Evaluation and Endoscopic Management of Esophageal SMT

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Introduction

Submucosal tumors (SMTs) are those that originate from cells constituting the submucosal layer and the muscularis propria and are covered by normal mucosa. Esophageal SMT is a rare disease accounting for less than 1% of overall esophageal tumors, but recent wide use of endoscopy has led to a rapid increase in incident cases of such tumors. Esophageal SMTs are positive in 90% or more of the cases, but the possibility of malignancies, such as gastrointestinal stromal tumor (GIST) and malignant leiomyosarcoma, still exists. Therefore, patients undergo a resection in the presence of symptoms or the possibility of a malignant tumor. For resection of esophageal subepithelial tumor, surgical resection was the only option available in the presence of the possibility of malignancy, but minimally invasive surgery by endoscopic resection is becoming more preferable to surgical resection with the development of endoscopic ultrasonography, endoscopy techniques and devices. This article will discuss methods of diagnosing esophageal SMT and a variety of therapeutic endoscopy.

Epidemiology of Esophageal SMTs

The frequency of esophageal SMTs is relatively low, with leiomyoma accounting for the majority of the cases (70-80%). GIST is a very rare disease accounting for less than 5% of the cases but accompanies the possibility of a malignancy, even malignant leiomyosarcoma in rare cases, which can cause dysphagia, obstruction, pain, and other symptoms, or requires resection if the diagnostic testing does not rule out a malignancy. Esophageal leiomyomas are usually detected in people in their 20s to 50s. They are more frequent in men in the ratio 2:1 and about 80% occur in the mid- and lower esophagus. The exact prevalence is unknown, but a report in Korea states that SMTs were detected in 1.45% - mostly involving the stomach (0.89%) followed by the esophagus (0.45%) - out of 48,926 cases of upper gastrointestinal endoscopy performed as a part of medical examination. Overall prevalence of esophageal SMTs was estimated around 0.4-0.6%, which is equivalent to previous reports.
Diagnosis of Esophageal SMTs

1. Endoscopic findings

The first thing to do upon detecting a SMT is to determine whether the tumor is intrinsic or extrinsic to the GI wall. Intrinsic compressions can be caused by either normal structures or positive or malignant lesion of the mediastinum. The normal structures include the aortic arch, left bronchus, and spine; patients with a congenital deformation particularly require a differentiation. Lymph node metastasis of lung cancer, recurrent esophageal cancer, and mediastinal tumor are the reported causes of extrinsic compressions by tumors. Extrinsic compressions are characterized by a large mass, often relocating depending on the patient’s position or breathing.

2. Endoscopic Ultrasound (EUS)

EUS allows observation of the size, layer of origin, echo pattern, internal properties, and margin of SMTs and enlarged lymph nodes near them. Layer of origin and echo pattern are particularly useful for differential diagnosis of SMTs. The echo patterns appear as hyperechoic, isoechoic, hypoechoic, or anechoic compared with the echo pattern of surrounding normal organs. It is most likely that a hypoechoic muscular layer is a leiomyoma. In the submucosal layer, hypoechoic pattern implies granular cell tumor and carcinoid, while hyperechoic pattern may suggest lipoma. Anechoic pattern suggests cystoma, lymphangioma, and vascular lesion. Hypoechoic muscularis propria suggests leiomyoma and stromal tumor. For the observation of esophageal SMTs, catheter probe EUS using water or gel is convenient and thus commonly used. Despite being the most useful technique for the diagnosis of SMTs, EUS still accompanies the risk of inter-observer variation depending on the observers’ proficiency. The inter-observer agreement was reported to be high for the diagnosis of cystic disease, lipoma, and intrinsic compressions; low for vascular lesion, leiomyoma, and stromal tumor; and very low for carcinoid and metastatic cancer. One limitation of EUS is that it is not the final diagnosis and requires histologic confirmation.

3. Histology

Common histology, which collects tissues only from the mucosa and part of the submucosa, is not recommended for every SMT. However, lesions of some SMTs (those that originate from the muscular layer such as leiomyoma and granular cell tumor) can be identified by histology, and sometimes lipoma or lymphangioma can be inferred based on the exposed tissue after histology. According to studies on SMTs with the likelihood of malignancy, malignancy was observed with 3.7% of GISTs smaller than 2 cm, and metastasis was observed with 2% of carcinoids smaller than 1 cm. For this reason, recent guidelines recommend histology for SMTs not smaller than 2 cm (US and Europe) or 1 cm (Japan). However, current histologic techniques using EUS such as fine needle aspiration biopsy and Tru-cut biopsy have a diagnostic accuracy of not more than 52 - 86%. The fine needle aspiration biopsy does not provide enough amounts of tissues for differentiation of SMTs, and the Tru-cut biopsy is difficult to handle for less skilled operators.
Treatment of Esophageal SMTs

Recent widespread use of endoscopic submucosal surgery made it possible to perform endoscopic diagnosis and treatment of esophageal SMTs. Endoscopic submucosal dissection (ESD) allows histology of SMTs in the muscularis propria. This is called endoscopic unroofing technique because it involves incision of the mucosa covering a suspected GIST lesion to confirm the tumor tissue and to obtain biopsy samples with a forceps. Endoscopic resection alone is often enough to achieve complete resection of SMTs in the muscular layer or submucosal layer. However, the histological resection rate is still approximately 78% with this technique, which often results in intraoperative tumor rupture and the risk of complications such as perforation and hemorrhage in less than 5%. There are new techniques as well such as endoscopic submucosal tunnel dissection (ESTD) and endoscopic muscularis dissection (EMD). ESTD consists of 5 stages, which was first introduced for the treatment of esophageal achalasia. First, inject a submucosal solution about 5 cm proximal to the lesion and then create a 2 cm mucosal incision so that the endoscope could be inserted into the submucosal layer. Afterwards, dissect the submucosal layer with a knife to create a submucosal tunnel until reaching about 1 - 2 cm distal to the SET, and then perform the resection of the SMT. Control the hemorrhage and then close the proximal mucosal resection with clips. EMD differs from ESD in that it requires precutting of the mucosa. The lesion is then dissected by blunt dissection (by pushing between the tumor and muscular layer), without using electrical dissection, to minimize injury to the muscular layer. Afterwards, close the mucosa as much as possible with endoclips.

Conclusion

With growing interest in SMTs and development of endoscopy devices and technology, the diagnosis and treatment of SMTs have made a lot of progress recently. With large-scale studies on endoscopic submucosal surgery techniques and preventive measures of complications such as perforation, and with the help of safer and more precise endoscopy devices, more advanced diagnostic and therapeutic techniques would be available for esophageal SMTs.

References