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# CASE STUDY ESOPHAGEAL SCHWANNOMA PRESENTING WITH CHEST OPPRESSION; REPORT OF A CASE

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Abstract We report a rare case of esophageal schwannoma. The patient was a 41-year-old man who was admitted to our hospital due to chest oppression. Chest imaging showed a solid mass in the middle mediastinum compressing the esophagus and the trachea. Neurogenic tumor was suspected and the patient underwent extirpation of the tumor through right thoracotomy. The tumor,  $4.5 \times 4.3 \times 3.4$  cm in size, was strongly attached to the esophageal wall. Pathological examination revealed myxomatous pattern in the tumor. Immunohistochemical staining for S-100 protein was positive and the diagnosis of schwannoma was made. The majority of mediastinal schwannomas arise from sympathetic nerve cells in the posterior mediastinum. Esophageal schwannoma is rare and only 27 cases were reported in literature to date.

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Key words: middle mediastinal tumor; chest oppression; esophageal Schwannoma.

#### 症例研究

## 胸部圧迫症状を呈した食道神経鞘腫瘍の1例

藤	田	和歌子	福	田	幾	夫	北	Л	理映	子
木	村	大 輔	山	田	芳	嗣	對	馬	敬	夫

**抄録** 著者らは、まれな食道神経鞘腫の1例を経験したので報告する. 症例は41歳の男性. 胸部圧迫症状があり、当科に紹介入院となった. 胸部画像検査では中縦隔に食道および気管を圧迫する腫瘤を認めた. 神経原性腫瘍が疑われ、右 開胸による腫瘍摘出術を行った. 腫瘍は食道壁に連続しており、摘出標本は4.5×4.3×3.4cm 大であった. 術中所見およ び病理組織所見から、食道より発生した schwannoma と診断した. 食道神経鞘腫は稀であり、文献検索では27 例しか 報告されていない.

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キーワード:中縦隔腫瘍;胸部圧迫感;食道神経鞘腫.

### Introduction

Neurogenic tumors are uncommon, though they comprise 15-25% of all primary mediastinal tumors<sup>1)</sup>. Thoracic neurogenic tumors are often

Department of Thoracic and Cardiovascular Surgery, Hirosaki University School of Medicine, Hirosaki, Aomori, Japan Correspondence: I. Fukuda Received for publication, April 3, 2008 Accepted for publication, May 27, 2008 found in the posterior mediastinum or in the chest wall<sup>2)</sup>, and schwannoma is the most common neurogenic tumor seen in adults. However, schwannoma arising from esophagus is rare. We herein report a case of esophageal schwannoma in

弘前大学大学院医学研究科胸部心臓血管外科学講座 別刷請求先:福田幾夫 平成20年4月3日受付 平成20年5月27日受理 the middle mediastinum that posed a diagnostic difficulty.

### **Case Report**

A 41-year-old man was referred to Hirosaki University Hospital for evaluation of a mediastinal mass and a surgical treatment. He had a several episodes of chest oppression in the last 5 years. Coronary-artery angiography was performed at a local hospital in 2002 when he had his first chest symptom. It demonstrated no stenosis or obstruction of the coronary artery. His most recent episode of chest pain occurred one month before admission of our hospital. Sublingual administration of nitroglycerin in the emergency room did not improve his chest pain, but eructation eliminated the symptom. Chest computerized tomography (CT) revealed well-circumscribed mass 4.3 x 4 x 4 cm at the posterior of trachea (at the level of T3 - T5) compressing the esophagus to the right (Fig 1). Upon admission, he complained of chest discomfort. He had no symptom of dyspnea or dysphagia.

On physical examination, the patient's height was 177 cm and his weight 105 kg. The body mass index was 33.5kg/m<sup>2</sup>. The blood pressure was 130/93 mmHg and the heart rate was 79 beats per minute. Lymph nodes in the cervical, infraclavicular and axillary arteries were not palpable. Physical examination of the lungs and heart showed no abnormalities. The abdomen was obese and nontender, with no organomegaly. There were no focal neurologic deficits. Laboratory data on admission, including electrolyte levels, white blood cell count, hematocrit, and urinalysis, were within normal limits. His chest X-ray revealed an enlarged mediastinal shadow (Fig. 2). Gastrointestinal fiberscopy was normal, and it did not demonstrate esophageal wall invasion. The barium esophagogram showed a round tumor compressing the upper thoracic esophagus (Fig. 3). Bronchoscopy revealed a



Figure 1 Chest computerized tomography (CT) revealed well-circumscribed mass at the posterior of trachea compressing the esophagus.



