Imaging features and literature review of rare primary giant liposarcoma of the esophagus

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Introduction

Liposarcoma, a prevalent form of soft tissue malignancy in adults, accounts for approximately 20% of all soft tissue malignancies [1]. While it commonly occurs in the retroperitoneum, trunk and extremities, its occurrence within the gastrointestinal tract is rare, with an autopsy-reported occurrence rate ranging from 0.1% to 5.8% [2]. Specifically, esophageal liposarcoma represents an extremely uncommon subset, comprising only 1.2% to 1.9% of all gastrointestinal liposarcomas [2].

The initial documentation of esophageal liposarcoma dates back to 1983, when Mansour first described its presence [3]. Since then, few cases of this rare condition have been reported in the available literature, with approximately 60 cases documented to date [4]. Notably, the existing literature primarily focuses on the pathological features and surgical treatment, without comprehensive exploration of the associated imaging manifestations.

In this study, we present a case of giant liposarcoma of the esophagus, accompanied by detailed imaging findings. Our aim is to contribute to the limited body of knowledge surrounding the radiological features of this extremely rare entity.

1 Case report

A 41-year-old female patient visited our outpatient department complaining of a six-month history of swallowing obstruction without hoarseness, chest and back pain, fever, irritating choking cough, or vomiting blood. Physical examination did not reveal any positive signs. The patients were thoroughly informed about the study and gave their written informed consent. The study was approved by the hospital ethics committee.

Imaging findings: Computed Tomography (CT) revealed a mixed density mass consisting of fat and soft tissue within the esophageal lumen, accompanied by enlargement of the esophageal lumen. Notably, there was no thickening observed in the esophageal wall (Figure 1 Plain CT scan). Enhancement CT demonstrated heterogeneous enhancement of the lesion (Figure 2 Enhanced CT). Esophageal X-ray barium meal imaging revealed a significant filling defect in the middle segment of the esophagus, with an intact esophageal mucosa. The presence of the contrast agent between the tumor and the esophageal wall exhibited the characteristic smear sign (Figure 3 X-ray barium meal of the esophagus). Ultrasonographic gastroscopy findings indicated a smooth, elongated elevation on the posterior wall of the esophagus, located 19-38 cm from the incisors, accompanied by luminal narrowing.

Treatment Process: Upon admission, the patient underwent middle segment resection of the esophagus and lymph node dissection. The tumor was located approximately 17-38 cm from the incisor and measured approximately 21 cm in length and 6.5 cm in width. Macroscopically, the tumor exhibited a gelatinous appearance and a soft texture. There was no evidence of tumor invasion into the surrounding tissues,
with pleural swelling observed around the esophagus. Postoperatively, the patient received hemostasis, fluid replacement, and prophylactic antimicrobial therapy. The patient experienced a smooth recovery and was discharged 15 days after the surgery. At one-year follow-up after discharge, no tumor recurrence or other abnormal conditions were detected.

Pathological diagnosis: The pathological examination confirmed the diagnosis of well-differentiated liposarcoma (Figure 4). The immunohistochemical results showed positive staining for Vimentin, Ki-67 (5%), and S-100 (focal), while staining for Smooth muscle actin, Desmin, and Lysozyme was negative.

Discussion

Clinical manifestations and pathological features

Primary esophageal liposarcoma is commonly observed in elderly individuals over the age of 60 [5], albeit sporadically seen in adolescents. It exhibits a higher incidence in males compared to females. Predominantly, this malignancy is situated in the upper region of the esophagus [6], although there are occasional in the lower segment. The principal clinical features encompass the sensation of a foreign object in the esophagus, throat discomfort, and progressive dysphagia. In the specific case under consideration, a 41-year-old of middle age presented with a tumor located in the mid-segment of her esophagus. The primary clinical manifestation in this instance was difficulty swallowing, which aligns with the findings reported in existing literature.

Liposarcoma arises from primitive mesenchymal cells and immature adipocytes. As per the soft tissue classification outlined by the World Health Organization (WHO) in 2013, liposarcoma encompasses several subtypes [7-8]: well-differentiated liposarcoma, myxoid liposarcoma, dedifferentiated sarcoma, and pleomorphic liposarcoma. Well-differentiated liposarcoma and myxoid liposarcoma are categorized as low-grade malignant tumors, characterized by a low metastasis rate but a high of local recurrence. Conversely, dedifferentiated liposarcoma and pleomorphic liposarcoma are highly malignant tumors, prone to both recurrence and metastasis.

Among these subtypes, well-differentiated liposarcoma is the most prevalent histological type, accounting for 30% to 40% of all liposarcomas [9-10]. It is characterized by a low level of malignancy, favorable prognosis, absence of metastasis, although local recurrence and dedifferentiation may occur. The case at hand represents the most common variant of well-differentiated liposarcoma and exhibits the following marker characteristics: Vimentin (+), Ki-67 (+, 5%), S-100 (+, focal), Smooth muscle actin (-), Desmin (-), Lysozyme (-). These markers align with the well-differentiated subtype of the tumor.

