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Robotic resection of mediastinal left vagus neurofibroma

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ABSTRACT

Neurogenic tumors represent 10 to 34% of all mediastinal tumors and among them, neurofibroma originating from the vagus nerve are rare entities. We present a case of a neurofibroma with cystic degeneration originating from the left branch of the vagus nerve in a 27-years-old man without von Recklinghausen disease. A complete robotic resection of the mediastinal mass has been performed, with amputation of the vagus nerve enclosed in the mass. The postoperative course was uneventful and the patient was discharged in two days.

INTRODUCTION

Intrathoracic neurogenic tumors are unusual neoplasm and represent for 10-34% of all mediastinal tumors; the most common site is the posterior mediastinum, although chest wall, anterior mediastinum, and intrapulmonary localization are the other usual areas. Benign schwannoma or neurofibroma is the most frequent neurogenic tumor, with a rate of malignancy of 5-10% in adults, and 40-60% in children [1].

Among these neoplasms, neurofibroma originating from the vagus nerve are very rare entities and often associated with von Recklinghausen disease [2]. We report a case of a left vagus nerve neurofibroma with cystic degeneration of the upper mediastinum successfully resected by robotic approach.

CASE REPORT

A 27-years-old man was admitted to our department with an occasional finding of a left mediastinal mass during a chest radiography screening. No previous medical history was referred; physical examination, pulmonary function, and laboratory data were normal.

Computer tomography (CT) scan of the chest showed a rounded, well-defined lesion with a mainly peripheral vascularization and some internal vascularized septa (CT value of about 22 HU); the tumor had a maximum transverse diameter of 4.7 cm and a craniocaudal extension of 6.9 cm, with a develops on the left side of the arch of the aorta extending upwards along the course of the left carotid and left subclavian. The mass presented relationships of close continuity with the arterial vascular structures for which a clear adipose tissue cleavage plane was determined all around the tumor (**Fig 1**). A slight compression of the homolateral anonymous trunk at the origin was also present.

A positron emission tomography (FDG PET/CT) revealed a slight standardized uptake value (SUV) of 3 along the wall of the mass, with a hypometabolic central portion (**Fig 2**).

A benign or low-grade anterior mediastinal tumor, such as thymic cyst, thymoma or bronchogenic cyst, was suspected and surgical treatment was planned.

The patient underwent a left-sided robotic surgery in which the whole mass was resected; the procedure was carried out using the Da Vinci Xi surgical system with a three arms approach. The camera port was placed in the 6th intercostal space (ICS), closer the posterior axillary line (PAL), for the use of a 30-degree robotic camera; the following 8 mm robotic instrument ports were placed in the 4th ICS, 2 cm medial to the scapula, and in the 8th ICS, near the scapular line. CO2 insufflation was also used, at a pressure of 6 mmHg and a flow of 6 L/min. The robotic instruments used during the whole procedure were the Maryland bipolar forceps and Permanent Cautery Spatula (Surgeon right hand; arm 1), and the Fenestrated bipolar forceps (Surgeon left hand; arm 2).

The mediastinal pleura was circumferentially opened around the mass; dissection proceeded upwards along the course of the left carotid and left subclavian artery, visualizing a clear plane between the mass and the nearby structures. No nodal or vascular infiltration was seen. As dissection proceeded, we appreciated how both the left proximal and distal portion of the vagus nerve were enclosed by the tumor, which

arose cranially of the left recurrent laryngeal nerve; the tumor was radically excised, with transection of the left vagus nerve (**Fig 3-A, B**).

Macroscopically, the specimen was a mass, with a cystic degeneration, measuring 7 cm in diameter. The outer surface was grey and smooth. On sectioning, the mass was cavitated, the wall appeared 1 to 2 mm thick with a prevalent hemorrhagic and brown appearance. No significant necrosis was seen in the tumor.

The histologic examination showed wall composed by prevalent spindle cell in absence of epithelial lining. The cellularity was moderated and composed of spindle cells and round to ovoid cells (**Fig 4-A, B**), a proliferation of elements of peripheral nerves intermixed with fibroblasts, collagen, slightly myxoid, and mastocytes.

The immunohistochemical profile showed a strong cytoplasmatic stain for S-100, SOX 10 (**Fig 4-C**), CD34 and collagen IV ERG, smooth actin muscle and HMB45 were negative. Proliferation index evaluated with Ki67 was extremely low (5%). A diagnosis of neurofibroma with cystic and hemorrhagic modifications was rendered.

