CASE REPORT

Adult Asymptomatic Hamartoma in the Distal Esophagus: A Rare Case

Wen Ming Wu, Xiang Dong Wang, Gang Sun, Ling Hu En Qiang and Yun Sheng Yang

Abstract

An esophageal hamartoma is a rare, benign germ-cell condition. Most hamartomas are intraluminal tumors located in the upper third of the esophagus. We herein report an unusual case of a pedunculated hamartoma that involved the lower third of the esophagus that was diagnosed incidentally during investigations for epigastric pain. The features noted on endoscopy, endoscopic ultrasound/Doppler endoscopic ultrasound and computed tomography of the chest are also presented.

Key words: hamartoma, esophagus, adult, male, endoscopic, ultrasonography

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Introduction

A hamartoma is an uncommon congenital abnormality that can contain a range of structures, from primitive somatic tissues to highly organized structures, and is heterogeneously cystic or solid (1). Since the first case of an esophageal hamartoma was described in 1963 (2), only 11 cases have been reported in the worldwide literature, six arising in the upper third and one in the lower third of the esophagus. All of the tumors have been located in the lumen, except for one that was intramural. Most have occurred in infants and children. In previously reported cases, the tumors were confirmed during surgery. We herein report a rare case of an adult with a tumor located in the lower third of the esophagus and in whom the diagnosis was made using multimodality imaging tools.

Case Report

A 40-year-old man presented with epigastric pain after drinking. The patient’s past history showed only an occasional choking feeling, with no difficulties in swallowing. At our outpatient clinic, upper endoscopy demonstrated an irregular esophageal mass located approximately 31-36 cm from the incisor. The mass consisted of two adjacent ovoid cystic tumors with a broad fibrous band that were found to originate from the lateral esophageal wall and extend into the lumen. The surface of the mass was covered with sparse hair (Fig. 1). Subsequent endoscopic ultrasonography (EUS; UM-2R, Olympus Optical, Tokyo, Japan) revealed a hyperechoic, cystic structure in the upper portion of the lesion, while the lower portion was formed of multiple separated cystic masses; partial muscularis propria was absent (Fig. 2). In addition, Doppler endoscopic ultrasound (GF-UE260, Olympus Optical) demonstrated distinct arteries in the tumor (Fig. 3). Chest computed tomography (CT) with or without contrast medium confirmed a lower esophageal mass measuring 2.6 cm with fat and calcification, suggestive of an esophageal hamartoma (Fig. 4). The radiographic findings were negative.

The results of a physical examination were normal. The only abnormal laboratory test was the detection of a positive expression of hepatitis B surface and core antibodies. We suggested performing thoracotomy; however, the patient refused the operation due to the potential for surgical injury and because he had been asymptomatic for many decades. The patient was discharged on day 6 and remains well to date.

Discussion

Benign esophageal tumors are rare, accounting for fewer than 1% of all esophageal neoplasms. These tumors are di-
provided into three categories based on location in relation to the esophageal wall: intramural, extramural or intraluminal. Only one-third of these lesions are pedunculated. A hamartoma is a benign, focal malformation that originates in pluripotent cells associated with the development of the foregut and tracheobronchial groove. Hamartomas contain diverse tissues that are foreign to the organ or anatomic site at which the hamartoma arises. Hair, teeth, bone, mucous glands, muscle, cartilage and adipose tissue have all been observed in such lesions. Due to its rarity, there are no epidemiological data regarding the occurrence of this disease in the esophagus. Limited data have been published as case reports, scattered in sometimes obscure or relatively inaccessible journals (Table) (2, 4-13). In most of the reported cases, the hamartoma has arisen from the proximal third of the esophagus and belonged to the category of intraluminal tumors. Most lesions have been discovered as a result of the patient’s clinical presentation, for example, the presence of obstruction due to intraluminal growth (4, 11) or the regurgitation of a pedunculated tumor (9, 13). Other tumors have been vague or asymptomatic and discovered incidentally in late childhood or adulthood (2, 5, 10), as in the case of the current patient. In the present case, the mass arose in the lower third of the esophagus and only occasionally caused a choking feeling; it was not discovered until the patient underwent esophagogastroscopy incidentally. We presume that the lack of symptoms is the result of the slow, gradual growth of these tumors, combined with the ability of the esophagus to dilate and accommodate such expansion. The indolent nature of these lesions is also common among other benign tumors, which can remain stagnant for years (3).

