Primary Tracheal Neurilemmoma Mimicking Asthma: A Case Report

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Primary tracheal tumors are rare; most of them are malignant, and benign tracheal tumors are seldom seen. Because of their non-specific symptoms or signs, late diagnosis may be attributed to a silent tumor, to a denying patient, or to a physician’s delay. We report a case of primary tracheal neurilemmoma which was initially diagnosed and treated as asthma. However, the symptoms were not relieved. Five months later, he received chest CT and bronchoscopic examinations. The results revealed an endotracheal tumor. Segmental resection and reconstruction with primary anastomosis were performed. Pathology showed a neurilemmoma. After operation, this patient returned to normal life without sequelae, and regular follow-up at the OPD showed good surgical results. *(Thorac Med 2005; 20: 684-689)*

Key words: tracheal neurilemmoma, asthma, tracheal reconstruction

Introduction

Primary tracheal tumors are rare; most of them are malignant, and benign tracheal tumors are rarely encountered [1]. According to Weber and Grillo and Masaoka *et al*., the majority of benign tracheal tumors are squamous papilloma in adults, followed by leiomyoma, hemangioma, and neurogenic tumor [2]. Of the neurogenic group, neurofibromas are the most common, with neurilemmoma forming approximately one third of this group [3]. We report a case of primary tracheal neurilemmoma which had a delayed diagnosis due to an initial presentation mimicking asthma.

Case Report

A 20-year-old man without underlying disease or a smoking history had suffered from cough and repeated upper respiratory infection since he was 15 years old. He went to a local medical department and asthma was suspected. The symptoms persisted although medication was given. The productive cough progressed for 5 months, and he went to another medical hospital where expiratory wheezes were noted. Flexible bronchoscopic examination revealed a 1.8x1.5 cm protruding mass, located at the lower trachea, with nearly total obstruction of the lumen. Hyperemia with irregular vessels was noted on the tumor surface. Because of the nature of the mass,
no attempt was made to advance the bronchoscopy through the lesion, and biopsy was not performed due to the risk of bleeding. Computed tomography was performed to delineate the distal extension of the mass, and demonstrated an oval endotracheal lesion about 1.5 cm in diameter, occupying the low trachea, 2 cm proximal to the carina (Figure 1). There was no mediastinal extension or adenopathy. Under the impression of tracheal tumor, he was referred to our hospital for further management. Inspiratory wheezing was more prominent during auscultation of the trachea. The remainder of the physical examination was normal. With careful inspection, the chest roentgenogram showed a mass shadow in the lower trachea, which represented the tumor (Figure 2). Pulmonary function studies demonstrated a severe obstructive ventilatory defect (FEV1: 0.97L, 20.05% of predicted value; FEV1/FVC: 19.2%). The flow-volume curve displayed a fixed airway obstruction (Figure 3).

During operation, a 2.5 cm segment of the trachea was resected, and primary anastomosis was performed. The initial approach was through a right posterolateral thoracotomy. Intraoperative bronchoscopy was used to locate the exact tumor site and its extension. A U-shaped incision was made at the right hilar area, and the pericardium was opened for hilar release. In this way, primary tracheal anastomosis could be achieved without tension.

The specimen of resected trachea was measured at 2.5x1.5x1 cm in size, in a fresh state. Grossly, there was an oval, pink, well-defined and thinly encapsulated tumor, which grew from the membranous portion, and nearly totally occluded the tracheal lumen (Figure 4). The tumor was yellowish-white, gelatinous, and soft on cutting. Microscopically, the endotracheal tumor was composed of spindle cells in a loose reticular background (Antoni B); some tumor cells were in a palisading pattern (Antoni A) (Figure 5). The overlying epithelium revealed focal squamous
metaplasia. The tumor cells were immune reactive to S-100 protein (Figure 6), but not smooth muscle actin. Thus, a neurilemmoma was confirmed.