The postoperative recovery was uneventful except of hoarseness; the patient was discharged on postoperative day 2 after chest tube removal. After 6 months of follow-up, no evidence of recurrence was observed, the patient is in good health and with improving dysphonia.

DISCUSSION

Neurofibroma accounts of about 25.3% intrathoracic neurogenic tumors [3]; neurogenic tumors originating from the intrathoracic vagus nerve are very rare [4]. These tumors often occur in association with neurofibromatosis (von Recklinghausen's disease); neurofibroma arising from the vagus nerve and not related with von Recklinghausen disease is instead very rare (about 0.1%) [5].

The patients are usually asymptomatic and the diagnosis is accidental (75-80% of cases); nevertheless, symptoms like dysphagia, chest pain, dyspnea, cough, atrial fibrillation and hoarseness may be present [6].

In our case, the tumor grew from the left vagus nerve before the origin of the recurrent laryngeal nerve; to date, there are few similar cases described in the literature.

Mediastinal vagus nerve neurofibromas are often difficult to recognize before surgery; radiological differential diagnosis comprises mediastinal masses like intrathoracic goitre, thymic carcinoma, thymoma, germ cell tumors, lymphoma, parathyroid adenoma and lymphangioma.

The highly magnified three-dimensional visualization of the operating field, the EndoWrist technology of the robotic instrument and the tremor reduction, allow a depth perception of the structures and permits a clear visualization of the tissue planes, especially in mediastinal procedures in which robotic peculiarities have an important advantage over classical VATS.

CONCLUSION

This case documented the clinical and surgical features of this extremely rare example of neurofibroma with cystic and hemorrhagic modification and should be added to the benign mediastinal tumors with a cystic appearance.

Intratumoral hemorrhage and cystic modifications could develop in neurofibroma and rarely these tumors could arise in anterior mediastinum; this variable presentation and location make its diagnosis difficult and poses greater diagnostic difficulty from a radiological and histological point of view, which may lead to a misdiagnosis of a mediastinal cyst. This case documented the clinical and surgical features of this extremely rare example of neurofibroma with cystic and hemorrhagic modification and should be added to the benign mediastinal tumors with a cystic appearance.

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Figure 1. Computed tomography scan showing a well-defined, anterior mediastinal mass, with peripheral vascularization and some interior vascularized septa.

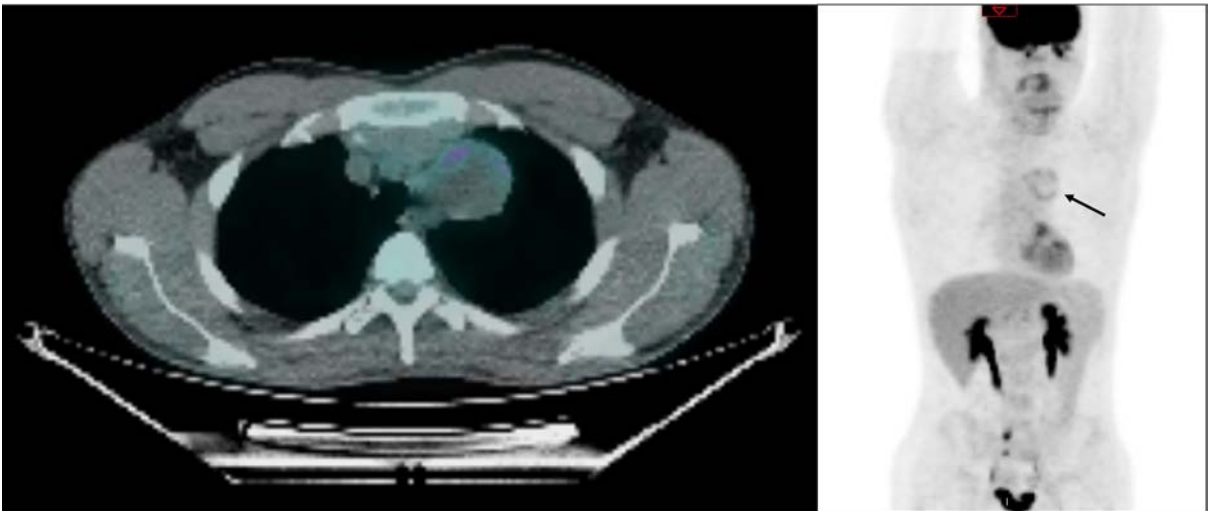


Figure 2. Positron emission tomography demonstrating a slight standardized uptake value (SUV) of 3 along the wall of the mass, with a hypometabolic central portion.

