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CASE REPORT

Vagus nerve schwannoma in the right upper mediastinum

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Abstract

A 35-year-old woman was admitted to our hospital with an abnormal shadow on her chest roentgenogram. Computed tomography showed that a tumor was located in the right upper mediastinum. Resection of the tumor by video-assisted thoracoscopic surgery was performed. Operative findings determined that the tumor originated from the right vagus nerve and was diagnosed as schwannoma by pathological examination.

Introduction

Although a neurogenic tumor is the most common type of tumor in the mediastinum, they rarely involve the vagus nerve. As a type of neurogenic tumor, schwannomas originating from the intrathoracic or mediastinal vagus nerve are relatively rare. Schwannomas are sporadic and benign in the vast majority of cases. Mediastinal vagus nerves, multiple schwannomas, and malignant schwannomas are usually associated with neurofibromatosis. Herein, we report a case of a vagus nerve schwannoma in the right upper mediastinum. The tumor originated near the branch point of the recurrent laryngeal nerve, and although the operation was difficult, we managed to remove the tumor and completely preserved recurrent laryngeal nerve function.

Case report

A chest roentgenogram of a 35-year-old Chinese woman showed a well defined mass located in the right superior lung field. Further contrast-enhanced computed tomography (CT) of the chest showed a clear boundary mass, 40 × 43 × 46 mm in size, in the right superior mediastinum. The mass, with a CT value of about 23 HU, had roughly uniform density. The arterial and venous phase of the enhanced CT showed slightly heterogeneous enhancement of the tumor (Fig 1). The patient denied suffering any symptoms or history of disease in her family, and her physical examination and laboratory test results showed no significant abnormalities. Neurofibromatosis was excluded, as the patient did not exhibit café au lait spots, hyperpigmented skin lesions, multiple neurofibromas, or vestibular tumors. Right-sided video-assisted thoracoscopic surgery (VATS) was performed. A tumor with a round shape was identified in the right superior mediastinum. It originated from and encased the right vagus nerve. Although the tumor seemed to be very close to the recurrent laryngeal nerve, after careful dissection and separation we found that the tumor was fortunately located at a site distal to the branching point and did not directly invade the recurrent laryngeal nerve. The tumor, together with part of the vagus nerve, was completely excised by vagus nerve amputation (Fig 2). The boundaries of the tumor were relatively clear, and the operation was successful without any damage to surrounding structures. The tumor was an encapsulated solid tumor measuring about 40 mm at its greatest dimension. The cut surface was smooth and was a pale yellow color. The diagnosis according to tumor pathology was