Esophageal teratomas may be detected on either CT or magnetic resonance imaging (MRI) at any age; these imaging modalities can be used to delineate the size and location of the tumor. CT is particularly useful for identifying fatty and calcified tissue within tumors (14). If possible, the mass should be visualized on endoscopy. In the present case, EUS provided important information for preoperative planning, including the location of the pedicle and the vascularity and tissue elements of the mass. Generally, surgical resection is the standard treatment for hamartomas. The procedure is usually performed via esophagotomy or thoracotomy, as ap-

Figure 1. Upper endoscopy demonstrated an irregular esophageal mass consisting of two adjacent ovoid cystic tumors with a broad fibrous band that were found to originate from the lateral esophageal wall and extend into the lumen. The tumor surface was covered with sparse hair.

Figure 2. Endoscopic ultrasonography (UM-2R, Olympus Optical, Tokyo, Japan) of the mass. (A) A hyperechoic, cystic structure (arrows) was observed in the upper portion of the lesion. (B) Doppler endoscopic ultrasound (GF-UE260, Olympus Optical) revealed that the lower portion of the lesion was septated with cystic masses, while the partial muscularis propria (wide hollow arrow) was absent.

Figure 3. (A) Color Doppler endoscopic ultrasound (GF-UE260, Olympus Optical) showed a well-defined mass (white arrows) with distinct arteries (dotted circle) in the tumor and a regional blood flow. (B) Pulsed-wave Doppler ultrasound suggested that the blood vessels were arteries.
A limitation in this case is that the lesion was not assessed using tissue pathology. Although such examinations are optimal for diagnosis, imaging technologies, such as esophagoscopy, EUS/Doppler EUS, CT and MRI, are effective in making a diagnosis and can provide useful information.

The authors state that they have no Conflict of Interest (COI).

References


Table. Clinical Features of Patients with Oesophageal Hamartoma

<table>
<thead>
<tr>
<th>Year of publication (Reference)</th>
<th>Age/sex</th>
<th>Initial symptom</th>
<th>Location</th>
<th>Histopathologic finding</th>
<th>Surgical procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1963[2] 61Y/male</td>
<td></td>
<td>Difficulty in swallowing solids</td>
<td>Upper oesophagus /Intraluminal</td>
<td>Fibro-adipose connective tissue intermingled with mucous glands and cartilage</td>
<td>Right lateral oesophagotomy</td>
</tr>
<tr>
<td>1975[5] 60Y/male</td>
<td></td>
<td>Vague midchest pain</td>
<td>Cervical esophagus /Intraluminal</td>
<td>Cartilaginous tissue, glandular structures and adipose tissue</td>
<td>Esophagotomy</td>
</tr>
<tr>
<td>1980[8] Infant[11m]/Male</td>
<td></td>
<td>Dysphagia and drooling</td>
<td>Upper oesophagus /Intraluminal</td>
<td>Skeletal muscle fibers, fibrous connective tissue and hyaline cartilage</td>
<td>Surgically removed</td>
</tr>
<tr>
<td>1993[12]</td>
<td></td>
<td></td>
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For some benign tumors with thin pedicles, endoscopic excision (e.g. electrocautery) may be the approach of choice (4, 13). In this case, the thick pedicle, deep continuity into the esophageal wall and abundant blood supply indicate that the tumor was not a candidate for minimally invasive procedures, resulting in the recommendation for an open surgical technique.

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