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Case Report

A glomus tumor of the lung of uncertain malignant potential: a case report

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Abstract: Glomus tumors are relatively rare tumors, especially those of bronchopulmonary origin. Fewer than 50 cases of glomus tumors of the lung have been reported in the English medical literature, and the majority of them were clinically benign. We report on a 15-year-old boy with a nodule in the left lung bronchus. The microscopic examination revealed a glomus tumor with an uncertain malignant potential.

Keywords: Glomus tumor, uncertain malignant potential

Introduction

Glomus tumors are unusual neoplasms composed of cells resembling a glomus body with the immunohistochemical and ultrastructural properties of smooth muscle [1]. Most glomus tumors occur in the subcutis or dermis of the distal extremities, but in rare cases they have been reported in uncommon locations such as the visceral organs of the gastrointestinal tract, the genitourinary tract, the liver, and the lungs [1-3]. The majority of glomus tumors are solitary with a benign clinical behavior, and a malignant glomus tumor is extremely rare [4]. Herein we present the first Taiwanese case of a glomus tumor of uncertain malignant potential in the respiratory tract.

Case report

A 15-year-old boy with a persistent cough for several weeks was admitted to the hospital, and the chest X-ray showed the left lower lobe of lung with haziness, suggesting lobar pneumonia. The subsequent computed tomography (CT) scan revealed an endobronchial mass, measuring 3 × 1.7 × 1.6 cm in size, obstructing the main bronchus of the left lower lobe (**Figure 1**). The initial radiological impression was carcinoid tumor. The patient received a video-assisted thoracoscopic lobectomy of the lower lobe of the left lung.

The pathological examinations of the endobronchial lesion showed a glomus tumor, composed of nests and sheets of round to polygonal cells of a relatively uniform size, distinct cell borders, amphophilic cytoplasms and centrally-located, inconspicuous nucleoli (**Figure 2A**). The tumor nests were surrounded by intervening, capillary-sized thin-walled vessels and fibrous stroma with mild hyalinization and myxoid change (**Figure 2B**). There was no marked nuclear atypia, and the mitotic count was low (less than one mitosis per 50 high power fields).

Immunohistochemically, the tumor cells expressed smooth muscle actin and pericellular type IV collagen (**Figure 3A** and **3B**), but they were negative for epithelial markers (pan-cytokeratin and epithelial membrane antigen), neuroendocrine markers (synaptophysin, chromogranin A and CD56), and S100 protein. The melanoma and perivascular epithelial cell tumor (PEComa) marker HMB-45 was also not expressed (not shown). The Ki-67 proliferation index was about 10% (**Figure 3C**).

The entire tumor was confined within the left main bronchus lumen without infiltration or destruction of the bronchial wall. There was no invasion into the lung parenchyma or metastasis to the regional lymph nodes. The final diagnosis of glomus tumor of uncertain malignant potential was made according to histomorphol-

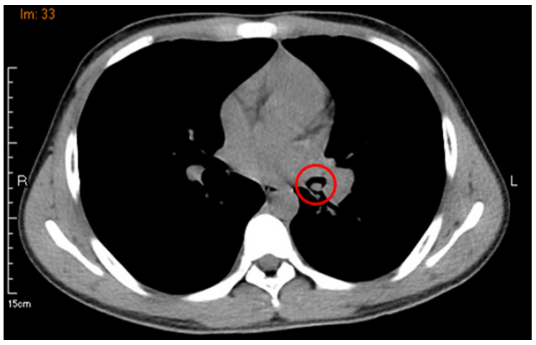

Figure 1. The CT revealed an endobronchial mass (red circle) obstructing the main bronchus of the left lower lobe.

