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Case report

A rare case of bilateral vagus nerve schwanomatosis

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ABSTRACT

Schwanomatosis is the third most common form of neurofibromatosis. Schwanomatosis affecting the vagus nerve is particularly rare. In this report, we describe an extremely rare case bilateral vagus nerve schwanomatosis in a 45-year-old male patient. The patient initially presented with bilateral neck tumors and hoarseness arising after thoracic surgery. We performed left neck surgery in order to diagnose and resect the remaining tumors followed by laryngeal framework surgery to improve vocal cord closure and symptoms of hoarseness. Voice recovery was successfully achieved after surgery. An appropriate diagnosis and surgical tumor resection followed by phonosurgery improved patient quality of life in this rare case.

1. Introduction

Schwannoma is a neural-origin benign tumor that is derived from Schwann cells that constitute the peripheral nerve sheath. While schwannomas usually arise from the auditory nerve, they have also been reported to develop from other cranial nerves including the vagus nerve. Multiple schwannomas are most commonly identified in cases of neurofibromatosis 2 (NF2), presenting with acoustic neurinomas and other peripheral nerve schwannomas but rarely affecting the other cranial nerves. Recently, the term schwanomatosis was introduced to describe multiple schwannomas arising from the cranial nerves, spiral nerve roots, and/or peripheral nerves without accompanying auditory neuromas or NF2 symptoms [1,2].

Vagus nerve schwanomatosis is a rare disease, with only a handful of cases reported in the literature [3]. Bilateral vagus nerve schwanomatosis is extremely rare [4]. Accordingly, the preoperative diagnosis of vagus nerve schwanomatosis is difficult and requires histopathological confirmation in most cases. Recurrent nerve palsy causing hoarseness can be an unavoidable complication after surgical removal of a benign schwannoma. Thyroplasty type-1 is one option for the treatment of postoperative recurrent nerve palsy. Here, we report a very rare case of bilateral vagus nerve schwanomatosis and postoperative vocal impairment that was successfully treated by thyroplasty.

2. Case presentation

A 45-year-old male patient presented at our department with bilateral neck tumors and hoarseness arising after thoracic surgery. Five years prior, he had abnormal findings on chest X-ray, but did not complete a follow-up medical examination. He eventually noticed a cervical mass and developed chronic cough lasting for one year before he visited a physician's office. Upon close inspection, the patient was diagnosed with a mediastinal tumor and multiple neck masses. Two months before presentation at our clinic, he underwent mediastinal

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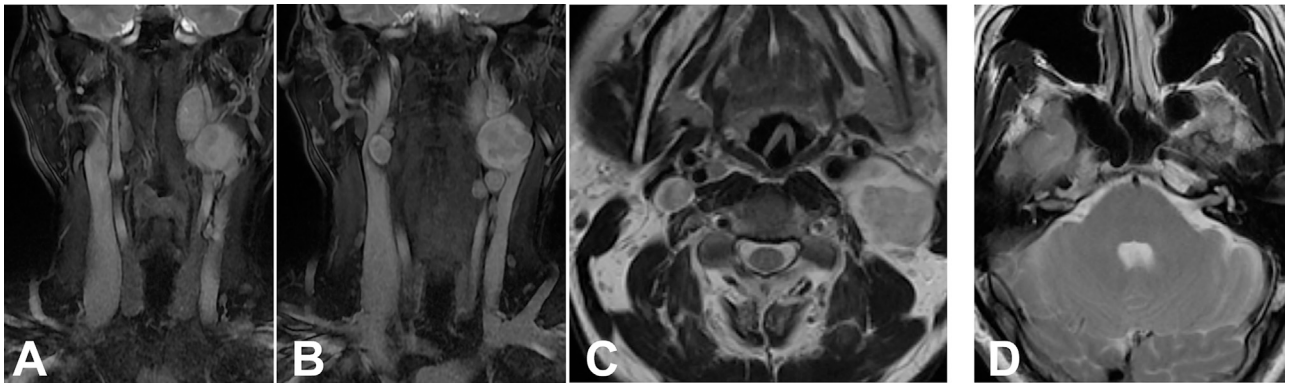

Fig. 1. Preoperative magnetic resonance imaging. Images showing bilateral neck tumors (A–C). No obvious vestibular neuronal tumors were observed (D).

tumor resection; the two tumors that were resected were classified as mediastinal vagus nerve schwannomas based on intraoperative findings and histopathological examination.