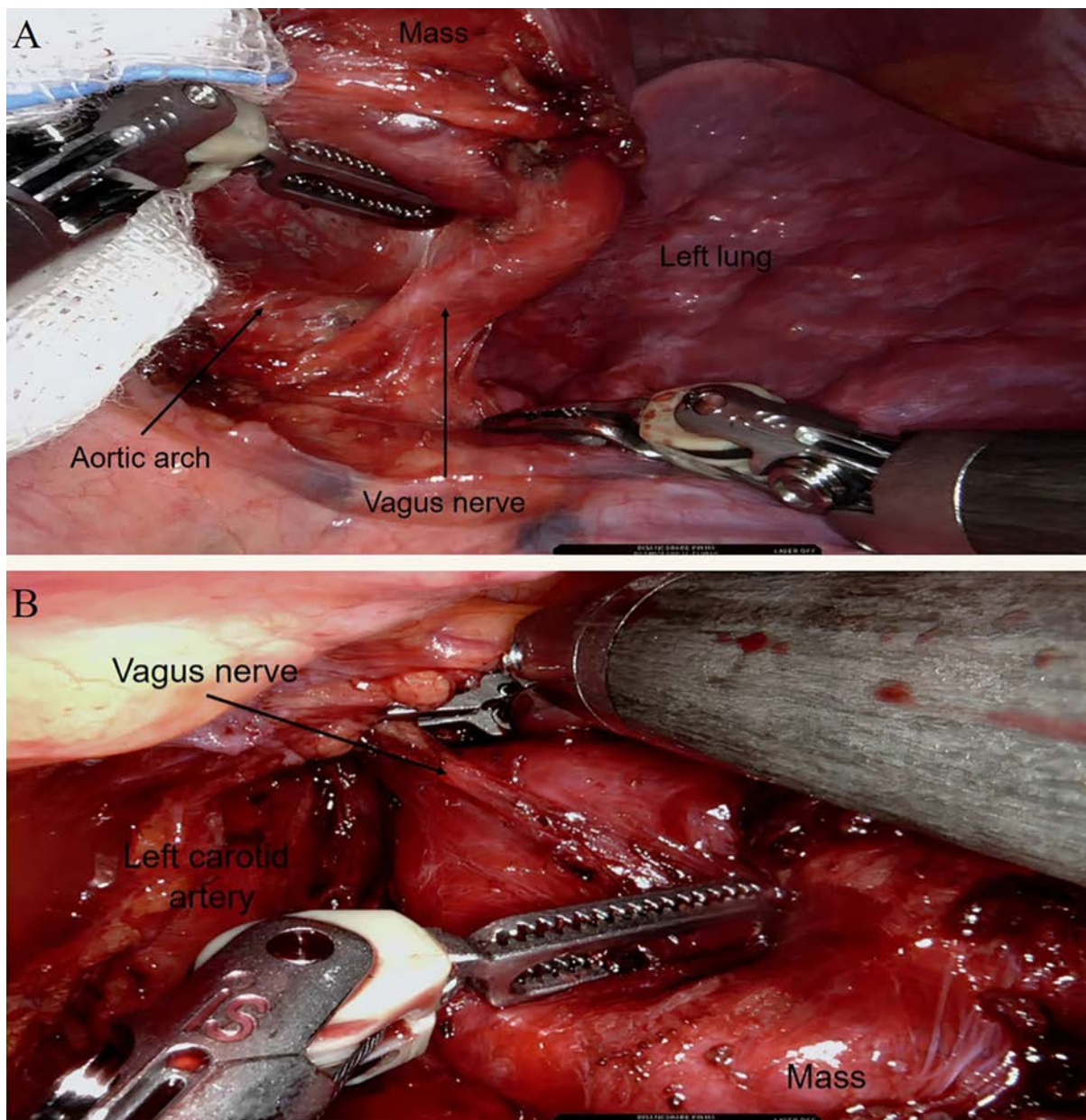


Figure 3. (A-B) Intraoperative view of the mediastinal mass originating from and enclosed the vagus nerve.