Imaging manifestations

Esophageal x-ray barium meal examination findings: the esophageal lumen displayed noticeable expansion and dilation, with observable filling defects within. The barium contrast agent was observed to be situated between the tumor and the esophageal wall, exhibiting a distinct “smear sign.” The term “smear sign” is employed to describe the extensive contact observed between the contrast agent and the tumor during barium esophagography. This contrast agent coating serves as an indicator of adhesion between the tumor and the esophageal wall, or provides insights into the surface morphology of the tumor. The presence of the smear sign aids physicians in determining the precise location of the tumor within the esophagus and its relationship with the esophageal wall. Importantly, there were no indications of esophageal mucosa destruction observed in this case.

CT and MRI findings: Well-differentiated liposarcoma typically comprises a fat component that accounts for over 75% of the tumor volume, with non-fat components appearing as septate or focal nodules. The tumor manifests as a mass predominantly exhibiting fat density/signal, while the fibrous and solid components demonstrate densities and signals similar to that of muscle tissue. Blood supply to the tumor is often not extensive, with slight enhancement observed in the fibrous septa and solid components after contrast enhancement, while the fat component remains unenhanced. Mucinous liposarcoma, characterized by a fat component comprising less than 10-25%, displays a mass with density and signal reminiscent to water
on CT and MRI scans, with the presence of minuscule fat nodules proving crucial for diagnosis. These tumors demonstrate an abundance of dendritic vascular structures, resulting in pronounced enhancement on contrast-enhanced scans. Dedifferentiated sarcoma typically possesses a fat component ranging from 25-50%. On CT, it appears as a mass displaying a mixture of soft tissue and fat densities, with signals akin to that of muscle tissue. Additionally, ossification or calcification can occur within the tumor, indicating heterologous differentiation or metaplasia, which often corresponds to a poor prognosis [11]. The solid component of dedifferentiated liposarcoma exhibits significant enhancement after contrast administration. The degree of differentiation is typically low in pleomorphic liposarcoma, making it difficult to identify the fat component. These tumors are rich in blood vessels and are prone to hemorrhage, necrosis, and a poor prognosis, often resulting in misdiagnosis [12]. CT and MRI scans depict solid masses without fat density or signal, with evidence of liquefaction necrosis and significant enhancement after contrast administration. In the current case, X-ray barium meal of the esophagus revealed a substantial filling defect associated with a smear sign. CT imaging demonstrated a tumor primarily consisting of fat density, while no MRI examination was conducted case.

Differential diagnosis

The giant liposarcoma in the esophagus in our case required differentiation from other esophageal conditions, such as giant leiomyoma, pedunculated polyps, intraluminal esophageal carcinoma, and polyloid sarcoma. First giant leiomyomas of the esophagus are typically found in the middle and lower segments of esophagus. X-ray barium meal examinations reveal a significantly dilated esophageal lumen, with a lobulated eccentric filling defect observed within the lumen. The contrast agent flows smoothly between the esophageal wall and the tumor, creating a characteristic ring-like sign. Additionally, the esophageal wall is compressed and thinned, and there may be liquefaction necrosis and calcification within the tumor. Secondly, pedunculated polyps of the esophagus often exhibit movement during swallowing and deep breathing, and these polyps are prone to ulceration and bleeding. Thirdly, intraluminal esophageal cancer damages the adjacent esophageal wall and mucosa. Lastly, polyloid sarcoma of the esophagus presents as irregular lobulated soft tissue masses, which are prone to ulceration and early hematogenous metastasis.

Treatment

Surgical resection stands as primary treatment method for esophageal liposarcoma [13]. Depending on the tumor's size and location, options such as partial esophagectomy, or combined resection other organs can be considered. The primary objective of resection is a complete eradication of the tumor while preserving the esophageal function. Given the rarity of esophageal liposarcoma, most documented cases are reported as isolated instances, with limited systematic follow-up. Consequently, the prognosis for esophageal liposarcoma following curative surgery remains largely unknown. The implementation of adjuvant radiotherapy and chemotherapy remains a subject of debate, necessitating close monitoring. In the presented case, the patient underwent tumor resection and lymph node dissection in the middle segment of the esophagus. After one year of follow-up, the patient currently demonstrates favorable clinical status. However, continued long-term observation is warranted.

In conclusion, despite its rarity, esophageal liposarcoma presents certain distinctive imaging features that can assist in preoperative qualitative diagnosis. Surgical resection remains the primary approach for managing esophageal liposarcoma.

Conflict of Interest: The authors declare that they have no conflict of interest.

For this type of study formal consent is not required.

This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent: Informed consent was obtained from all individual participants included in the study.

References


Figure 1 Plain CT scan shows that the lumen of the esophagus is enlarged, and there is a mass of fat and soft tissue density, but no thickening of the esophageal wall.

Figure 2 Enhanced CT scan showing heterogeneous enhancement of the mass.

Figure 3 X-ray barium meal of the esophagus shows a huge filling defect throughout the esophagus without damage to the esophageal mucosa, and the barium is located between the tumor and the esophageal wall, showing a smear sign.

Figure 4 Postoperative pathology shows that the tumor consists of differentiated mature adipocytes, a small amount of mesenchymal cells, and blood vessels. HE ×100