Int Surg 2013;**98:**461–465
DOI: 10.9738/INTSURG-D-12-00021.1

Case Report

Video-Assisted Thoracoscopic Surgery for Localized Neurofibroma of the Esophagus: Case Report and Review of the Literature

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Esophageal submucosal tumors are less common than other gastrointestinal tract tumors. Leiomyoma is the most common benign esophageal SMT, accounting for more than 70% of these tumors. We report on a case of a 56-year-old woman with a 3-cm diameter midthoracic esophageal submucosal tumor. Magnetic resonance imaging suggested leiomyoma or neurofibroma. Video-assisted thoracoscopic surgery was performed to enucleate the tumor from the esophageal wall by splitting the muscle layers. The postoperative course was uneventful, and the patient was discharged on postoperative day 8. Immunohistochemical staining confirmed the diagnosis of esophageal neurofibroma. Gastrointestinal tract involvement of neurofibromatous lesions is rare and occurs most frequently as a systemic manifestation of von Recklinghausen disease. Cases of localized esophageal neurofibroma with prior or subsequent evidence of generalized neurofibromatosis have rarely been documented. This is a rare case of isolated esophageal neurofibroma without classic systemic manifestations of generalized neurofibromatosis, and it is the first reported case treated by video-assisted thoracoscopic surgery.

Key words: Neurofibroma – Esophagus – Thoracoscopic surgery

Benign esophageal tumors are relatively rare, and leiomyomas account for most of these tumors. Rarely, gastrointestinal stromal tumors, papillomas, hemangioma, granular cell tumors, neurofibromas, and myxofibromas occur. Neurofibromas are generally associated with hereditary diseases and are usually a manifestation of von Recklinghausen disease (VRD).[1] However, neurofi-

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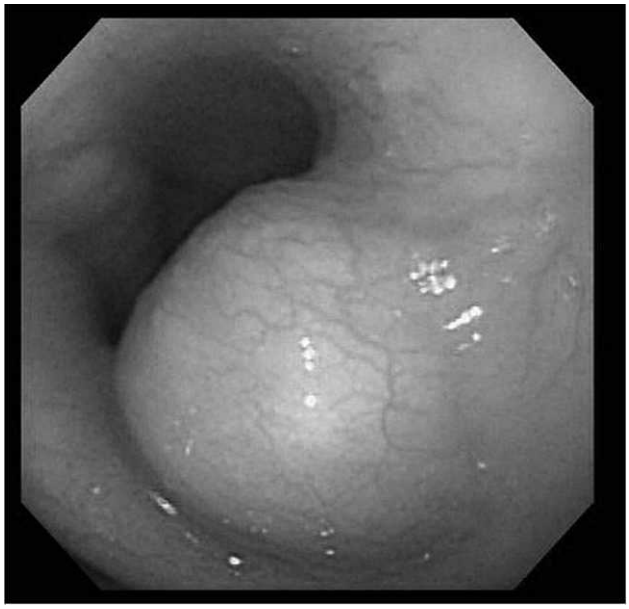

Fig. 1 Endoscopic examination revealed an esophageal tumor at the midthoracic esophagus, which was covered with normal esophageal epithelium.

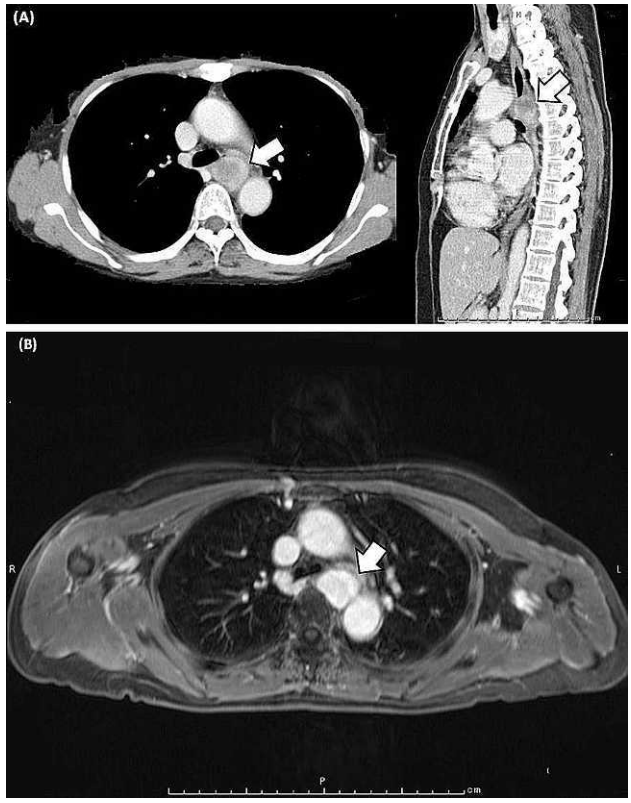

Fig. 2 Chest computed tomography shows a well-circumscribed tumor (arrows) measuring 25 × 33 × 24 mm located in the posterior mediastinum at the level of the tracheal bifurcation (A). A heterogenous inner structure was revealed by magnetic resonance imaging (B).

bromas rarely occur in the gastrointestinal tract (GI) as isolated tumors outside the classic clinical picture of the VRD.[2–8]

We report on a rare case of isolated esophageal neurofibroma in a patient without systemic manifestations of generalized neurofibromatosis, which is the first reported case treated by video-assisted thoracoscopic surgery (VATS).

