Huge Myxoid Liposarcoma of the Esophagus: A Case Report

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Abstract- Although esophageal liposarcoma is an extremely rare tumor, liposarcoma is the most common soft tissue sarcoma in adults. Liposarcoma is currently classified into the types of well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated liposarcoma. Up to now only a few cases of esophagus liposarcoma have been described in the world literature. We describe a myxoid type liposarcoma of the esophagus in a 68 year old man presented with hoarseness and intermittent dysphagea to solid food. He had a huge mass in his mouth which was mobile with gag reflex. A barium swallow, esophageal manometery and CT scan of the esophagus have not clearly revealed the mass. After endoscopic surgical resection of the tumor the histological examination revealed a myxoid liposarcoma. Both the presenting signs and symptoms and the histology type are rare for such tumor. This case demonstrate a rare differential diagnosis of intermittent dysphagia as early diagnosis is so important in those tumors and should kept in mind them, although they are quite rare.

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Introduction

Liposarcoma is still a relatively rare tumor (1) but it is the ubiquitous soft tissue malignancy. It is the most common soft tissue tumor of the lower extremity (2) and also the most common sarcoma of the retroperitoneum (3). Although liposarcoma most frequently involve those sites, other sites have been reported such as the mediastinum and head and neck (4). Liposarcoma rarely arises in the gastrointestinal tract and the incidence of gastrointestinal tract involvement is 0.1% to 5.8% at autopsy and esophagus is the least common location (1.2% to 1.5% of all gastrointestinal lipoma) (5).

Histopathological classification of liposarcoma has been diverse because of its complex histological components (6, 7). Enzinger and Weiss (5) classified it into five basic histological categories, namely, well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated, which had previously been subclassified under "well-differentiated," with a footnote that very occasional cases (5%–10%) show a combination of two or three components (so-called mixed-type liposarcoma). Primary liposarcoma may arise wherever adipose tissue is present. The tumor usually involves older individuals with male predominance (2). Aggressively wide excision with or without local radiation recommended for well-differentiated and myxoid liposarcoma arising in more usual sites.

Primary liposarcomas of the esophagus are extremely rare and usually slow growing, arising from mucosa and submucosal esophageal layers (8, 9). The first case of primary esophageal liposarcoma was described by Mansour *et al*. in 1983 (9). Up to 2006, 14 cases of esophageal liposarcoma have been reported, with the age range of 43-73 years and a male-female ratio of 1 to 1.1, which is consistent with the slight male predominance observed in the soft tissue liposarcomas (10,11). The predominant clinical symptoms included progressive dysphagea, nausea, throat discomfort, retrosternal pain, odynophagea, respiratory distress, anemia, tumor regurgitation and sudden death by

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asphyxia and foreign body sensation (12). The first case of pedunculated myxoid liposarcoma of esophagus was reported in a 53-year-old with respiratory rather than gastrointestinal symptoms (9). In this report, we described a large liposarcoma of the esophagus in a man which was presented predominantly with hoarseness and intermittent dysphagea. After surgical resection, the histological examination revealed a myxoid liposarcoma.

Case Report

A sixty-eight year old Persian man who lived in Tehran was presented to the surgery clinic of Imam Khomeini Hospital affiliated to Tehran University of Medical Sciences in September 2006, with a 3 years history of increasing hoarseness and intermittent dysphagea with solid food. He also complained of nausea and vomiting. In physical examination a huge pedunculated pink mass was found in his mouth (just over the tongue) which was moving with gag reflex and coming up and down out of the mouth. The palpable mass and the symptoms disappeared when patient was swallowing (Figure 1). He had no history of shortness of breath, no past history of pulmonary disease, gastrointestinal bleeding, weight loss and fever. He was a chronic hepatitis B carrier and had a 5 years history of cirrhosis. He also had a history of lower gastrointestinal bleeding two years ago which treated conservatively. In physical examination although no mass was seen primarily but after gag reflex stimulation a non-vegetative pink mass with smooth surface appeared in the mouth, Figure 1. True vocal cord (TVC) motion was normal. A barium swallow, esophageal manometery and CT scan of the esophagus have not clearly revealed the mass. The remaining physical examination was also normal and the patient appeared otherwise healthy condition.

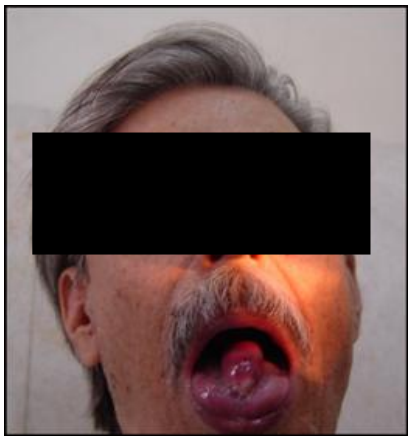

Figure1. Huge pink mass which was moving with gag reflex

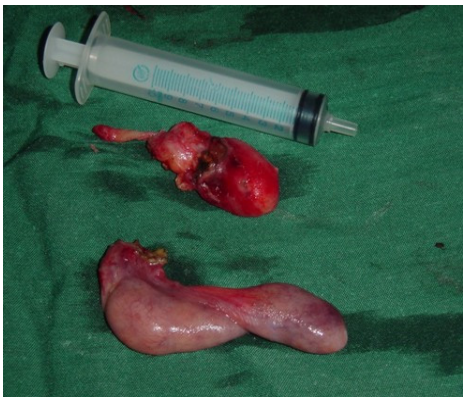

Figure 2. A smooth pink oval 17×3×2 centimeter mass with intact surface was seen in the posterior wall of the esoghagus and another 10×3×2 cm mass was found in 2cm lower than the first one with the same characteristics

