A Case of Benign Esophageal Schwannoma Causing Life-threatening Tracheal Obstruction

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A 59-year-old woman presented with a 1-year history of dysphagia. She suffered from a large mediastinal mass obstructing trachea and bilateral main bronchus, which led to dyspnea and disturbed consciousness. Immediate intubation and surgery was required. A solid tumor that included esophagus and right vagal nerve, and adhered to the membranous part of the bronchus was found. However, the tumor could be resected en bloc and the patient has been free from recurrence. Pathologically, the tumor exhibited proliferative spindle cells and was diffusely positive for S-100 protein. It was therefore diagnosed as a benign esophageal schwannoma. To our knowledge, this is the first report of tracheal obstruction from a benign esophageal schwannoma, which we successfully treated with emergency subtotal esophagectomy.

Keywords: esophagus, schwannoma, dyspnea, tracheal obstruction

Introduction

Schwannoma of the esophagus is rare and most frequently located in the upper thoracic esophagus.1 Dyspnea frequently occurs and reports about it are recently increasing.2–6 However, esophageal obstruction has not been previously reported, nor has acute surgical intervention been needed. We describe a case of a major schwannoma of the esophagus, which required emergency esophagectomy, and review 5 publications about tracheal compression.

Case Report

A 59-year-old woman presented with a history of diabetes mellitus. She had been aware of dysphagia in the past 1 year. However, since this symptom seemed mild, she rejected further evaluation. She was transported to emergency room of another hospital after an emergency medical services (EMS) call, resulting from dyspnea and disturbed consciousness. Her condition required emergency intubation, which did not significantly improve her state. Laboratory data from arterial blood showed respiratory acidosis with hypoxemia and hypercapnia (pH 7.165, PaO2 79.8 mmHg and PaCO2 99.5 mmHg at bilateral positive airway pressure [BIPAP], FiO2 0.8, positive end-expiratory pressure [PEEP] 8 cmH2O, f 15). Hyperglycemia was also found. A computed tomography (CT) scan revealed a posterior mediastinal mass, about 10 cm in diameter. The tumor showed a heterogeneous pattern and strongly compressed the esophagus and trachea (Fig. 1a, b). The tracheal lumen narrowed to 9 × 4 mm at the minimum point. The CT scan also showed the pneumonia in the left lower lung lobe. The patient was then taken and
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admitted to our hospital. Esophagogastroduodenoscopy revealed esophageal obstruction, but the mucosa was intact (Fig. 1c). We regarded this as a submucosal tumor of the esophagus causing airway obstruction. Emergency surgery was performed.

The patient underwent a right-sided thoracotomy with bilateral lung ventilation. The tumor was solid and existed among the vertebrae, trachea and right main bronchus. The tumor strongly adhered to the membranous part of the bronchus, but was able to be released. The tumor, esophagus and right vagal nerve were compacted into a solid mass, so a subtotal esophagectomy was performed. Cervical esophagostomy and tube gastrostomy for decompression and further feeding were also done. Reconstructive surgery was performed 8 months later. The tumor was 10.9 cm × 7.2 cm × 7.1 cm in size, with two components visible macroscopically, as had been revealed by the CT scan. The cut surface of the tumor showed an ivory area and a brown area (Fig. 2a). Pathological examination revealed that the tumor continued to the muscularis propria of the esophagus and consisted of atypical spindle-shape cells (Fig. 2b, c). A lymphoid cuff was also detected, and there was no mitosis. Immunohistologically, the cells were positive for S-100 protein (Fig. 2d), but negative for CD34, c-kit, α-smooth muscle actin (SMA) and vimentin. The tumor was diagnosed as a benign esophageal schwannoma. The surgical margin was pathologically negative. The patient has shown no signs of recurrence for 8 months.

Discussion

Esophageal tumors predominantly originate in the epithelium, and those of submucosal origin are rare.7) Most submucosal tumors of the esophagus are leiomyoma,4) and schwannomas are less common. Esophageal schwannomas are commonly benign, with only a few reports of malignant cases.8,9) After complete resection, patients make good progress. Esophageal schwannomas typically present in middle aged women and the most frequent symptom is dysphagia.4) While an esophageal schwannoma occasionally causes obstructive dyspnea, the immediate surgical treatment is rarely required. If the tracheal lumen narrows to 75% of that under the vocal cord, the patient generally presents with obstructive respiratory failure.10) In the current case, the minimum tracheal cross-section area was 46 mm², an 80% decrease, compared with the area under the vocal cord. The causes of acute upper airway obstruction include many clinical conditions such as infections, trauma, tracheal intubation and tumors.11) Similar to the endotracheal neoplasms, tumors located outside of the trachea can straighten the inner cavity. CT scan allows differentiation between causes of airway obstruction.

Ever since Chaterlin, et al. described the esophageal schwannoma in 1967,12) only 29 cases have been reported.
According to a PubMed search using the word “esophageal schwannoma”. Six cases, including ours, presented with dyspnea, and all of them had tracheal compression (Table 1). The sizes of the tumors in these reports were over 70 mm. Treatments included four enucleations, one partial resection and one subtotal esophagectomy. The common factor leading to esophagectomies was possibility for malignancy,4–15 Preoperative diagnosis of schwannoma was regarded to be difficult,4 so many cases needed surgical resection to diagnose. In some reported cases, although a benign schwannoma was diagnosed before surgery, subtotal esophagectomy was performed, because the tumor size was too large to repair without stenosis after partial resection.7,16 We judged that subtotal esophagectomy was inevitable for this large tumor broadly adhered to the esophagus and not preoperatively diagnosed.

This patient presented here had been in a critical state with dyspnea and complicating pneumonia. A deeper tracheal intubation was thought to be ineffective, as the tumor was presented in the thoracic esophagus, including a more peripheral position beyond the tip of the inserted tube. While placing a tracheal stent might be an optional treatment, it seemed difficult to keep enough lumen for its length. Therefore, for this case, an emergency esophagectomy was necessary to release the occlusion promptly and to make a definite diagnosis pathologically. Thoracoscopic esophagectomy might be a treatment option, but is usually performed under one-lung ventilation. We thought that her respiratory condition would not allow this option. In case that her respiratory failure was too severe to continue the operation, we had prepared a percutaneous cardiopulmonary support device (PCPS).

The preoperative diagnosis of this case was a submucosal esophageal tumor and we tried to perform an en-bloc resection. Under the assumption that the tumor is malignant, an en-bloc resection should be done. We achieved it because the tumor did not invade the bronchus or other surrounding organs.

**Conclusion**

We successfully performed a life-saving emergency resection for a patient with life-threatening tracheal stenosis due to a giant esophageal schwannoma. As the symptom and surgical procedure depend on size and location of the tumor, proper and timely treatment is essential.

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**Disclosure Statement**

All of authors have no conflict of interest.

**References**