

A Case of Primary Endobronchial Neurilemmoma Without Intraspinal Extension

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Neurilemmoma is a benign and slowly growing neurogenic tumor. Intrathoracic neurilemmoma often develops in the chest wall and posterior mediastinum, but endobronchial neurilemmoma is extremely rare. The diagnosis of endobronchial neurilemmoma with preoperative imaging findings is challenging and is usually made via postoperative pathological examination. These authors encountered a case of primary endobronchial neurilemmoma in a 52-year-old woman who had no symptoms. A 3.0 × 2.6 cm mass in the right lower lobe projecting into the mediobasal segmental bronchus was shown in the results of the contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) of the chest. Benign neurilemmoma was confirmed via bronchoscopic biopsy, and surgical resection (sleeve bronchial excision and end-to-end anastomosis) was performed.

Key Words: Neurilemmoma, Bronchi, Pulmonary surgical procedures

INTRODUCTION

Neurilemmoma is a benign, slowly growing tumors of Schwann cells that may arise in any nerve, including cranial nerves, spinal roots or peripheral nerves. Neurilemmoma occurs with equal frequency in men and women. The tumor has been reported in patients of all ages (mean age 41 years).¹

Neurilemmoma of the endobronchial origin is rare and only few cases have been reported previously in the literature. It is conceivable that neurilemmoma rarely occur in the trachea, bronchus, bronchioles and alveoli, even though the exact frequency is unknown.²

Prognosis is excellent, and recurrences are rarely reported after surgical resection. We report a case of primary endobronchial neurilemmoma in a 52-years-old woman. Surgical resection (sleeve bronchial excision and end to end anastomosis) was performed.

CASE

A 52-year-old woman who was taking medication for anxiety disorder presented with anterior chest pain. She was diagnosed with unstable angina, and underwent a coronary angiography with percutaneous transluminal coronary angioplasty. She presented ambiguous abdominal pain and was found to have lung mass in right lower lobe by abdominal CT. But, she had no respiratory symptom and never smoked.

The patient's vital signs included blood pressure of 150/100 mmHg, temperature of 36.5°C, respiratory rate of 20 breaths per minute and oxygen saturation of 97% on room air. There is no abnormal finding by lung auscultation.

Chest X-ray were normal (Fig. 1) and routine laboratory tests revealed only increased LDL-cholesterol (142 mg/dL) with normal liver function, normal renal function and normal coagulation tests.

Contrast enhanced CT of chest showed about a 3.0 × 2.6 cm sized, well-demarcated round mass in right lower lobe projecting into mediobasal segmental bronchus. Bronchoscopy showed a small polypoid lesion in mediobasal segment of the right lower bronchus (Fig. 1). The bronchoscopic biopsy showed that tumor was histopathologically and immunohisto-

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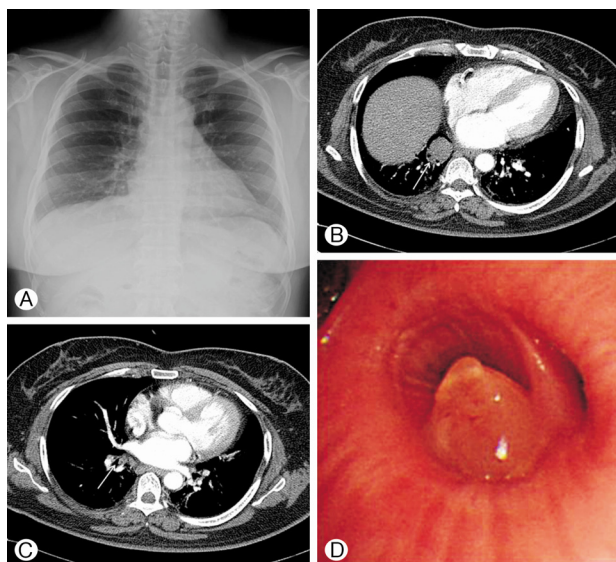


Fig. 1. Chest X-ray is shown to be normal (A), and the chest CT scan image shows an about 3.0×2.6 cm well-demarcated mass in the right lower lobe (B) projecting into the mediobasal segmental bronchus (C) (white arrow). The bronchoscopic view shows a polypoid protruding mass in the mediobasal segmental bronchus (D).

