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Imaging Findings of Giant Liposarcoma of the Esophagus

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A giant esophageal liposarcoma showing rapid growth over 7 months is presented in 56-year-old man. It originated from the pharyngo-esophageal junction with a short stalk, and extended downward to the distal esophagus. A barium swallow study showed a large, sausage-like intraluminal mass in the dilated esophagus. CT and MR imaging showed a heterogeneous mass with a fatty component in the esophagus. A total laryngopharyngo-esophagectomy was performed and the histological diagnosis was of a well-differentiated liposarcoma.

Key Words: Esophagus, liposarcoma, CT, MR, esophagography

INTRODUCTION

Although the predominant malignant tumor of the esophagus is squamous cell carcinoma, a variety of sarcomatous tumors are occasionally noted, though they account for less than 0.5% of all primary malignant esophageal tumors.[1] Gastrointestinal lipomatous tumors have an incidence of 0.1 to 5.8% at autopsy[2] and the esophagus is the least common location for these tumors, and is only found in 1.2 to 1.5% of all gastrointestinal lipomas.[2]

Esophageal liposarcomas less than 10 cases have been reported in the literature,[3-7] and most have originated from the cervical esophagus. Both benign and malignant esophageal lipomatous tumors usually grow as polypoid masses, and show few symptoms until the tumor has attained a considerable size.[3-7] Liposarcoma is known to originate from primitive mesenchymal cells rather than mature adipose tissue; the presence of normal fat tissue is not a prerequisite of its development.[5]

We report the imaging features of a giant esophageal liposarcoma that showed rapid interval growth over a period of 7 months.

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CASE REPORT

A 56-year-old man was initially admitted to our hospital because of throat discomfort and a voice change over 6 months. He also complained of mild but progressive dysphagia with a foreign body sensation. A physical examination revealed a supraglottic soft tissue mass with a slightly prominent covering vein. Laboratory data were normal.

Initial esophagography showed a lobulated mass involving both pyriform sinuses (more prominent in the left side) in the hypopharynx, which extended downward into the proximal esophagus with a tongue-like projection (Fig. 1A). Esophagoscopy revealed a lobulated soft tissue mass with slightly firm consistency in the throat. Computed tomography (CT) revealed an ill-defined fat containing mass in the left pyriform sinus area and well-circumscribed fat containing mass within the cervical esophageal lumen. The mass was diagnosed as a liposarcoma in the hypopharynx via peroral biopsy. The patient refused surgical removal of the mass because he feared voice loss from laryngectomy.

After 7 months, he was admitted again for operation due to an aggravation of the dysphagia. Follow-up esophagography showed a more

