy was proposed as the primary treatment, taking into account the deterioration of his dysphagia.

After informed consent was obtained from the patient, surgery was carried out. Thoracotomy on the right through the fifth intercostal space was performed. The tumor derived from the left-anterior wall of the esophagus above the azygos vein showed a dense consistency and had a smooth surface (Figure 3). The tumor sharply constricted the trachea and superior vena cava. Abutting mediastinal structures showed no signs of involvement in the tumor process. The affected part of the esophagus was mobilized, and through a longitudinal esophagotomy over the mass, the tumor was enucleated from the esophageal wall. We carefully manipulated and secured the surgical margin for intact adventitia of the esophagus. Lymphadenectomy was performed to remove the regional lymph nodes (cervical, mediastinal and abdominal). Standard steps for McCeown’s surgery include the formation of an end-to-side anastomosis on the neck, creating a sufficient length of the gastric transplant and the formation of the least dangerous esophago-gastro anastomosis on the left neck. At the end of the operation, the patient had a double-lumen nasoenteral tube, which was subsequently used for decompression of the transplant and anastomosis as well as for enteral feeding (inner lumen) in the postoperative period. The total duration of the operation was 300 minutes (Figure 4).

The postoperative course was good. The nasoenteral tube was removed after satisfactory findings were obtained on oral contrast radiography (76% sodium amidotrizoate solution) of the anastomosis on the sixth and seventh days after surgery, and the patient was transferred to oral nutrition. He was discharged on postoperative day 13. After one, three and six months of follow-up, esophagoscopy and X-ray studies had not shown any evidence of pathology from the esophagogastroanastomosis or transplant.

A pathological examination revealed the tumor to be highly differentiated ELMS (Figure 5). Immunohistochemistry showed positive staining for alpha-SMA and negative for c-kit and S-100 protein (No images). No lymph nodes metastasis was histologically confirmed.

Given the low sensitivity of LMS for chemo-radiation therapy and the lack of any metastases in the regional lymph nodes, we refrained from adjuvant therapy, and the patient has been undergoing observation. Control esophagoscopy and X-ray contrast study of the esophagogastroanastomosis at one, three and six months after the operation did not detect any metastatic lesions. In addition, we plan to perform the periodical positron emission tomography every year as a further follow-up.
Figure 4  The resected specimen of the esophageal tumor
The removed tumor with dimensions 70 × 80 × 80 mm had dense consistency and a smooth surface. The split section was whitish in color, and the cellularity was high without necrosis.

Figure 5  The microscopic findings of the tumor
The findings of atypical smooth muscle cells and mitosis, which are characteristic of leiomyosarcoma, were confirmed on the pathological examination.

Discussion

ELMS is extremely rare, accounting for 0.1%–2.8% of all malignant neoplasms of the esophagus, according to the literature[1-6]. Approximately 200 cases of ELMS have been reported around the world. ELMS is characterized by a slow growth and late metastasis and thus has a better prognosis than squamous cell carcinoma and adenocarcinoma of the esophagus[7,8]. Similarly, in the present
case, no metastatic lesions of the lymph nodes or other structures were detected\(^2, 5, 6, 8\). The 5-year survival of patients with ELMS, depending on the nature of growth, ranges widely from 25.0%–83.3\(^1,3,5-8\). Infiltrative tumor growth is considered more malignant, while intramural and polypoid tumor growth have a more favorable outcome. The overwhelming majority of sources confirm that ELMS mainly occurs in middle-aged and elderly patients\(^3\). Our case was no exception in terms of the age, with his age falling in the average age range of patients with ELMS (50–60 years). Based on these data, we decided to perform initially surgical resection for this tumor through our multi-disciplinary conference.

ELMS is often found in the lower third of the esophagus, where the muscle layer of the organ consists only of smooth muscle, and cases where the process is located in the middle third of the esophagus are rare\(^1, 2, 5, 8\). However, the upper third of the esophagus is an extremely rare place for LMS. The clinical symptoms of ELMS are not specific, and the most common is dysphagia, the frequency of which ranges from 64.7%–91.7% according to various sources\(^5, 8\). Other symptoms include retrosternal pain and back pain (approximately 75%), weight loss (approximately 62.5%), and emesis and respiratory symptoms. Dysphagia usually appears when the tumor reaches a large size. Indeed, in our case, the patient had a 6-month history of dysphagia, and the tumor was 80 mm in size.

Treatment for ELMS is carried out similarly to that for other soft tissue sarcomas. Surgical treatment is considered the treatment of choice for ELMS. Although local excision of the tumor has provided a good overall survival, resection of the esophagus is the standard for this pathology\(^5, 8\). Other treatment approaches, such as radiation therapy and chemotherapy, are limited in use, since malignant smooth muscle cells are resistant to radiation and chemotherapy\(^1, 7\). We performed McCoeown’s surgery to completely remove the tumor and achieve adequate lymphatic dissection. In the postoperative period, the patient was not prescribed adjuvant chemoradiotherapy, and we have been performing follow-up examinations.

The prognosis of the disease is more favorable for female patients than males and for cases with tumors of polypoid and intramural growth than squamous cell carcinoma. Accordingly, tumors with infiltrative growth have an unfavorable prognosis.

**Conclusion**

We described an extremely rare case of large ELMS in the upper third of the thoracic esophagus. Our report indicates that McCoeown’s operation, which made it possible to achieve adequate access to the surgical area, is a viable option for local control.

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