The patient visited our department complaining of postoperative hoarseness. Upon examination, there were multiple palpable masses on the left side of his neck. The patient had difficulty speaking and his left vocal cord was fixed in a paramedian position that prevented sufficient vocal cord closure. To further characterize the neck masses, we performed magnetic resonance imaging (MRI). Imaging revealed that the masses were well-defined, non-uniformly enhanced, and distributed along the bilateral cervical vagus nerves. The largest lesion was approximately 30 mm × 33 mm × 36 mm and located at the para-pharyngeal space and carotid sheath. Other lesions were 20 mm, 16 mm, 13 mm, and 10 mm in diameter on the left side and 14 mm and 8 mm in diameter on the right side (Fig. 1A–C). No intracranial tumors were detected (Fig. 1D). There was no obvious cervical lymph node enlargement and the thyroid gland appeared normal. We considered thyroplasty to improve the patient's hoarseness, but some of the left neck masses were large and surgical resection was deemed necessary to yield a definitive histopathological diagnosis given the rarity of bilateral vagus nerve schwanomatosis. Fine needle aspiration was not performed in this time, because at least two of these multiple tumors have been diagnosed already as schwannomas by a preceding mediastinal tumor resection and no obvious imaging findings suggest malignancy.

Five months after the patient's first visit to our department (seven months after thoracic surgery), we performed a trans-cervical left neck tumor resection. While there were also small masses on the right side, we only performed surgery on the left side to avoid bilateral recurrent nerve palsy as a complication. Intraoperatively, two well-encapsulated masses (about 50 mm and 20 mm in size, respectively) were identified superolateral to the carotid bifurcations beneath to the digastric muscle. We also found and resected two dominant masses arising from the vagus nerve and confirmed that the hypoglossal and accessory nerves were intact. Finally, we resected a swollen lymph node near the other resected masses. A small palpable mass was identified in a more cranial position extending beyond the digastric muscle, but we thought that resecting this mass would be too invasive and opted not to remove it.

The final histopathology report showed two schwannomas with palisading patterning and the presence of S100-positive cells (Fig. 2). On this premise, we concluded that the resected masses were vagus nerve schwannomas. Taken together with findings from the preceding thoracic surgery, we diagnosed the patient with bilateral vagus nerve schwanomatosis.

Eight months after the patient's first visit to our center (three months after cervical surgery), we performed a Type I thyroplasty under local anesthesia to address insufficient vocal cord closure and symptoms of hoarseness. We made a transverse neck skin incision in a natural skin crease. After raising the subplatysmal flaps, the strap muscles were dissected from the thyroid cartilage. The perichondrium was elevated and a small window was opened by excising a small piece of cartilage. The cartilage was pushed through the window and a Gore-Tex® sheet (1 cm × 1 mm × 6.5 cm) was inserted. We

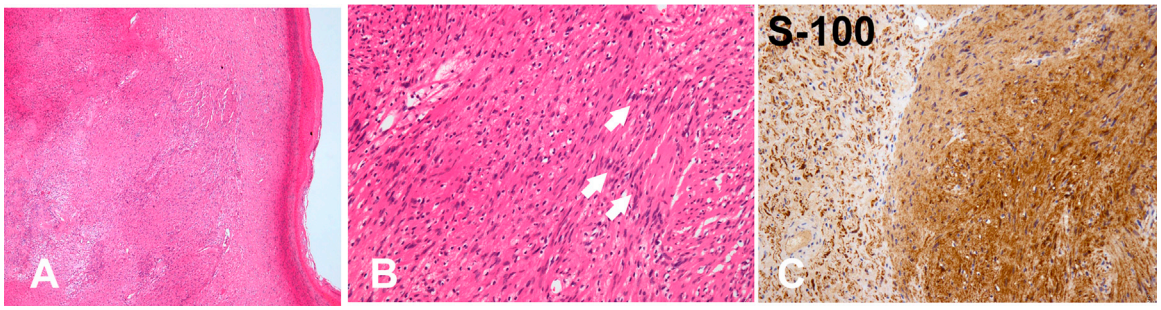

Fig. 2. Histological staining of resected tumors. Hematoxylin and eosin staining is shown in panels A and B; immunohistochemical staining of S-100 protein is shown in panel C. Palisading patterning, a characteristic future of schwannoma, was observed (Arrow in B). S-100 positive cells were also identified in the resected tumor (C).

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