Postoperative follow up by bronchoscopy at 3 and 6 months showed good healing of the anastomosis without granulation tissue or stricture.

**Discussion**

The neurilemmoma was first described by Verocay, and later named neurilemmoma by Stout, who attributed its histogenesis to an embryonic neuroectoderm; it is characterized by a proliferation of Schwann cells [3]. The neurilemmoma is a slow growing tumor that may occur at any age, but mostly in middle-age. A malignant variety exists, but no transformation from benign to malignant neurilemmoma has been reported. Grossly, neurilemmomas are typically solitary, circumscribed and encapsulated lesions that are pink, yellow or pearl-gray in color. The most common site of tracheal neurilemmoma is the distal trachea, followed by the proximal trachea and the middle third of the trachea [4-5]. Histolo-
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Historically, Antoni described 2 types of neurilemmomas: Type A has an orderly arrangement of elongated cells with blunted ends, anastomosing cell bodies, and most characteristically, palisading nuclei; wire-like reticular fibers also are present. Type B lacks orientation and consists of haphazardly arranged cells with intercellular edema imparting a loose-textured appearance. Microcysts are present as well. These 2 configurations often coexist in a given tumor [3]. In addition, in immunohistochemical stains, we find that neurilemmomas are positive for S-100 protein, an acidic protein common to supporting cells of the central and peripheral nervous system [1-2].

Like other intraluminal tumors, symptoms of airway obstruction predominate and are determined by tumor bulk and location, including dyspnea, cough, hemoptysis, wheezing and/or by the effects of distal atelectasis or pneumonia. However, these symptoms can also be present in other pulmonary diseases, such as asthma or chronic obstructive pulmonary disease (COPD). As tracheal neurilemmomas manifest with a cough and wheezing or stridor, several cases (including ours) have been mistakenly treated as asthma [5-8]. Due to the slow growth and rarity of these tumors, the average diagnosis is 10-15 months from the onset of symptoms [4]. By this time, it is not uncommon for 50-75% of the tracheal lumen to be occluded by the tumor before leading to symptoms [7]. But the diagnosis may be suggested, as in this case, by a flow-volume loop demonstrating localized fixed obstruction of the central airways. Thus, this combination of atypical symptomatology and spirometry indicated that further evaluation was necessary, leading to a correct diagnosis.

Another diagnostic challenge of tracheal tumor is the fact that routine posteroanterior chest X-rays are frequently unrevealing [9]. However, with close inspection of the tracheal air column, some cases (as in ours) will reveal a filling defect that can be easily missed on an otherwise normal chest radiograph. CT scan of the chest or magnetic resonance imaging may yield more valuable data on the tumor size, location and extension. The cornerstone diagnostic modality is bronchoscopy [10].

Labeling a wheezing patient as “asthmatic” without a demonstrable reversibility of airway flow obstruction may lead to a significant delay in diagnosis and proper therapy. Cases such as this should observe the age-old adage that, “not all that wheezes is asthma”. So those patients with adolescent or adult onset wheezing, cough and shortness of breath who do not respond to accepted medical regimens deserve systematic evaluations of their entire airway [4].

References

以氣喘症狀表現的原發性氣管纖維鞘瘤：病例報告

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原發性氣管腫瘤很罕見，大部分是惡性，良性的更少遇見。由於症狀的表現並無特異性，診斷的延遲可歸因於安靜的腫瘤，病人的否認或是檢查者的延遲。我們在此報告一個原發性氣管纖維鞘瘤的病例，因開始被當作氣喘治療，但症狀並無改善，四個月後，病人接受了電腦斷層與支氣管的檢查，兩個結果都顯示為氣管腫瘤，經由手術做部分氣管切除及重建，術後病人恢復良好，門診定期追蹤的結果並無氣管息肉或狹窄的情形發生。(胸腔醫學 2005; 20: 684-689)

關鍵詞：氣管神經纖維鞘瘤，氣喘，氣管重建