Figure 2 Chest X-ray showing enlarged mediastinal shadow.

tracheal deviation and protruding membranous wall of trachea. Magnetic resonance imaging (MRI) showed a tumor with isointensity on T1 weighted image and high intensity on T2 weighted image (Fig. 4). Considering these results, neurogenic tumor was suspected.

The patient was placed in the left lateral decubitus position after induction of general anesthesia, and one-lung ventilation was started. Posterolateral thoracotomy was performed over the 4<sup>th</sup> intercostal space. The mediastinal pleura around the tumor was incised and right



Figure 3 Barium esophagogram shows a round tumor compressing the upper thoracic esophagus.



Figure 4 Magnetic resonance imaging (MRI) showed a tumor with isointensity on T1 weighted image and high intensity on T2 weighted image.

phrenic nerve was retracted with tape. The tumor located near the arch of azygos vein was attached to the esophagus and there was no clear separation between the tumor and the esophagus wall. There was no adhesion between the tumor and the membranous wall of trachea. The tumor was completely extirpated without



Figure 5 The resected tumor was an encapsulated elastic mass, measuring  $4.5 \times 4.3 \times 3.4$  cm in size and 32g in weight.

injuring the muscular layer of the esophagus. The resected tumor was an encapsulated elastic mass, measuring  $4.5 \times 4.3 \times 3.4$  cm in size and 32g in weight (Fig. 5). The cut surface of the tumor was solid and yellowish white. The patient had an uneventful postoperative course and he was discharged on postoperative day 12. Histologically, the tumor showed areas of edema and myxomatous changes with less cells, which is characteristic of benign schwannoma (Antoni B). The immunohistochemical staining for S-100 protein was positive.

### Discussion

Schwannoma is a benign tumor that is derived from Schwann cells. It is usually asymptomatic but becomes symptomatic when it invades or compresses adjacent structures, as in this case. Takeda et al.<sup>1)</sup> reported that 83.7% of the adult patients with intrathoracic neurogenic tumors were asymptomatic and their tumors were found on routine chest X-rays by chance. Most mediastinal schwannomas arise from sympathetic nerve cells in the posterior mediastinum. Esophageal schwannoma is rare and only 27 cases were reported in the literatures to date. In the cases of esophageal schwannoma, the most common symptom was dysphasia<sup>3)</sup> and chest symptom was seen in only a few patients<sup>4)</sup>. In our patient, schwannoma originating from the esophageal wall kept causing him chest oppression and acute myocardial infarction was once suspected in a local hospital. According to Saito et al.<sup>5)</sup> benign schwannomas are usually located in the upper esophagus and occur frequently in middle-aged women.

Perioperative evaluation of mediastinal tumor is done by CT scan and MRI, as they enhance the accuracy of diagnosis. In the case of schwannoma, the tumor margin or the entire tumor is usually enhanced in CT scan using contrast medium. MRI image shows low to high signal intensity on T1 and high intensity on T2. However, there is no specific sign for schwannoma and no reports have been published on making accurate preoperative diagnosis. Differential diagnosis usually includes esophageal submucosal tumor, leiomyoma and mediastinal tumor<sup>6, 7)</sup>. In our patient, tracheal cyst was suspected upon admission. After MRI evaluation of the tumor, differential diagnosis of neurogenic tumor, angioma, esophageal leiomyoma were made.

Mediastinal neurogenic tumors are usually asymptomatic and benign in nature. Since it leads to definite diagnosis and provides therapy, surgical resection is preferred. Benign intrathoracic neurogenic tumors are good candidates for Videoassisted thoracoscopic surgery (VATS) resection, which can be achieved safely and effectively with rapid recovery and less pain resulting in shortening hospital stay<sup>8)</sup>. VATS often provides an appropriate view for the operation, especially for the posterior mediastinum<sup>2</sup>. VATS resection of the tumor was originally planned in our patient but his thick subcutaneous tissue made it impossible to insert a trocar. Therefore, standard posterolateral thoracotomy was performed instead. In general, the prognosis of esophageal schwannoma after surgical removal is excellent and no cases with postoperative complications have been reported. The frequency of malignancy among intrathoracic neurogenic tumors is relatively low<sup>2)</sup> but a few cases of malignant esophageal schwannoma have been reported.

### Conclusion

We conclude that making preoperative diagnosis of schwannoma with chest imaging is challenging. Complete surgical resection of the tumor is the treatment of choice and pathological analysis should always be made. Long-term follow up is recommended after the removal of esophageal schwannoma.

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