Case Report

A 56-year-old woman underwent GI endoscopy for epigastric discomfort at an outpatient clinic in her neighborhood. Endoscopic examination revealed a 2-cm–long submucosal tumor (SMT) in the midthoracic esophagus. Since then, the tumor size was examined annually by GI endoscopy. GI endoscopy in the fourth year showed tumor enlargement, and the patient was eventually referred to our institution. Endoscopic examination was repeated for precise tumor review. The esophageal SMT measuring 3 cm in diameter was located in the midthoracic esophageal region 25 cm from the incisor teeth (Fig. 1). The tumor appeared to have grown approximately 1 cm relative to the size observed the previous year. Endoscopic ultrasonography demonstrated a hypoechoic and homogeneous mass in the submucosal layer. Following endoscopic ultrasonography, a fine-needle aspiration biopsy of the tumor was performed. However, the biopsy specimen size was too small for diagnosis. Computed tomographic and magnetic resonance imaging revealed a well-circumscribed tumor measuring 25 × 33 × 24 mm located in the posterior mediastinum at the level of tracheal bifurcation, and with a heterogeneous inner structure (Fig. 2).

Radiologic and endoscopic findings suggested esophageal leiomyoma or neurogenic tumor. Although the patient did not show any clinical symptoms, surgical treatment was chosen because of the tumor enlargement.

The patient underwent enucleation of the tumor by VATS. A small 2.5-cm diameter thoracotomy was first created at the right fifth intercostal space on the anterior axillary line, with the patient in the left lateral decubitus position. Four additional thoracic ports (11.5 mm) were created on the posterior axillary line at the fifth intercostal spaces, medial

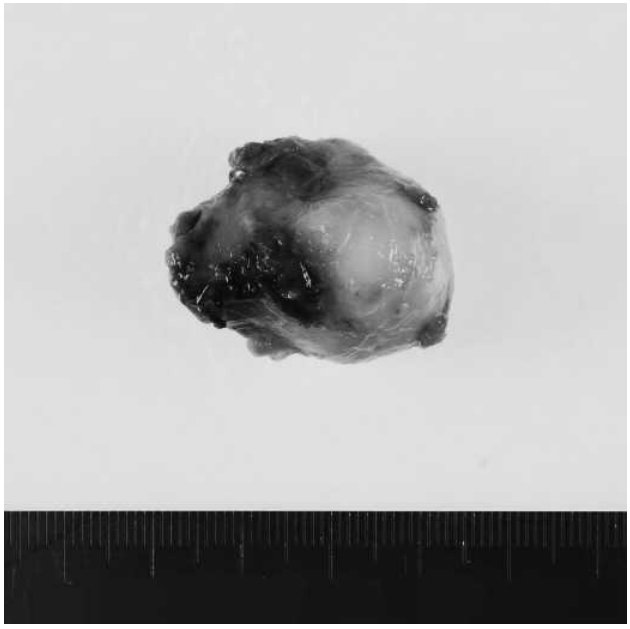

Fig. 3 Macroscopic appearance of the tumor. The resected tumor was soft in elasticity and measured 34 × 28 × 22 mm.

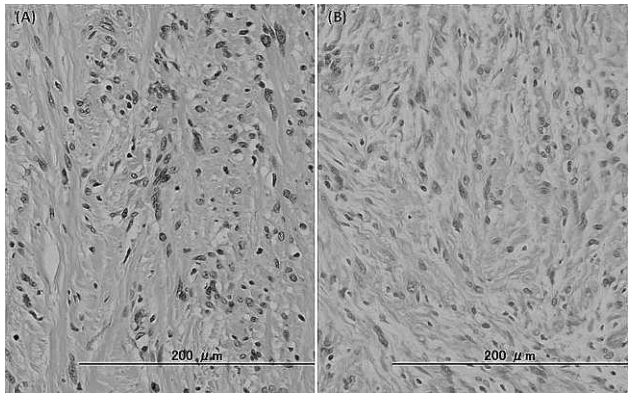

Fig. 4 Histologic appearance of the tumor. A tumor comprised a mixture of fibrillary collagen sheets and cords of spindle cells with nodular growth (A). Immunohistochemical staining results were positive for S-100 (B).