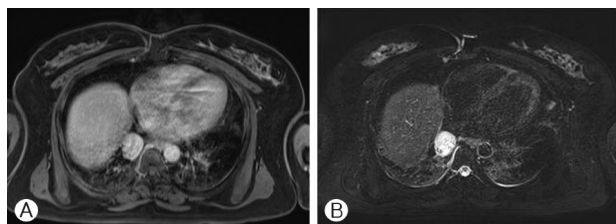


Fig. 2. Enhanced T1-weighted fat suppression image shows a soft-tissue mass with a globular enhancing portion in the right mediobasal segment (A). Enhanced T2-weighted fat suppression image shows a mainly high-signal-intensity lesion at the same site as with A (B).

chemically compatible with neurilemmoma. On MRI, mass had globular enhancing portion on T1-weighted fat suppression images and had areas of high signal intensity on T2-weighted fat suppression images without intraspinal tumor extension (Fig. 2).

The patient underwent an operation for surgical resection. The mass was occupying the mediobasal segment of right lower lobe and connected to mediobasal segmental bronchus. It was measured a $3.5 \times 2.9 \times 2.4$ cm sized oval mass (Fig. 3). A postoperative biopsy was conducted and the subsequent hematoxylin and eosin (HE) stain revealed that the lesion consisted of dense lymphoid infiltration and spindle cell (Fig. 3). The S-100 immunohistochemical stain was positive, leading to our diagnosis that the lesion was a neurilemmoma (Fig. 3).

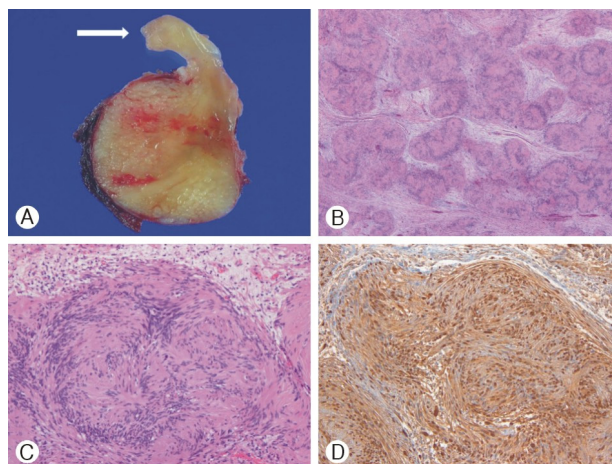


Fig. 3. A $3.5 \times 2.9 \times 2.4$ cm, oval mass is connected to the bronchus (white arrow). The cut section shows a yellowish translucent glistening appearance and the focal area of hemorrhage (A). The tumor shows hypercellular Antoni A and hypocellular Antoni B areas (B: hematoxylin-eosin stain, $\times 40$). The tumor cells are spindle-shaped and have wavy, tapering nuclei (C: hematoxylin-eosin stain, $\times 100$). The tumor cells are positive for S-100 protein (D: immunohistochemical stain, $\times 100$).

