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Case Report:

The value of endoscopic ultrasonography on diagnosis and treatment of esophageal hamartoma

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Abstract: Objective: To examine the values of endoscopic ultrasonography (EUS) on diagnosis and treatment of esophageal hamartoma. Methods: We compared and analyzed various kinds of imaging examinations such as barium esophagram, contrast-enhanced computed tomography (CT) and conventional gastroscopy in retrospectively reviewing the clinical data of an esophageal hamartoma patient seen in our clinic in the First Affiliated Hospital, School of Medicine, Zhejiang University, Hangzhou, China. Having received various imaging examinations, this patient was finally diagnosed with esophageal hamartoma and underwent gastroscopic resection of hamartoma with the diagnostic information obtained from EUS. The patient had been regularly followed up for 13 months after treatment. Results: Barium esophagram, CT and conventional gastroscopy detected the lesion, but were unable to distinguish it from common esophagopolypus and other submucosal lesions, and unable to determine etiopathogenisis. EUS detected the hamartoma and identified its internal structure, echo, exact size, depth of invasion, origin and the relationship between adjacent tissues and organs, differentiating the lesion from other submucosal tumors and clearly defining the diagnosis. EUS-guided fine needle aspiration (FNA) also helped to identify the etiological diagnosis. Conclusion: EUS was superior to other imaging means in diagnosis and treatment of hamartoma.

Key words: Esophageal hamartoma, Endoscopic ultrasonography (EUS), Gastroscopic resection
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INTRODUCTION

"Hamartoma" was first brought forward by Albrecht in 1904 (Wang, 1992), and can be originated from any tissue. Esophageal hamartoma is rare (Song *et al*., 2004), and its clinical manifestation and auxiliary examination such as barium esophagram, contrast-enhanced computed tomography (CT) and conventional gastroscopy are not specific, adding a difficulty to make an accurate preoperative diagnosis. Endoscopic ultrasonography (EUS) can clearly show the structure of gastrointestinal wall, and identify the gastrointestinal lesion's surface and internal structure, echo, exact size, depth of invasion, origin, as well as the relationship between adjacent tissues and organs (Chen *et al*., 2001; Gress *et al*., 2001; Fusaroli and Caletti, 2005). EUS can gain more diagnostic information about submucosal lesions, which is helpful in differentiating hamartoma from other submucosal tumors. In this study, we report the procedure of diagnosis and treatment of a patient with esophageal hamartoma who was seen in our clinic in the First Affiliated Hospital, School of Medicine, Zhejiang University, Hangzhou, China, to examine the value of EUS on diagnosis and treatment of hamartoma.

CLINICAL DATA

A 45-year-old male with a 20-year history of foreign body in the esophagus had developed a neoplasm 1.5 cm in diameter in his esophagus 20 years before. The neoplasm could protrude 2 cm outside of his mouth when vomiting and could retract into the esophagus by itself. He had frequently heartburn behind the sternum when hungry, but no chest distress,

chest pain, haematemesis, dysphagia, fever and chilly. The patient was hospitalized in our clinic for further diagnosis and treatment. Physical examination showed that there was no abnormal finding except for a long-shape neoplasm getting out of his mouth when he nauseated (Fig.1, see Page 664); the neoplasm could retract into the esophagus by itself. Blood, stool and urine routines, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), liver and kidney functions, and prothrombin time (PT) were all normal. Serum tumor markers were in the normal range, and anti-neutrophil cytoplasmic antibodies (ANCA) and anti-nuclear antibody (ANA) were also negative.

Chest X-ray showed no abnormality. Barium esophagram indicated a little left-bias flow sign when passing through the upper esophagus and a filling defect on the right of the esophagus (Fig.2). The filling defect was about 11.5 cm long, with distinct and smooth border; esophageal peristalsis was normal and mucous membrane appeared to be not obviously destroyed or interrupted. The diagnosis was esophageal neoplasm. CT of chest revealed an occupying lesion in esophagus (Fig.3). Its density was uneven, with the centre being low-density and the periphery being a little intensified. There was no swelling lymph node in mediastinum. The diagnosis was esophagus occupying lesion.

Gastroscopy showed a long-shape submucosal tumor in the esophagus about 20~35 cm distant from incisor and 1.2 cm in diameter. Its basilar part was located in upper section of the esophagus about 20 cm distant from incisor; the surface was smooth and regular, and the color was normal (Fig.4 see Page 664). Gastroscopic diagnosis was esophageal submucosal lesion (adipose sarcoma?). EUS with 12 MHz micro-ultrasonic probe showed that the submucosal tumor in the esophagus was a hypoechoic lesion which was originated from submucosa. The internal echos of the hypoechoic lesion were uneven with many lumen-like structures. The submucosal tumor was measured 1.21 cm×0.84 cm in cross-section diameter and 15 cm in overall length (Fig.5). The surrounding esophagus of the tumor presented normal structure. There was no obvious bleeding when punctured by EUS-guided fine needle aspiration (FNA). Microscopically, the biopsied specimen was thick-wall vessels with fibrofatty in inequality of size, mixed with inflammatory granulation and necrotic tissue. Pathologic diagnosis was fibrofatty tissue mixed with thick-wall vessels, consistent with hamartoma.