axillary line at the fourth and seventh intercostal spaces, and proximal axillary line at the third intercostal line for the endoscope, lung retraction, and surgical assistance, respectively. After the esophagus was freed from the posterior mediastinal space, the tumor was found to bulge dorsally from the upper thoracic esophagus. A Sengstaken-Blakemore tube was inserted orally, and esophageal balloon inflation was used to push the tumor outward from the esophagus. The tumor was easily enucleated from the esophageal mucosa and muscular layer. The esophageal muscular layer was closed roughly with 3 single stitches using extracorporeal knotting technique. The resected tumor was soft in elasticity, it measured 34 × 28 × 22 mm, and the cut surface was whitish (Fig. 3). Histologic examination revealed that the tumor was ill defined with indistinct borders and comprised mixed fibrillary collagen sheets and cords of spindle cells with nodular growth. No signs of atypia or significant mitotic activity were observed. Immunohistochemical staining results were positive for S-100 but negative for c-KIT, CD34, αSMA, and desmin (Fig. 4). The above morphologic and immunohistochemical characteristics were consistent with the diagnosis of neurofibroma.

Postoperative nasogastric tube or esophageal contrast study was considered unnecessary because the tumor was enucleated without mucosal damage and additional muscular coverage was obtained. Therefore, the patient was allowed to consume water on the next day after surgery and a solid meal on the second day after surgery. The postoperative course was uneventful, and the patient was discharged on the eighth postoperative day. She remains tumor free without any postoperative complications 2 years after surgery.

Discussion

Esophageal SMTs are less common than other GI tract tumors and are classified as benign or malignant disease. Gastrointestinal stromal tumors are the most common malignant SMTs. Leiomyoma is the most common benign esophageal SMT, accounting for more than 70% of these tumors.[9] Neurofibromas associated with esophageal SMTs have been reported by Plachta[10] to be 0.9%.

Neurofibromas are generally associated with hereditary diseases and are usually manifestations of VRD.[1] They are associated with benign tumors comprising neural and connective tissue components, such as Schwann and perineural cells and myofibroblasts. Gastrointestinal involvement is reported in 25% patients with VRD.[11] There are 3 types of neurofibromas: localized, diffuse, and plexiform. Localized neurofibromas of GI tracts, unlike VRD, are benign nerve sheath tumor in the peripheral nervous system originating from the Auerbach plexus and Meissner submucosal plexus.[12,13] Our patient had a localized neurofibroma,

and a type that is far more common in GI tracts. Cases of localized neurofibromas of the esophagus with prior or subsequent evidence of generalized neurofibromatosis, including our case, have rarely been documented.[2–8] According to our Medline search, 1 case of plexiform neurofibroma of the esophagus was reported in association with neurofibroma and VRD.[14] Most neurofibromas are benign tumors; however, nodular and particularly plexiform types may demonstrate a malignant transformation in 2% to 16% of affected individuals.[15]

We found 16 reported cases of localized esophageal neurofibromas, and Fujita et al[2] and other researchers[3–8] summarized 11 cases in the literature. The mean age of the affected patients was 51.8 years (range, 26–75 years), and there was no gender preference (male to female, 7:9). Symptoms of dysphagia were observed in 32% of patients. Tumors were located mainly in the upper to middle sections of the thoracic esophagus (83%), and the mean tumor size was 5.7 cm (range, 0.5–22.5 cm), whereas leiomyomas were located in the middle to distal regions of the esophagus.[16]

Histopathologic diagnosis of neurofibroma is necessary in most cases because neurofibroma or other submucosal tumors, such as leiomyoma, are often difficult to diagnose using diagnostic imaging. The histopathologic characteristic appearance of neurofibroma mainly comprises spindle-shaped cells associated with collagen fibrils. In addition, immunohistochemical staining helps distinguish neurogenic from myogenic tumors. In our case, the tumor comprised a mixture of fibrillary collagen sheets and cords of spindle cells with modularly growth; it was positive for S-100 but negative for c-KIT, CD34, αSMA, and desmin.

The treatment for localized neurofibromas in GI tracts is primarily surgery and depends on the location and size of the lesions. Solitary well-circumscribed lesions may be only incidental findings at the time of screening endoscopy and require no further therapy, but larger solitary lesions may be noticed clinically because of intestinal obstruction or impingement on adjacent structures, and they may require resection. In fact, 14 of 16 reported cases were surgically treated. In the case of esophageal neurofibromatosis, especially associated with VRD, surgical resection should be performed promptly even if there are no clinical symptoms because malignant degeneration may occur prior to surgery. In our case, we selected surgical treatment because the tumor had enlarged over a relatively short period and may have had the potential for malignant transformation.

To date, all reported surgical treatments have been performed through thoracotomy or laparotomy. Our case appears to be the first where VATS was used for successfully treating a patient with esophageal neurofibroma. Surgical procedures can be performed in a manner similar to those used for leiomyomas and other benign tumors.[17]

We conclude that enucleation of esophageal neurofibromas is feasible and is an ideal indication for a thoracoscopic approach. However, the procedure should be performed with extra care, especially if malignancy is suspected.

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