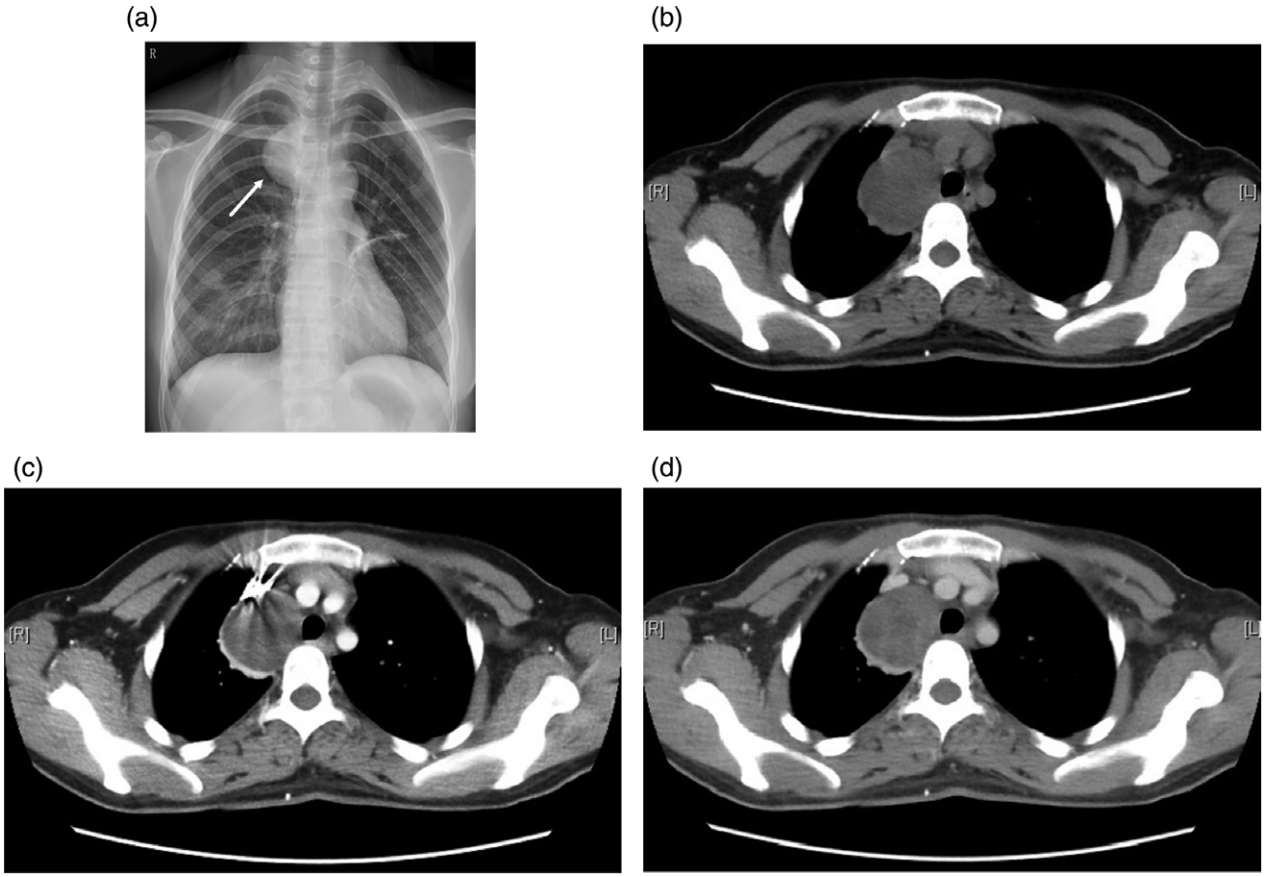

Figure 1 (**a**) Chest roentgenogram showed a right superior lung field mass protruding from the mediastinum (white arrow). (**b**) Computed tomography (CT) scan showed a clear boundary mass in the right superior mediastinum but no evidence of invasion into adjacent tissue. (**c**) The arterial phase of the contrast enhanced CT scan showed slightly heterogeneous enhancement of the tumor. (**d**) The venous phase showed a similar result.

benign vagus nerve schwannoma (Fig 3). The patient's postoperative recovery was uneventful and she did not experience hoarseness.

Discussion

Schwannoma, also known as neurilemmoma, is a tumor originating in the Schwann cells that surround the peripheral nerve fibers. In some earlier studies, neurilemmomas, schwannomas, and neurofibromas were considered the same clinical entity, and the terms are used interchangeably. If not carefully distinguished, this will lead to confusion of some concepts.

Less than 9% of schwannomas occur in the mediastinum[1]; however this kind of tumor is a relatively common mediastinal neurogenic tumor, accounting for about 25.3% of intrathoracic neurogenic tumors.[2] Intrathoracic schwannomas most often arise in a paravertebral location from the sympathetic chain or intercostal nerve. Schwannomas originating from the intrathoracic or mediastinal vagus nerve are rare and atypical.

Mediastinal vagus nerve schwannomas are almost twice as likely to be located on the left than on the right, may occur at any age, and do not show a gender preference.[3] In rare instances, they could be derived from the nerve endings within the esophageal muscularis propria, similar to other primary tumors of the esophagus. Mediastinal vagus nerve schwannomas are difficult to identify before surgery.[4] Generally, they are solitary, multiple schwannomas without malignant components arising from the unilateral or bilateral intrathoracic vagus nerves.[5] These tumors are usually slow growing and asymptomatic; however, a number of symptoms, including chest pain, dysphagia, and dyspnea to varying degrees, may occur when the tumors reach huge dimensions or compress the adjacent structures. Hoarseness may occur when the tumor involves the recurrent laryngeal nerve. Mediastinal vagus nerve schwannomas can become life threatening because of cardiopulmonary complications or tracheal obstruction.[6,7]