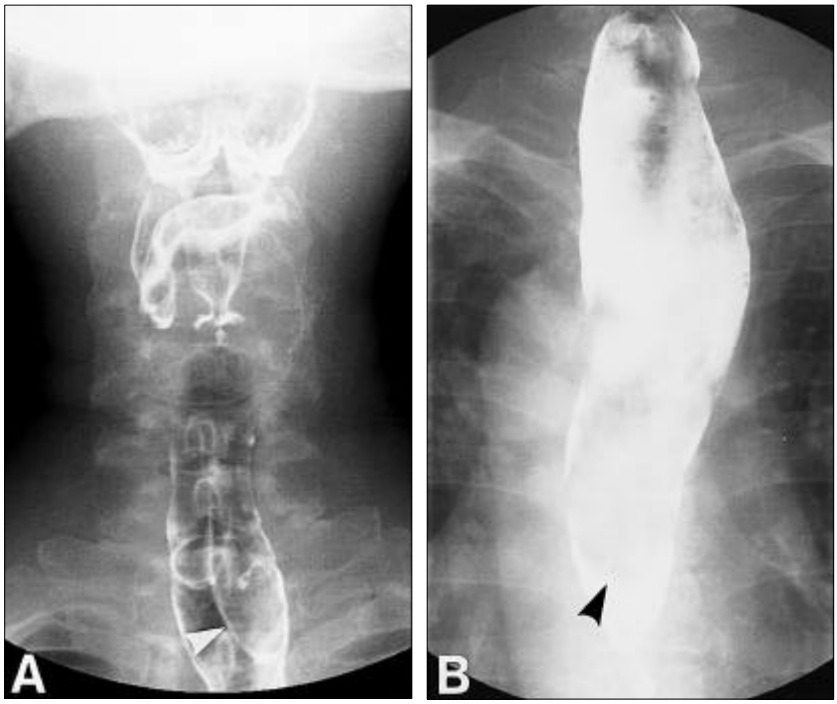

Fig. 1. (A) Initial esophagogram showing a lobulated mass involving both pyriform sinuses (more prominent in the left side) in the hypopharynx, and extending downward to the second thoracic vertebral body level with a tongue-like projection (arrowhead). (B) A follow-up esophagogram taken in the prone position after 7 months later showed a more downward growing mass (arrowhead) within the megaesophagus, extending to the eighth thoracic vertebral body level with a sausage-like appearance.

Fig. 2. Postcontrast CT scan revealed a well-circumscribed low attenuation mass in the dilated esophagus at the level of the aortic arch.

downward growing mass within the megaesophagus, extending to the eighth thoracic vertebral body level (Fig. 1B). Follow-up CT showed a more enlarged and downward extending fat containing mass in the dilated esophagus (Fig. 2). Magnetic resonance (MR) images demonstrated an inhomogeneous signal intensity mass with internal high signal intensity foci in the dilated esophagus on in-phase T1-weighted images (TR/TE, 150/4 msec, flip angle 80°, section thickness 8-10 mm) (Fig. 3A), and a decreased signal intensity of the high signal intensity foci on out-of-phase T1-weighted images (TR/TE, 150/2.2 msec, flip angle 80°, section thickness 8-10 mm) (Fig. 3B), indicating a fat containing mass. After the administration of gadolinium, the mass was inhomogeneously enhanced.

The patient underwent total laryngopharyngoesophagectomy and reconstruction with gastric substitution. The large and long soft tissue mass originated from the posterior wall of the pharyngo-esophageal junction with a short stalk, which extended downward to the distal esophagus (Fig. 4). The maximal size of this tumor was 21 × 6 × 2 cm (craniocaudal × transverse × sagittal).

DISCUSSION

Although gastrointestinal lipomatous tumors are predominantly found in the distal ileum and in the large bowel, the esophagus is the most infrequent alimentary tract location.[8] Esophageal liposarcomas are exceedingly rare and slow growing tumors that arise from the mucosal or submucosal layers of the esophagus.[3-7] All 10 cases of this malignant tumor reported to date

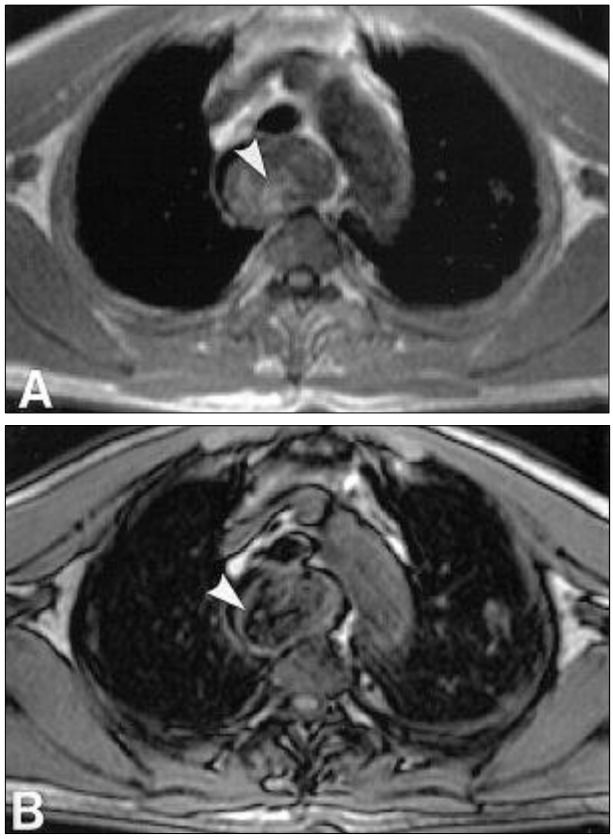

Fig. 3. (A) In-phase T1-weighted MR image showing an inhomogeneous low signal intensity mass with internal high signal intensity foci (arrowhead) in the dilated esophagus. (B) Out-of-phase T1-weighted MR images showing decreased signal intensity (arrowhead) of the high signal intensity foci within the esophageal mass, indicating a fat containing mass.