The patient's laboratory assessment was within normal limit. He was scheduled for esoghagoscopy and laryngoscopy and underwent laryngoscopy at first visit. The laryngeal elements were normal. On esophagoscopic examination a smooth pink oval 17×3×2 centimeter mass with intact surface was seen in the posterior wall of the esoghagus. In addition in the middle and lower part of the esophagus four columns of grade II varices were identified. The patient underwent surgery under general anesthesia. The base of the mass was hold by clips and coagulated with coutery. Another 10×3×2 cm mass was found in 2cm lower than the first one with the same characteristics and both of them were removed, Figure 2.

The patient was discharged after 8 days admission. Radiological follow up with chest X-ray and upper GI Endoscopy showed no recurrences or metastasis until the 12th months. After 12 months, barium swallow was performed and no intrathoracic, mural and intramural esophageal abnormalities have been found. Follow-up endoscopic examinations were performed in 3, 6 and 9 months later which have shown no recurrence. In the last upper GI endoscopy there were 2 columns of grade I and two columns of grade II and some sclerosed varices, without active bleeding .After one year follow up the patient was in good condition without any complication.

Pathology

The microscopic appearance showed squamous mucosa with a delineated myxoid tumor in the subepithelial stroma. The tumor consisted of ovoid plump spindle cells with scattered predominantly mono and rarely multivacuolated fat cells and typical plexiform vascular pattern.

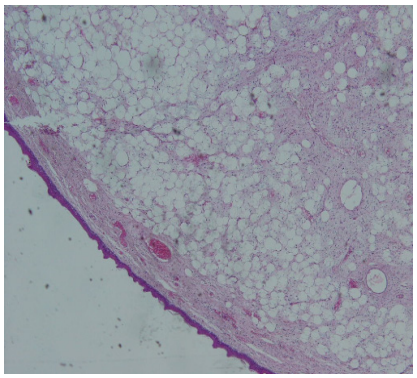

Figure 3. The pathologic appearance of the mass

Isolated moderately polymorphic nuclei were also noted. In some other areas there are lobules of mature adipose tissue with scattered trabeculation of myxoid spindle-cells. The mucosal epithelium was intact and showed no atypia, Figure 3.

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Discussion

Although liposarcoma is the most common tumor of mesenchymal origin in adult, its esophagus origin is extremely rare. Our report is an unusual case of primary liposarcoma with a myxoid pathology. Among the reported liposarcomas of the esophagus, a few subclassified as myxoid (8, 10). Although squamous cell carcinoma is a common malignant tumor of the esophagus but other malignant tumors such as fibrosarcoma, leiomyosarcoma, rhabdomyosarcoma, pseudosarcoma, carcinosarcoma, and reticulum cell carcinoma have been reported. According to the previous studies (13,14) this group of sarcomas consisted only 0.5% of all primary esophageal malignant tumors. However due to clinical presentation and special features of the tumor in our case it differed to common esophageal neoplasm's. Although in a case of esophageal cancer marked abnormalities were seen in liver function during viral infection and deterioration of the hepatic disorder during viral infection and surgical aggression (16); in our case there was no significant change in hepatic enzyme or markers was identified after operation. Surgery is the only diagnostic method of the esophageal malignancies such as liposarcoma. Different options of treatment have been advocated. This treatment varies from simple enucleation or endoscopic resection to partial or total esophagectomy or trans-cervical, transthoracic, transgastric resections (8-11). The most reliable prognostic factors are liposarcoma grade, histological subtype, location and sufficiency of surgical treatment (11, 15). As in our case the treatment of choice is surgery and surgical intervention in this condition seems to favor the quality of life and prolong survival. Based on the location of tumor surgical approach may be differed. In an interesting report by Arteaga *et al*., they resected a large tumor in a 72 years old man through left cervicotomy with thoracoscopic assistance (17).

Esophageal liposarcoma can be diagnosed if a patient has a history of slow growing esophageal mass with a low tumor density in computerized tomography (CT) in combination with surgical resection and histological examination. Upper aerodigestive tract liposarcoma especially esophageal types are very rare and the rate of metastatic disease and mortality rate is low. However, high recurrence rates may be presented in these tumors especially when less radical surgery is in use. The present case was treated by a conservative endoscopic surgery and no metatstasis and relapse were found during follow up. Also this treatment was not affected the grade of varices. In conclusion, this rare case of esophageal liposarcoma demonstrates an important different diagnosis for intermittent dysphagia as diagnostic delay is a very ominous event. Multicentricity of this case describes the importance of examination enteric length of organ due to synchronous tumors.

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