Calcifying fibrous pseudotumor (CFT) is an uncommon lesion that was first described by Fetsch et al., and is histologically characterized by hyalinized collagen tissue with focal lymphoplasmacytic infiltrate, and psammomatous and dystrophic calcifications. It can develop at any site, but esophageal cases are extremely rare and have never been reported before. A 54-year-old woman underwent upper gastrointestinal endoscopy and endoscopic ultrasound because of intermittent dysphagia. The results showed an isoechic esophageal submucosal tumor over the deep mucosa and submucosal layers, with calcifications inside. The patient underwent tumor excision, and the diagnosis was confirmed by pathological features, with abundant collagen, calcification and inflammatory cell infiltration. She received regular follow-up at the clinic and no evidence of tumor recurrence was found.

Key Words: calcification, esophagus, fibrous tumor

CASE REPORT

Calcifying Fibrous Pseudotumor of the Esophagus

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Calcifying fibrous pseudotumor is an uncommon lesion and has recently been recognized as a distinctive fibrous lesion. Esophageal calcifying fibrous pseudotumor is extremely rare and, to the best of our knowledge, has never been reported before. A 54-year-old woman underwent upper gastrointestinal endoscopy and endoscopic ultrasound because of intermittent dysphagia. The results showed an isoechic esophageal submucosal tumor over the deep mucosa and submucosal layers, with calcifications inside. The patient underwent tumor excision, and the diagnosis was confirmed by pathological features, with abundant collagen, calcification and inflammatory cell infiltration. She received regular follow-up at the clinic and no evidence of tumor recurrence was found. [J Chin Med Assoc 2010;73(11):599–601]

Key Words: calcification, esophagus, fibrous tumor

Introduction

Calcifying fibrous pseudotumor (CFT) is an uncommon lesion that was first described by Fetsch et al., and is histologically characterized by hyalinized collagen tissue with focal lymphoplasmacytic infiltrate, and psammomatous and dystrophic calcifications. It can develop at any site, but esophageal cases are extremely rare and have never been reported previously, to the best of our knowledge. We present a case of CFT that developed in the esophagus, with specific imaging and pathological characteristics.

Case Report

A 54-year-old woman visited our hospital because of an esophageal tumor found accidentally by upper gastrointestinal (UGI) endoscopy at another hospital. She could not recall nausea, retrosternal pain, body weight loss or poor appetite, but she had experienced a foreign body sensation in the throat for several weeks. On physical examination, the patient had a calm appearance. No lymphadenopathy or splenomegaly was found by palpation. Laboratory data demonstrated 14 g/dL hemoglobin (normal, 11.3–15.3 g/dL), 2 ng/mL carcinoembryonic antigen (normal, < 5 ng/mL), and 0.3 ng/mL squamous cell carcinoma antigen (normal, < 1.5 ng/mL). Chest X-ray and abdominal sonography showed no active lesion.

Repeated UGI endoscopy found an esophageal submucosal tumor, 4 cm in size, which was located 1 cm above the esophagocardiac junction (Figure 1). Endoscopic ultrasound (EUS) revealed an isoechic tumor over the deep mucosa and submucosal layers, with hyperechic lesions inside. The muscularis propria layer was intact, without tumor invasion (Figure 2). Mediastinal computed tomography revealed a thickened esophageal wall with calcification (Figure 3). Esophageal carcinoid tumor was suspected initially, and surgical intervention was suggested.

The patient underwent surgical tumor excision, and the specimen was found to be composed of abundant collagen, with calcification and inflammatory cell infiltration (Figure 4). These cells were immunohistochemically negative for CD32, CD117, S-100 protein and desmin. A diagnosis of CFT of the esophagus was confirmed by specific imaging and pathological features.

The patient received regular follow-up at the clinic and no evidence of recurrent tumor was noted by UGI endoscopy and mediastinal computed tomography for 1 year after tumor excision.
Discussion

CFT is rare and has recently been recognized as a distinctive fibrous lesion with specific characteristics. Both sexes are equally affected, and although age distribution ranges from 1 to 65 years, children and young adults are affected most often. It can develop at a wide range of anatomic locations, including the extremities, trunk, neck, mesentery, mediastinum, and paratesticular area, but esophageal cases are extremely rare. A previous case report demonstrated calcification with a crown-like shape in the gastric submucosal layer by EUS, but it might have been undifferentiated from other similar lesions, such as solitary fibrous tumor and inflammatory myofibroblastic tumor. CFT can be suspected early by the typical

Figure 1. Endoscopy of the esophagus shows a submucosal tumor.

Figure 2. Endoscopic ultrasound of the lower esophagus shows a submucosal tumor originating from the deep mucosa and submucosal layers (2nd and 3rd layers), with: (A) intact muscularis propria (4th layer); and (B) calcification inside.

Figure 3. Computed tomography of the mediastinum reveals thickening of the esophageal wall with calcification inside (arrow).

Figure 4. Pathological examination shows dense fibrous tissue, inflammatory cell infiltration, and psammomatous and dystrophic calcification (arrow).
EUS characteristics: an isoechoic tumor originating from the deep mucosa and submucosal layers. The differential diagnosis is carcinoid tumor, for which the mainstay of treatment is surgery.

An adequate diagnosis of CFT depends on pathological studies from local excision of this tumor. Pathological features include a non-encapsulated, densely hyalinized, fibrotic proliferation, with slight infiltration of lymphocytes and plasma cells. Two types of calcifications have been described, including psammomatous and dystrophic, but a previous study reported that calcifications exist in only 5% of cases. Treatment is surgical excision, although therapeutic recommendations have not been established to date due to the relatively small number of cases. Malignant transformation of CFT has not been reported, and it is a recognizable entity with good prognosis.

We have described an esophageal CFT that presented as a submucosal tumor with calcification inside. Definite diagnosis relies on typical pathological characteristics observed after local excision of the tumor.

References