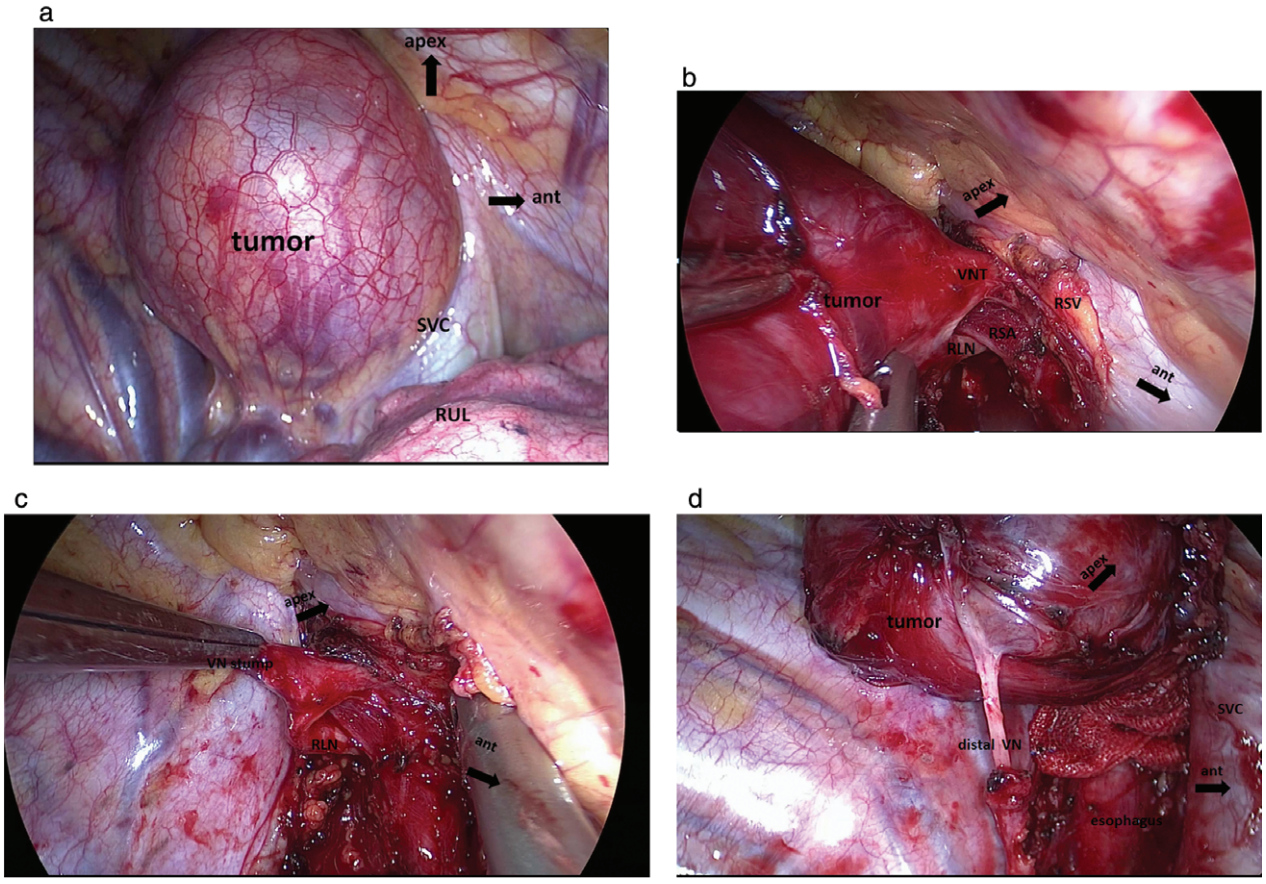

Figure 2 (**a**) Appearance of the tumor under thoracoscopy. ant, patient's front; apex, patient's apex; RUL, right upper lobe; SVC, superior vena cava. (**b**) The tumor is near and distal to the branching point of the recurrent laryngeal nerve. RLN, recurrent laryngeal nerve; RSA, right subclavian artery; RSV, right subclavian vein; VNT, vagus nerve trunk. (**c**) Tumor resection through vagus nerve amputation. VN stump, vagus nerve stump. (**d**) Distal side of the tumor. distal VN, distal side of the vagus nerve.

Schwannomas are benign and well encapsulated most of the time, but a few cases of malignant, very aggressive, locally invasive schwannomas, with a tendency to relapse and metastasize have been reported. Preoperative induction chemoradiotherapy may be beneficial when accompanied by surgery.[8]

Computed tomography and magnetic resonance imaging are only helpful to confirm the size of the tumor and the relationship to adjacent structures. Position emission CT has poor sensitivity and specificity. Fine needle aspiration biopsy is also not recommended. The tumor is difficult to puncture, and the paucity of the specimens obtained will prevent a definitive diagnosis.[9]

Surgical resection via thoracotomy or VATS can confirm the diagnosis and is considered curative. Thoracoscopic surgery is preferred because of its less invasive nature, which is beneficial when the boundaries of the tumor are clear. Both subcapsular enucleation and tumor resection with vagus nerve amputation are optional surgical procedures. It depends on the relationship between the tumor and nerve: either alongside or encased in the vagus nerve. Although subcapsular enucleation appears to be minimally invasive, nerve conduction damage does still occur.[10] The most important issue during the procedure is whether to resect the recurrent laryngeal nerve. Intraoperative nerve monitoring is increasingly used and is worthwhile, as it can help the surgeon effectively identify the recurrent laryngeal nerve when the tumor is closely located and, as a result, achieve a more precise resection to avoid damaging these fragile nerves by accident. However, if the tumor originates from the recurrent laryngeal nerve, and subcapsular enucleation is difficult to realize, sacrificing the laryngeal recurrent nerve is inevitable in this situation in order to resect the tumor completely. During surgery, the patient should be closely observed as severe bradycardia or asystole can occur.[11]

