

Case Report

Giant Esophageal Liposarcoma Diagnosed by Endoscopic Ultrasound

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Abstract

We report a case of giant well-differentiated esophageal liposarcoma in a Chinese patient. Liposarcoma is a common soft tissue malignant tumor that usually arises in the lower extremities, trunk, and the retroperitoneum, but it is very rare that liposarcoma locates in the gastrointestinal tract, especially in the esophagus. Endoscopic ultrasound could be an efficient tool for preliminary examination. Moreover, the patient underwent combined thoraco-laparoscopic surgery, after reviewing the published articles, this surgery was first applied in the excision of esophageal liposarcoma due to the detailed preoperative information from the ultrasound technique.

Keyword: Endoscopic ultrasound, esophageal liposarcoma, minimal invasive method

INTRODUCTION

Esophageal liposarcoma was first reported by Mansour in 1983,^[1] and this extremely rare disease accounts for only 1.2% to 1.5% of gastrointestinal liposarcoma,^[2] merely 70 cases were published in the English literature as so far. It was reported that the average age of onset was 58.4 years old, men were more susceptible to the disease, and the average size of the specimen was 13.3 cm. Eighty percent of esophageal liposarcoma is situated in the cervical segment of the esophagus.^[3,4] The tumor usually grows slowly and the common symptoms include dysphagia and regurgitation, after surgery, the favorable prognosis is usually achieved. The imaging diagnosis of this disease mainly depends on the examination of computed tomography (CT) and endoscopic ultrasound (EUS), the preoperative pathology is usually acquired by the operation of EUS-guided or esophagoscopy biopsy. The tumor may present as an intramural submucosal mass or intramural mass with a vascularized pedicle, or transmural mass which invades the wall of the esophagus, so the surgical option varies with the location of the tumor.^[5]

CASE REPORT

A 52-year-old Chinese man presented with a 10-year-history of intermittent dysphagia for solid food without complaints of chest pain and regurgitation, and the symptom worsened after drunk 2 years ago. The patient was performed a contrast-enhanced CT, a heterogeneous mass can be observed growing along the wall through the entire course of the esophagus, covering an area of 25 cm in length and 3.2 cm in width [Figure 1a]. Flexible esophagoscope visualized a large submucosal lesion at 17 cm to 40 cm from the incisor teeth [Figure 1b]. Further examination of EUS was performed, the examination was well tolerable, and there were no complications, such as perforation and bleeding. The B-mode ultrasonogram revealed a mixed echogenic mass originating from the entrance to the dentate line of the esophagus. Meanwhile, the outer membrane was not invaded, the upper part of the mass was hyperechoic which was consistent with the ultrasonic characteristic of fat [Figure 2a], and the lower part was hypoechoic which meant more solid composition than the upper part [Figure 2b]. Color Doppler detected no blood flow

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signal in the mass; meanwhile, an artery was showed aside the mass [Figure 2c], and the boundary between the mass and artery was clear. During the combined thoraco-laparoscopic surgery, pectoral, abdominal portion of the esophagus and the proximal lymph nodes were removed, and the specimen of this tumor had a smooth outer membrane [Figure 3]. Histopathological examination of this 22.5 cm × 5.2 cm × 3.1 cm mass was consistent with well-differentiated heterogeneous esophageal liposarcoma, the pathology of hyperechoic part of the tumor showed that the fat cells were dominant, only a few spindle cells and lymphocytes were found [Figure 4a]. The immunochemistry showed the expression of S-100 (fat 2+), which marked the adipocytes or fat cells in the tumor tissue. The pathology of hypoechoic part of the tumor showed that a large area was covered by spindle cells, and proliferation of fibrous tissue was obvious [Figure 4b]. The immunochemistry showed the expression of CD34 (2+), which mainly expressed in the spindle cells in the tumor of liposarcoma.