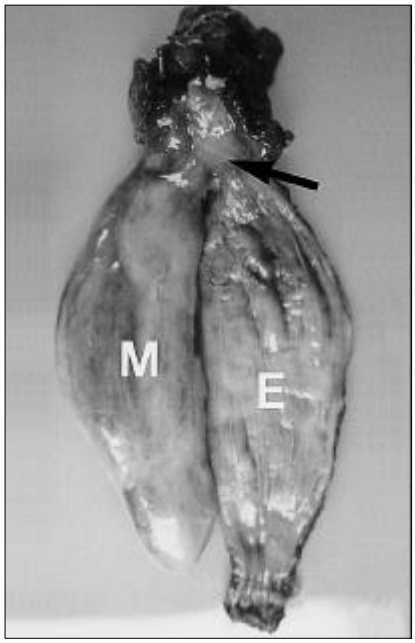

Fig. 4. Gross specimen demonstrating a large and elongated mass (M) arising from the posterior cricoid wall with a short stalk (arrow) in the pharyngo-esophageal junction. The maximal extent of this tumor was 21 × 6 × 2 cm. E=esophagus.

have been pedunculated tumors, similar to other esophageal mucosal tumors.[3-7,9] Pedunculated tumors may become large and occupy the entire esophageal lumen.

Esophageal benign tumors usually arise from the lower portion of the esophagus, however, pedunculated tumors commonly arise from the cervical esophagus.[4] The wall of the cervical esophagus is relatively thin and habitually approximated by tonic muscular contractions. Hypothetically, these tumors may arise from the mucosal fold or as a small tumor, which is gradually forced downward and is elongated by peristaltic waves.[9] These intraluminal pedunculated tumors are largely mesenchymal in origin.

At barium swallow study, lipomatous tumors may indeed be confused with the more usual esophageal pathologies such as megaesophagus or bezoars, and cannot be easily distinguished from other more common extramucosal tumors such as leiomyomas,[7] because the normal swallowing mechanism and the unobstructed passage of barium through the whole of the esophagus can cause misdiagnoses.[9] Moreover, misdiagnoses, such as achalasia, have also been made after both radiological and endoscopic examinations, because this tumor has a soft consistency, is pliable and has a smooth surface, nearly always covered with normal esophageal mucosa. The only reliable diagnostic method for esophageal lipomatous tumors is surgical removal and histological examination.[7]

This is the first reported case of an esophageal liposarcoma, which showed rapid growth over a period of 7 months. Initially this tumor had a craniocaudal length of 9 cm, which increased to 21 cm over 7 months, with corresponding marked increases in transverse and sagittal distances. The barium swallow study showed the megaesophagus with a large filling defect and focal irregular stalk-like adhesion in the pharyngo-esophageal junction. It is known that the attachment of the

stalk is frequently not identified, as it is often thin. CT and MR images may give added information from tissue density (i.e., fat tissue of low density on CT image, high signal intensity on in-phase T1-weighted image and low signal intensity on out-of-phase T1-weighted MR images), and the detection of characteristic adipose tissue that can be helpful in diagnosing lipoma or liposarcoma. Moreover, MR direct multiplanar images can enhance the likelihood of precisely locating the tumor.

Although the radiological appearance of giant esophageal liposarcoma may be similar to that of other tumors, such as giant esophageal polyps, the histological diagnosis may be variable, and includes lipomas, fibromas, fibrolipomas, myxolipomas, leiomyomas, fibrosarcomas, and leiomyosarcomas.[9] Some fatty tumors among these can be easily differentiated from non-fatty tumors, such as, leiomyomas and fibromas, on CT and MR images. However, the fatty esophageal tumors can be differentiated only by histological diagnosis. Only 20% of the esophageal tumors are benign and the majority is non-epithelial. Leiomyoma is the most common extra- and intramural non-epithelial tumor and the fibrovascular polyp is the most common intraluminal non-epithelial tumor of the benign esophageal tumors. Most liposarcomas arise in the cervical esophagus or, as in this case, within the hypopharynx at the level of the cricoid cartilage.[7]

In conclusion, a clinical history of a rapid growing esophageal mass, with no sign of either local or distant spread, and a typical low-density appearance on the majority of CT sections or high signal intensity on in-phase T1-weighted MR images, may suggest a diagnosis of esophageal lipoma or liposarcoma.

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