DISCUSSION

Neurilemmomas are typically single, circumscribed, encapsulated tumors attached to any nerve, including cranial nerves, spinal nerves or peripheral nerves.¹ More than 90% of neurilemmoma are benign.³ Neurilemmoma occurs with equal frequency in men and women.² The tumor has been reported in patients of all ages (mean age 41 years).¹⁻³

Neurilemmoma may occur in any peripheral nerve and is often found in the chest wall and the posterior mediastinum.⁴ Although being very rare, pulmonary neurilemmoma can occur in all respiratory tracts including the trachea, bronchus, bronchioles and alveoli.^{5,6} Only few cases of pulmonary neurilemmoma was reported in Korea.⁷

Neurilemmoma have an equal distribution in trachea, central bronchus and pulmonary parenchyma.⁶ They usually have a long natural history, causing symptoms only after they have attained a large size. The tracheobronchial tumors can present with respiratory dyspnea, wheeze, dry or productive cough, chest pain and hemoptysis.^{8,9} Bronchial obstruction can cause recurrent pneumonia and lung collapse.¹⁰

The Contrast enhanced CT of chest is helpful in assessing lung parenchyma distal to obstruction and delineation of the tumor inside the lumen as well as outside the wall of the respiratory tract. The computed tomographic appearance of neurilemmoma usually consists of a well demarcated round or

oval mass.

Neurilemmomas can cause deformity or erosion of the vertebra, neural foramen and ribs. Ten percent of neurilemmomas grow through the adjacent intervertebral foramen and extend into the spinal canal with a "dumbbell" or "hourglass" configuration. Tumors with dumbbell intraspinal extension with mass effect on the spinal cord are best excised with combined neurosurgical and thoracic procedures. So, MRI should be performed preoperatively in all patients to exclude definitely intraspinal tumor extension.¹¹

The MRI appearance of neurilemmomas arising from cranial or spinal nerves has been well described in the literature, but that of an endobronchial neurilemmomas has not been reported previously. On MRI, neurilemmoma typically had low to intermediate signal intensity on T1-weighted images and had areas of intermediate to high signal intensity on T2-weighted images. Very high signal intensity areas on T2-weighted images correspond pathologically to areas of cystic degeneration. Occasionally, gadolinium enhanced MRI images show contrast enhancement in peripheral portions of tumor and no enhancement in areas of cystic degeneration.¹² In our case, round mass in right lower lobe have typical MRI finding of neurilemmoma without evidence of intraspinal tumor extension. Histologically, they consist of two components. One is a highly ordered cellular component (Antoni type A tissue) and the other is loose myxoid component (Antoni type B tissue).¹³ Diagnosis of pulmonary neurilemmoma is made based on the typical Antoni A and B formation in HE stain and positive S-100, Leu-7, vimentin in immunohistochemical stains.^{2,10}

The definitive treatment is surgical resection with a healthy margin of tissue.^{9,14} Surgery include tumor resection with bronchial excision, sleeve bronchial excision and end to end anastomosis, lobectomy, pneumonectomy.

Endoscopic transbronchial electrical snaring and neodymium-doped yttrium aluminium garnet (Nd-YAG) laser abrasion are reported for histopathologically proven benign lesions.^{15,16} However, there is a possibility of local recurrence over a long interval. The extraluminal progression can lead to a recurrence after endoscopic resection of intraluminal lesion. Especially, in aged patients or patients with severe underlying diseases or malignancies, a bronchoscopic removal should be employed.⁶ An endoscopic approach was not ideal for our patient due to lung parenchymal involvement. So she underwent an sleeve bronchial excision and end to end anastomosis for curative resection. Postoperative histologic analysis revealed a benign neurilemmoma of Schwann cell origin. She is doing well at 2 months follow-up.

Neurilemmoma is a slowly growing benign tumor, so pro-

gnosis is excellent. Recurrences are rarely reported after surgical resection. Moreover, neurilemmomas can grow through the adjacent intervertebral foramen and extend into the spinal canal. In that case, tumors must excised with intraspinal extension on the spinal cord with combined neurosurgical and thoracic procedures. So, MRI may be helpful for providing the anatomic details and characteristic findings of neurilemmoma. If we have no consideration for intraspinal extension, incomplete resection can cause recurrences subsequently. Accordingly, MRI should be performed preoperatively in all patients. In previous other study, there was no mention about MRI. So, we was experienced a case of a primary endobronchial neurilemmoma without intraspinal tumor extension on MRI and thoracic surgical resection was performed.

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