DISCUSSION

Esophageal liposarcoma is normally classified into the types of myxoid, pleomorphic, well differentiated,

and dedifferentiated. Moreover, well-differentiated subtype accounts for approximately 72% of all the categories.^[6] In general, the mass grows slowly, most patients are hospitalized for dysphagia and regurgitation when the mass grows big enough. In this case, the patient presented with a gradual progression of intermittent dysphagia, the surgeons did an emergent operation to the ease this fatal symptom. Although much valuable diagnostic information was provided by the sonogram, EUS could not determine the pathology of this tumor; moreover, the lymph nodes around the esophagus were easy to escape the scanning of the probe, and it was not reliable to diagnose the lymph nodes as malignant or not by this way. Usually, during the preoperational period, EUS-fine-needle aspiration of the tumor and the proximal lymph nodes detected is necessary for the accurate diagnosis,^[7] but in this case, the precious time spent on the procedure may cause an unpleasant result. Therefore, the surgeons did the operation despite the lack of preoperative pathology, and an extended resection of tumor and lymph nodes was performed for

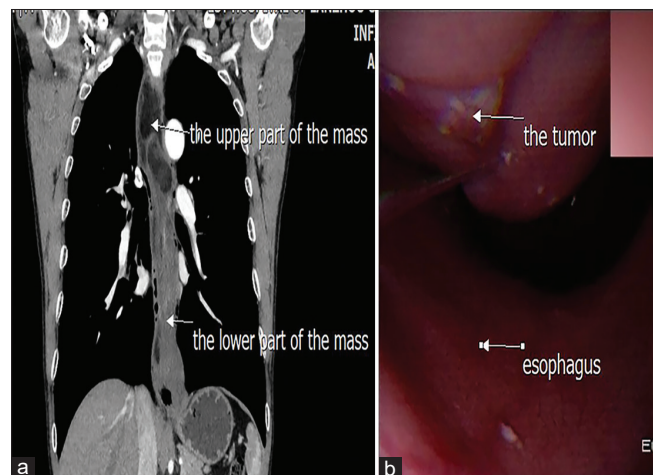


Figure 1: (a) The coronal plane of this heterogeneous tumor in the examination of enhanced computed tomography, the mass grown along the entire esophagus. (b) the endoscopy of the mass shows a huge eminence in the esophageal lumen

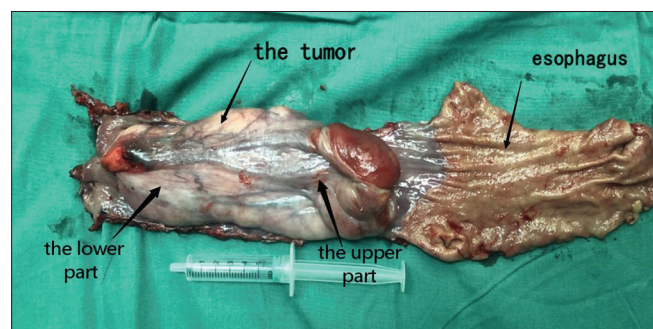


Figure 3: The specimen of this tumor, the outer membrane is smooth, a part of the esophagus was removed

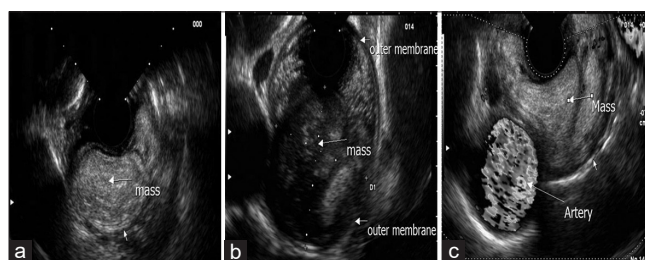


Figure 2: (a) The endoscopic ultrasound shows the upper part of the tumor, the boundary between the tumor and the normal esophagus is clear, and homogeneous hyperecho indicated the composition of fat. (b) The endoscopic ultrasound shows the lower part of the tumor, the outer membrane of the mass is clear, there is not proximal invasion, the outer membrane of the wall of the mass and the normal esophagus is intact, this tumor acquired the transmural nature. (c) The Doppler mode of ultrasound of this mass, there was no blood signal in the mass, and the artery aside the mass was invisible, the boundary between the mass and the proximal artery is clear

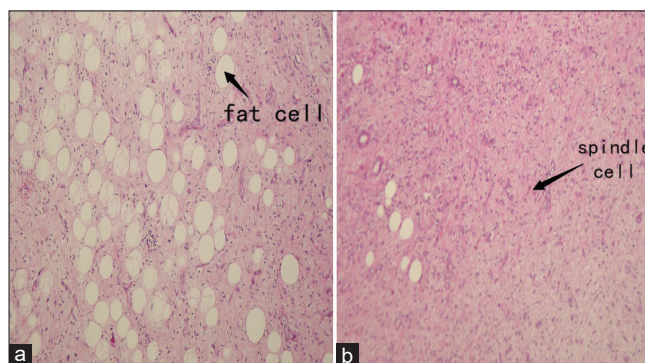


Figure 4: (a) This is the result of pathology of the upper part in this tumor, the fat cells were dominant in this picture, which is consistent with the findings of endoscopic ultrasound. (b) This is the result of pathology of the lower part in this tumor, the spindle cells were dominant in this picture, which is consistent with the findings of endoscopic ultrasound