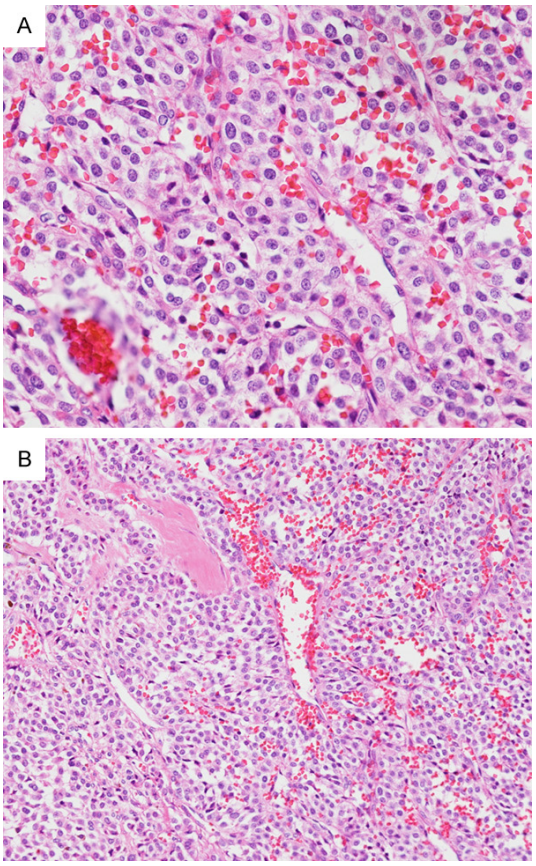

Figure 2. A. The tumor showed nests and sheets of round to polygonal cells of a relatively uniform size, distinct cell borders, amphophilic cytoplasms and centrally located, and inconspicuous nucleoli. B. The tumor nests were surrounded by intervening capillary-sized thin-walled vessels and fibrous stroma with mild hyalinization and myxoid change.

ogy, tumor size, and tumor location. The patient recovered well and showed no evidence of tumor recurrence after regular follow-up for two years up to date.

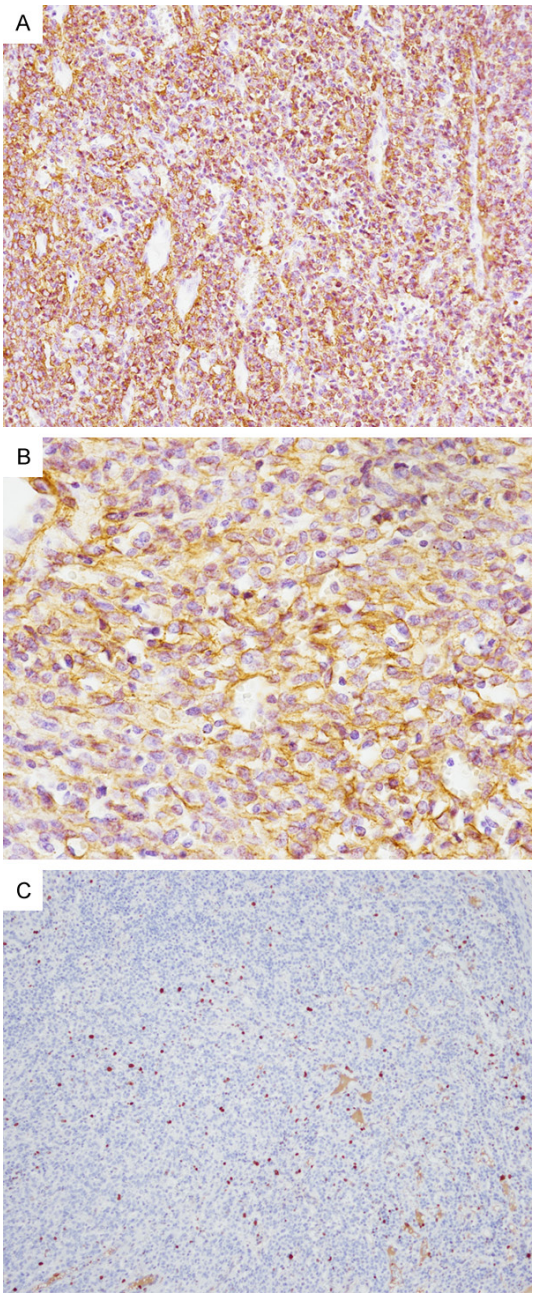

Figure 3. Immunohistochemical studies: (A) tumor cells express smooth muscle actin, (B) pericellular type IV collagen, and (C) the Ki-67 proliferation index is about 10%.

Discussion

The current WHO classification of soft tissue tumors divides glomus tumors into benign glomus tumor, glomangiomatosis (multiple/familial lesions) and malignant glomus tumor (glomangiosarcoma). Histopathologically, a malignant glomus tumor should satisfy one of the two following criteria: (1) marked nuclear atypia with

any level of mitotic activity, or (2) presence of atypical mitosis. Glomus tumors larger than 2 cm in size, located in deep visceral organs but not fulfilling the malignancy criteria, are designated as "glomus tumor with uncertain malignant potential", which has been shown to have much more indolent biological behaviors [5]. Up to date, fewer than 50 cases of pulmonary glomus tumor have been reported in the English medical literature [4]. The rarity of this tumor makes the differential diagnosis difficult, and the tumor may be misdiagnosed as a carcinoid tumor because of the uniformity of tumor cells and the relatively higher incidence of carcinoid tumors in the lungs, particularly from the imaging study. Other differential diagnoses of the glomus tumor include the solitary fibrous tumor, the sclerosing pneumocytoma, and PEComa. Although the tumor is rare, a thorough clinical and histopathological examination with ancillary immunohistochemical staining, such as smooth muscle actin and collagen IV, should help pathologists recognize this entity and avoid an erroneous diagnosis.

Disclosure of conflict of interest

None.

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