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Case Report

Pulmonary function test detects a case of primary tracheal neurilemmoma presenting as an asthma

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Abstract: In tracheobronchial tree, neurilemmomas are exceedingly rare tumors and their diagnoses are always delayed due to insidious and non-specific clinical presentation. We report a case of primary tracheal neurilemmoma in a middle-aged man who has had intractable cough and breathlessness for over 2 months. Physical examination was normal except for auscultated wheezes. Pulmonary function test suggested a platform-like change of expiratory phase and chest computed tomography (CT) demonstrated a nodule occupying 20% of the lumen in upper trachea. Then the mass was resected with snares and confirmed as neurilemmoma histologically. The patient reported symptomatic relief after the endoscopic intervention. This case shows that pulmonary function test may reveal an upper airway obstruction and aid in the early diagnosis of tracheal neurilemmomas. Moreover, clinicians should consider the possibility of tracheal masses when patients present with intractable asthmatic symptoms.

Keywords: Tracheal neurilemmoma, intractable cough, breathlessness, pulmonary function test

Introduction

Primary tracheal neurilemmomas are extremely rare among primary tracheal tumors and only around 36 cases have been reported in literature since 1950 [1]. The tumors may present with features of airway obstruction including dyspnea, wheezing, stridor and cough, so they are often mistakenly treated as asthma [2]. Although they are histologically benign, they can lead to recurrent infections [1], unnecessary long term use of corticosteroids [3] and even life threatening situations [4] due to missed or delayed diagnosis. We report a case of primary tracheal neurilemmoma in a 52-year-old male with complaints of intractable cough and breathlessness. In addition, we provide a review emphasizing the clinical presentation and diagnostic methods of this rare entity.

Case presentation

A 52-year-old man was admitted to our hospital for 2-month dry cough and breathlessness. He experienced aggravating breathlessness associated with wheeze in the preceding week prior to admission. The symptoms were more severe in squat body position and had progressed despite one-week treatment with bronchodilators and inhaled steroids. He denied expectoration, fever, chest pain, hemoptysis and weight loss. He had a 15-year smoking history, but had neither history of irritability rhinitis nor previous airway diseases, such as asthma, bronchiectasis and tuberculosis. On physical examination, auscultation revealed bilateral inspiratory and expiratory wheeze in both upper parts of chest. The remainder of the systemic examination was unremarkable. Laboratory tests including total IgE (TIgE), procalcitonin, erythrocyte sedimentation rate, carcinoembryonic antigen and total Cyfra21-1 were within normal range. Antibody of tuberculosis was also negative.

Pulmonary function test showed very severe obstructive ventilatory defect (FEV 1% Pred 27%) and a platform-like change of expiratory phase (**Figure 1**), suggestive of a variable intrathoracic upper airway obstruction caused by unknown reasons. Bronchial dilation test was negative. A nodule round with well-defined margins occupying 20% of the lumen in the upper trachea was revealed by chest computed tomography (CT) (**Figure 2**). Moreover, the nod-

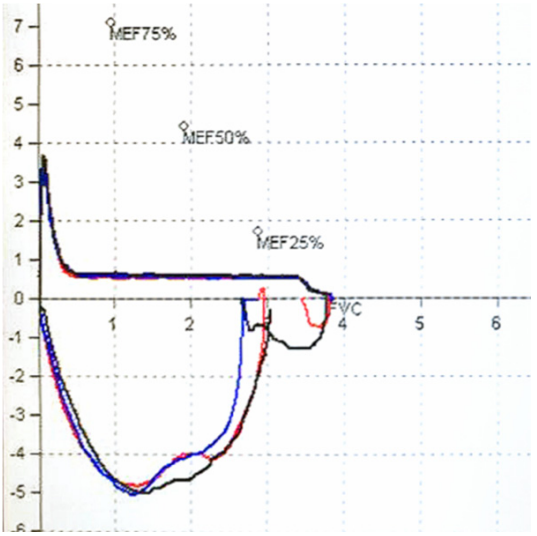

Figure 1. Pulmonary function test suggesting a platform-like change of expiratory phase.