a better prognosis. The postoperative pathology, in turn, proved the findings detected by EUS, the pathology of the upper part of the tumor showed many fat cells, which absorbed little ultrasound, and existed as hyperecho in sonogram, this was consistent with the findings of EUS. The pathology of the lower part of the tumor showed many spindle cells and proliferation of hyaline degeneration of fibrous tissue, this is the solid composition of the tumor, much ultrasound emitted by the EUS probe was absorbed, and this part was hypoechoic in sonogram. Some results of the immunohistochemistry such as MDM2 (–) and the low expression of ki67 (20%) revealed that the tumor had little aggressiveness, this is consistent of the intact and smooth outer membrane of esophageal wall and the negative findings of proximal tissue in the sonogram.

The key diagnostic tips of EUS for esophageal liposarcoma can be summarized as following four points: first, hyperecho is dominant in most parts of esophageal liposarcoma, and only just a fraction of which presents as hypoechoic sonogram. Second, the outer membrane of the esophageal wall is smooth and intact, the tumor may invade not only the submucosa but also the whole wall of the esophagus; meanwhile, the shape of esophageal liposarcoma is regular, and the boundary is smooth. Third, color Doppler imaging shows the lack of vascularity inside esophageal liposarcoma, proximal artery could be detected, but regular EUS cannot easily find the nutrient vessels of esophageal liposarcoma, even the solid part of the tumor which is hypoechoic in the sonogram. Fourth, the size of esophageal liposarcoma is large, reported tumor sizes ranged from 4 cm to a maximum length of 33 cm, with most of the sizes being over 10 cm.^[8]

Esophageal liposarcoma ought to be differentiated from benign diseases such as esophageal giant leiomyoma and hamartoma. Leiomyoma of the esophagus is often lobulated with central liquefactive necrosis, and endoscopic ultrasonography usually presents as a homogeneous submucosal eminence with clear boundaries in B-mode ultrasound; occasionally, the esophageal leiomyoma may present as an annular submucosal anechoic (cyst like) tumor, which is apparently different from the sonogram of esophageal liposarcoma.^[9,10] Esophageal hamartoma is also derived from the original mesenchymal cells as the same with esophageal liposarcoma, EUS shows heterogeneous hypoechoic mass with rough and unsmooth outer membrane;^[11] meanwhile, blood flow signal is visible inside the mass of esophageal hamartoma in color Doppler ultrasound. In this case, endoscopic ultrasonography of esophageal liposarcoma showed a mixed-echo region with hyperecho in the upper segment and hypoecho in the lower segment of the esophagus with smooth boundary and absence of blood flow signal. These sonographic characteristics are different from the common diseases above. In conclusion, EUS is an accurate and effective examination to exclude other similar diseases.

In general, esophageal liposarcoma is removed by classical cervical open surgery.^[5] However, in recent years, endoscopic

submucosal dissection emerges as a new minimal invasive strategy for the disease.^[12] In the preoperative EUS examination, a transmural nature of the tumor was revealed, which represented that the tumor had grown from inside to the outer membrane of the wall of the esophagus. Hence, an esophagectomy was preferred rather than an endoscopic submucosal dissection for the limitation of excising region during an endoscopic surgery.^[13] However, the patient was too weak to weather the damage of the open surgery. After additional detection of surrounding blood flow by the EUS examination, clear boundary with grand vessels encouraged the surgeons to remove the tumor with the assistance of combined thoraco-laparoscopy. After the operation, only four holes on the patient's body were left, and the bleeding volume was below 200 ml. Finally, the patient experienced a relatively short postoperative period for this minimally invasive method, and there have not been any recurrence or postoperative complications during the follow-up period.

CONCLUSION

Esophageal liposarcoma is a rare tumor. In this case, the mass was huge, transmural through detailed preoperative examinations. The patient underwent combined thoraco-laparoscopy, which was firstly applied in the treatment of esophageal liposarcoma. Preoperative EUS examination played a key role in the diagnosis and the choice of treatment strategy of this disease, which facilitated the minimally invasive treatment of esophageal liposarcoma which is not suitable for endoscopic submucosal dissection.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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