Pathological examination is required to diagnose schwannomas. Schwannomas usually have a complete capsule. A tumor consisting of both Antoni A and B type histologies could be revealed under microscopic examination. Positivity

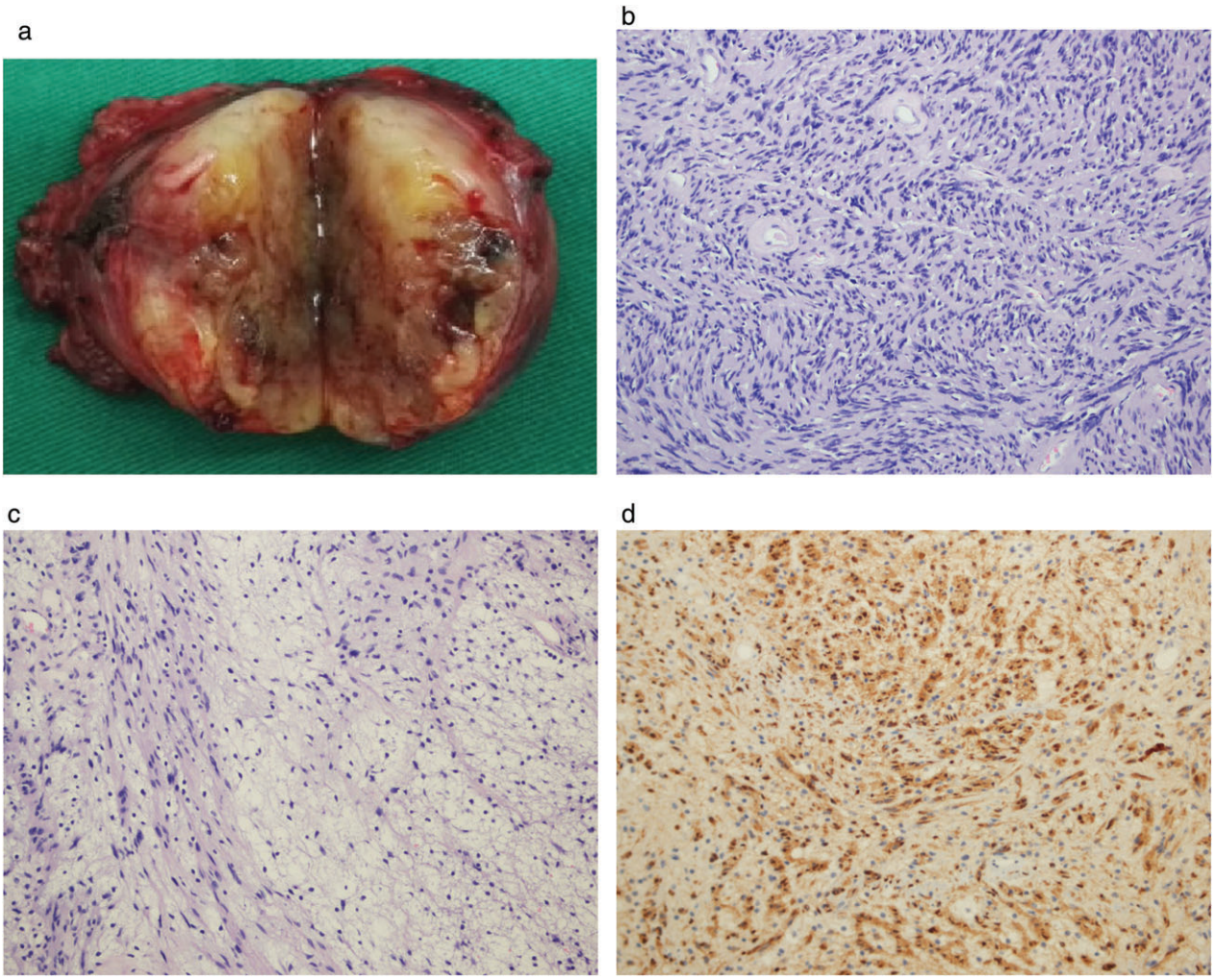

Figure 3 (**a**) Cut surface of the specimen. (**b**) Antoni A zone (hematoxylin and eosin; magnification ×200). (**c**) Antoni B zone (hematoxylin and eosin; magnification ×200). (**d**) Immunohistochemistry for S 100 protein exhibited strong positivity (magnification ×200).

for S-100 protein in immunohistochemical analysis may help to make a final diagnosis. If atypia, mitoses, pleomorphism, and necrosis are identified, a malignant schwannoma should be considered, even though they are extremely rare.[12]

Previous studies have reported satisfactory prognosis of mediastinal vagus nerve schwannomas following complete resection. However, the prognosis of malignant schwannomas is poor, and to date no convincing adjuvant therapy is available.

Acknowledgments

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Disclosure

No authors report any conflict of interest.

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