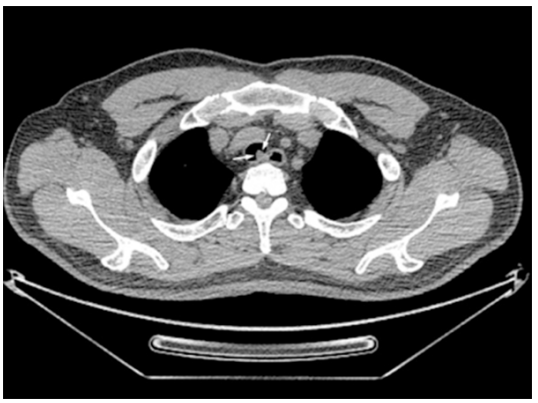

Figure 2. Chest CT revealing an intratracheal nodule occupying 20% of the lumen (arrows).

ule showed normal fluorodeoxyglucose (FDG) uptake in positron emission tomography (PET) with a standardized uptake value of 1.5. Bronchoscope examination demonstrated a 0.6 cm×0.7 cm mass located 3 cm distal to the vocal cords. It was pedicled on the posterior aspect of trachea and had a smooth surface rich in capillaries (**Figure 3A**). The mass was resected by using snare and there were no complications after the operation (**Figure 3B**). Microscopically, the mass revealed spindle-shaped cells arranged densely in fascicles in the submucosa and looser, myxoid regions in the center area. Focal nuclear palisading was present and no obvious mitotic activity or necrosis was observed (**Figure 4A**). Immunohistochemical staining was positive for S-100 protein and vimentin (**Figure 4B**), but was negative for synaptophysin, neuron specific enolase and actin. The diagnosis was consistent with tracheal neurilemmoma.

Fortunately, the patient reported symptomatic relief after removal of the tumor. He was discharged on the 10th postoperative day without cough and breathlessness. There have been no recurrent signs throughout 1 year of follow-up.

Discussion

Tracheal neurilemmomas are rare benign peripheral nerve sheath tumors affect patients of all ages with no predilection to sex, which do not transform to malignancy with few exception of atypical type [5].

Our patient was admitted to the Pneumology Department primarily for progressive cough and breathlessness. Similar to neurilemmomas in other parts of the body, the symptoms of the tracheal ones are mainly caused by involvement of surrounding tissues and mass effect. The main complaints of these patients are progressive dyspnea, recurrent cough, and other asthma-like symptoms, which are similar to our patient.

It occurred to us he might be suffering an asthmatic attack with the wheezing in his chest. However, the impotence of conventional drugs and laboratory tests failed to support the diagnosis of asthma. Then a pulmonary function test was conducted to assist diagnosis, interestingly, it showed a platform-like change of expiratory phase, and very severe obstructive ventilatory defect, suggestive of a variable intra-thoracic upper airway obstruction which greatly raised the possibility of an occupying lesion to us. Consequently, chest CT and bronchoscopy were performed and the results revealed a mass obstructing the lumen. Resected by snare, the mass was pathologically confirmed as neurilemmoma.

Although providing simply nonspecific results, radiologic exams are frequently used to help determine the location, size, degree of extension and metastasis of the tumors. Routine posteroanterior chest X-rays are frequently unrevealing and interpreted normal due to superimposed soft tissues and bone structures [6]. Though being effective means of delineat-