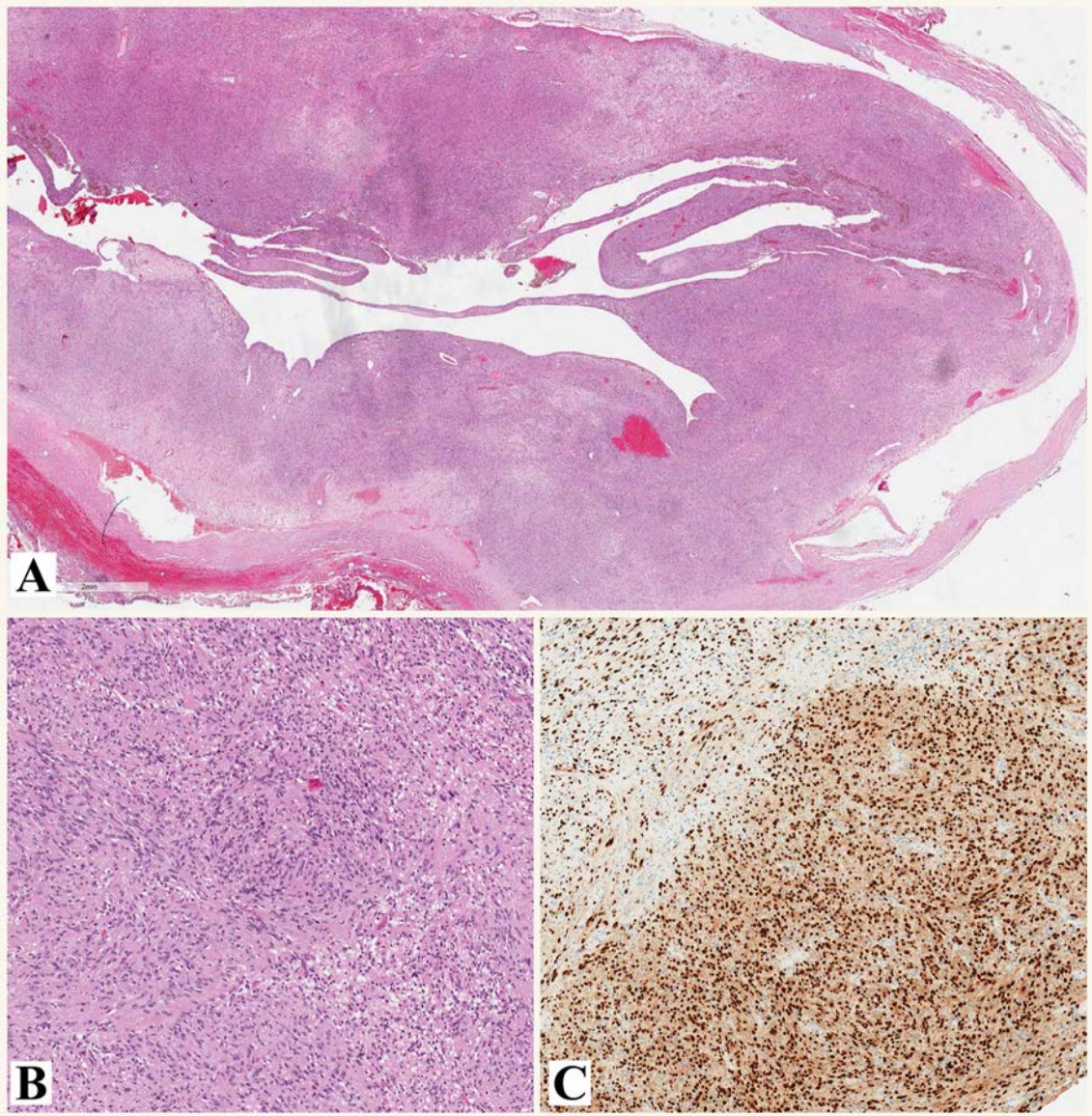


Figure 4. Microscopical section showed a pseudocyst wall without lining cells on the luminal side (A: hematoxylin and eosin 20x of magnification). The tumor was composed by spindle-shaped cells intermixed with bland spindle cells and “shredded” collagen and hemosiderin-laden macrophages (B: hematoxylin and eosin 100x of magnification). Immunohistochemistry revealed nuclear expression for SOX10 (C: immunohistochemistry SOX10 100x of magnification).