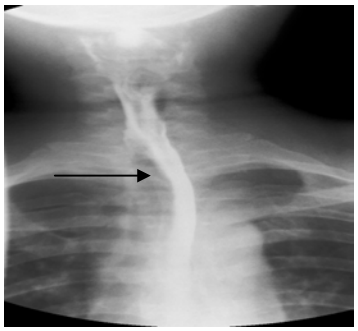


Fig.2 Barium esophagram: a little left-bias flow sign and a filling defect on the right of esophagus (arrow)

Fig.3 Contrast-enhanced computed tomography (CT): a occupying lesion in esophagus (arrow)

Fig.5 Endoscopic ultrasonography (EUS): a hypoechoic lesion, internal echos of which were uneven with many lumen-like structures, was originated from submucosa (arrow)

Having identified the etiopathogenisis of lesion and its origin of submucosa by EUS examination, we chose endoscopic resection for the treatment. In order to prevent lesion from bleeding during endoscopic resection, we ligatured basilar part of the tumor tightly with nylon string (OLYMPUS, Model No.

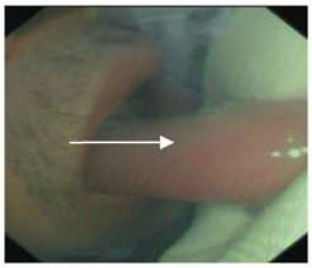
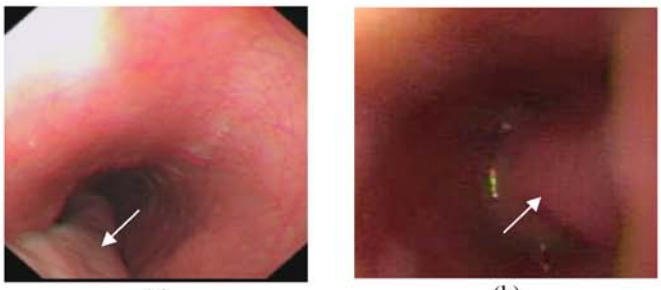


Fig.1 A long-shape neoplasm about 1.5 cm in diameter getting out of mouth (arrow)



(a) (b)

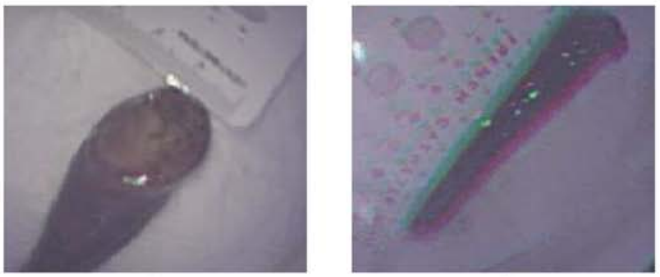
Fig.4 Endoscopy: (a) Submucosal lesion (arrow) in the esophagus. Its surface was smooth and regular, and the color was normal; (b) The basilar part of submucosal lesion (arrow) was located in esophagus about 20 cm distant from incisor

(a) (b) (c)

Fig.6 Ligate basilar part of the tumor tightly with nylon string under the gastroscope (a, b). And the mucous membrane of tumor presented ischemic change (color changed into gloomy purple) (c)

(a) (b) (c)

Fig.7 Tighten the tumor with a snare at 2 cm distant from the basal part ligated by nylon string (a, b), then resect the tumor (c)



(a) (b)

Fig.8 The resected specimen measuring about 11 cm×1.5 cm. (a) Cross-sectional view; (b) Lateral view

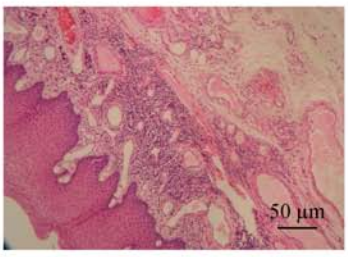


Fig.9 Pathological result: the specimen was covered with pavement-epithelium and contained a plenty of thick-wall vessels, fatty tissue, and many inflammatory cells (H & E stained)

MAJ-254) by the gastroscopy until the color of mucous membrane of tumor under the ligation changed into gloomy purple, and then left the nylon string (Fig.6, see Page 664). Then we resected the tumor at 2 cm distant from the distal end of the ligation position with a snare by 35 mV blend electric power of coagulation (Fig.7, see Page 664). Endoscopic resection was successful without complications such as hemorrhage and perforation. The size of resected specimen was about 11 cm×1.5 cm (Fig.8, see Page 664). Microscopically, the specimen was covered with pavement-epithelium and contained a plenty of thick-wall vessels, fatty tissue, and many inflammatory cells, mainly lymphocytes (Fig.9, see Page 664). Final pathologic diagnosis was esophageal hamartoma characterized in hemangioma.