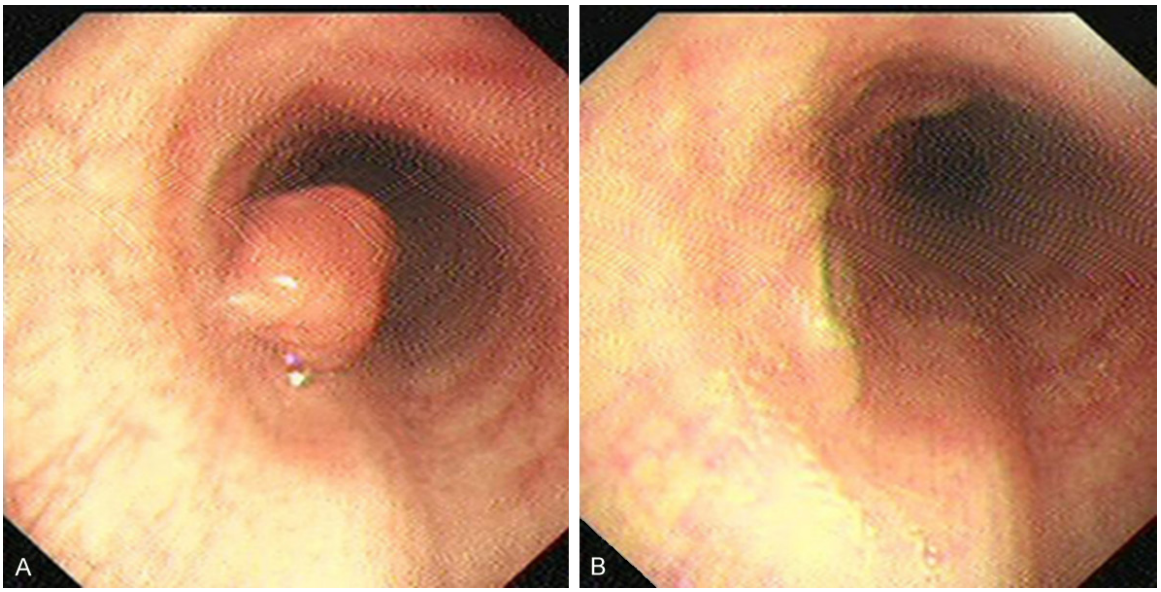

Figure 3. A. Bronchoscopy showing a neoplasm on posterior wall of the trachea with smooth surface rich in capillaries. B. There was no obvious bleeding after tumor resection.

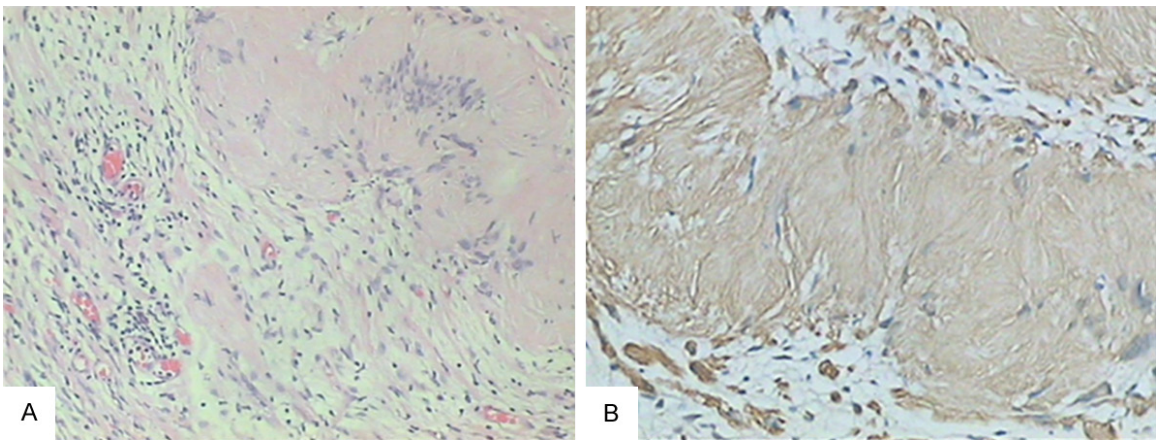

Figure 4. Pathological findings. A. Tumor cells growing in a typcal Antoni A pattern on the left and Antoni B pattern on the right (H&E stain, ×200). B. Immunohistochemical staining of the cells was positive for S-100 protein (×200).

ing lesions and their relationship with surrounding structures, CT and MRI are not able to distinguish benign from malignant tumors. Under conditions where biopsy is not allowed, PET scan can become an alternative modality to enable the diagnosis and facilitate clinical planning [7-9]. Undoubtedly, the definite diagnosis of tracheal neurilemmomas depends on pathological data.

This case remind us in clinical work, intratracheal masses should be considered under such conditions as patients present with breathelessness, other asthmatic symptoms invalid to conventional therapy [2] and pulmonary function test reveals an obstructive event. In previous reported cases, more than 70% of the lumen occluded by neurilemmoma would commonly cause such asthma-like symptoms [10, 11]. However, in our case, 20% occlusion of the lumen caused severe airway obstruction. It immensely draws our attention on the significance of early diagnosis and treatment of neurilemmoma because of the progressive symptoms. As a consequence, patient's life quality will be greatly improved after the resection.

Herein, we should note the significance of pulmonary function test in the diagnosis of this rare entity. The primary purpose of pulmonary

function test is to identify and evaluate the severity of pulmonary impairment [12]. It is one of the most common and simple clinical examinations and plays an important role in diagnosis of respiratory diseases such us chronic obstructive pulmonary disease and asthma. It can assist in the diagnosis of obstructive abnormalities in upper airway. In small-scale hospitals where CT and PET-CT are unavailable, it is a good option to estimate patient's symptoms. If the results imply obstructive abnormalities in upper airway, special attention must be paid to tracheal masses such as neurilemmomas.

Disclosure of Conflict of Interest

None.

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