The patient had been regularly followed up for 13 months after the endoscopic resection and had been in good condition without the feeling of foreign body and heartburn and recurrent lesion in the esophagus.

DISCUSSION

The etiopathogenisis of hamartomas is still unclear; however, it is considered to be tumor-liked hyperplasy formed by congenital developmental disorder of mesenchymal tissue of primitive mesoblast. They are rather a kind of developmental malformation tuberositas of tumor than real tumors (Wu, 1993; Hoffman *et al*., 1997; Hui, 2003). Esophageal hamartomas are rare, and there is only one case reported in China before this case (Utsunomiya *et al*., 1975; Dou and Liu, 1996; Song *et al*., 2004; Liu *et al*., 2006; Kevin and Charis, 2007). In the present case, the patient had found to have a tumor for more than 20 years. The tumor grew slowly and was visible outside the mouth when vomiting. As preoperative serological examination results were all in the normal range, it was unlikely to make the etiological diagnosis. The comparison of different imaging examinations demonstrated that both barium esophagram and CT scan could detect the lesion and provide relatively indirect image of lesion while CT can also provide information of various tissues and organs around the lesion. However, according to those imaging examination results plus medical history, only a diagnosis of benign lesion could be made, but not the differentiating diagnosis distinguishing the lesion from common esophagopolypus, nor the origin and etiopathogenisis of the lesion. Conventional gastroscopy is more direct than barium esophagram and CT scan, can show more precise understanding of location, size and surface structure of the lesion, and can identify a polyp originating from the epidermis or mucosa by biopsy. However, when gastroscopy showed that the lesion was a submucosal one, the conventional endoscopic biopsy was not able to confirm the diagnosis, for the specimen was too small and superficial (Sun and Liu, 2006). In this esophageal lesion case, conventional gastroscopy could only make the diagnosis of esophageal submucosal lesion, but was not able to distinguish the lesion from esophageal cyst, lipoma, hemangioma, leiomyoma and other submucosal lesions. While there is a parallel correlation between the histological structure of the digestive tract's walls and characteristics of ultrasonic echo (Rosch *et al*., 2002; Shen *et al*., 2002), addition to the ordinary functions of endoscopy, EUS can clearly show the layer structure of each gastrointestinal wall and identify the gastrointestinal lesion's surface and internal structure, echo, exact size, depth of invasion, origin, as well as the relationship between adjacent tissues and organs. So EUS can provide more diagnostic information about submucosal lesions, which will help to diagnose and differentiate submucosal lesions. At the present, EUS is the best means to diagnose submucosal tumors (Chen *et al*., 2001; Gress *et al*., 2001; Fusaroli and Caletti, 2005; Xu *et al*., 2008). Ultrasonic image of the present case showed a hypoechoic lesion, internal echos of which were uneven, with many lumen-like structures. The hypoechoic lesion was originated from the submucosa and had distinct borders as well as normal structures of surrounding esophageal wall of the lesion. According to those imaging features, it was quite easy to distinguish the lesion from cyst, lipoma, leiomyoma, GIST (gastrointestinal stromal tumor), hemangioma and other submucosal lesions. Given the focus in EUS with a certain characteristic and unique diagnostic value (Caletti and Ferrari, 1996), an accurate preoperative diagnosis may be made for gastrointestinal hamartoma in the future. If necessary, histopathological evidence by EUS-guided FNA could be also obtained to confirm the diagnosis.

Surgical operation and regular observation were usually taken to treat gastrointestinal submucosal tumors in the past. However, after identifying the lesion's origin, blood supply, and the relationship between adjacent tissues and organs, and achieving accurate positioning and nature diagnosis of submu-

cosal tumor, EUS can also provide reference and help to make reasonable and effective treatment plan (Takada *et al*., 1999; Song *et al*., 2004; Saito *et al*., 2005). Generally, it is thought to be feasible to perform a gastroscopic resection of tumors originated from the first to third layers of the gastrointestinal tract, while surgical operation and therapeutic laparoscopy are likely to cause perforation by an endoscopic resection when the tumors were originated from deeper layers (Lanzafame and Massimino, 1989; Kojima *et al*., 1999; Xiong *et al*., 2000). In this case, according to the information provided by EUS, we believed that the tumor was originated from the third layer (the submucosa), so the patient underwent the gastroscopic resection of hamartoma. As the tumor had a large basilar part, we ligatured the basilar part of the tumor tightly with nylon string before resection to prevent intraoperative and postoperative bleeding. Compared with conventional endoscopic resection, this therapy has advantages such as being safer, being more reliable, and having less complications; while compared with previous surgical operation, it is more convenient, less traumatic, having less complications, and more economic. We, therefore, conclude that EUS in diagnosis and treatment of esophageal and other gastrointestinal hamartomas is obviously superior to other types of imaging examinations.

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