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Case report

Primary pulmonary glomus tumor of uncertain malignant potential: A case report with literature review focusing on current concepts of malignancy grade estimation

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ABSTRACT

We report a 38-year-old woman with a left lung tumor presenting as obstructive pneumonia. Bronchoscopic examination revealed a polypoid tumor filling the left main bronchus. The tumor was partially resected by a snaring procedure for diagnostic purposes. Microscopic examination revealed a submucosal tumor located underneath normal bronchial epithelium. The tumor was composed of sheets of uniform oval to cuboidal cells encompassing numerous blood vessels. Immunohistochemically, the tumor cells exhibited smooth muscle markers, but were negative for neuroendocrine markers. The diagnosis of primary pulmonary glomus tumor was therefore made. Subsequent bronchoscopic intervention allowed us to pin-point the origin of the tumor: superior segmental $B^{6a/b}$. She underwent a left lower lobe superior segmental resection successfully. Glomus tumors are relatively rare soft tissue tumors, and those of bronchopulmonary origin are exceedingly rare clinical condition. Among primary lung tumors, the carcinoid tumor is a mimic of the glomus tumor, and differentiating these tumors is known to be difficult, especially using small biopsy samples. In the present case, a large tissue sample obtained by bronchoscopic snaring was quite useful for the correct preoperative diagnosis. Because of the disease rarity, malignancy grade estimation of visceral glomus tumors has not been clearly addressed. Recently, the histopathological diagnostic criteria for malignant glomus tumors was defined in the WHO classification of soft tissue and bone tumors 4th edition. Here we also reviewed the literature on primary bronchopulmonary glomus tumors with special attention to the current concept of malignancy grade estimation.

1. Introduction

Glomus tumors are relatively rare soft tissue tumors composed of cells that resemble the modified smooth muscle cells of the specialized form of arteriovenous anastomosis “glomus body” [1–4]. The most common site of the tumor is the subungual region; however, they occasionally occur in visceral organs such as airway tracts [1,2]. Primary glomus tumors of the lung are exceedingly rare, and the diagnostic and therapeutic strategies for this rare condition have not been well established. Here, we report a case of primary pulmonary glomus tumor that arose in a left segmental bronchus as a protruding polypoid mass with a clinical manifestation of obstructive pneumonia. In the present case, bronchoscopic intervention became a powerful tool not only for histological diagnosis but also for determining the proper operative procedure for the tumor. Ever since the criteria for the diagnosis of malignancy in glomus tumors was first established in 2001 [5], the malignancy estimation of visceral glomus tumors is a worrisome problem to be addressed because of the rarity of this condition. Recently, the criteria were modified and employed in the WHO classification of soft tissue and bone tumors 4th edition [4]. However, pulmonary glomus tumors diagnosed by the current WHO criteria have been scarcely reported. We also reviewed previous cases of primary bronchopulmonary glomus tumors in the literature, with special attention given to current diagnostic criteria.

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2. Case presentation

A 38-year old woman visited a local hospital with the chief complaint of high fever and was diagnosed with pneumonia. She had no history of smoking. She had a past medical history of bronchial asthma, but asthmatic symptoms had ceased for a long time. Although antibiotic treatment was properly initiated, her symptoms persisted. Chest CT examination revealed a left lower lobe atelectasis with a water density mass in the left bronchus (Fig. 1a). Bronchoscopic examination revealed a polypoid mass in the left main bronchus. She was suspected of having a primary bronchial tumor and was referred to our hospital for further examination. Physical examination revealed decreased air entry in the left lower lung. Routine hematological and chemical laboratory results were normal, except for a slight increase in CRP (1.0 mg/dl). Upon initial bronchoscopic examination, a polypoid tumor was observed that occluded nearly 90% of the lumen of the left main bronchus (Fig. 1b). For confirmation of the tumor type, a partial resection of the tumor was performed by bronchoscopic snaring. Postoperative chest X-ray revealed improvement of the atelectasis, and she was tentatively discharged. The partially resected surgical specimen consisted of tumor tissue that measured 1.5 cm in diameter (Fig. 1c). The tumor was well-circumscribed, firm, and tan in color (Fig. 1d). Microscopically, the tumor was located within the bronchial interstitial connective tissue covered by the bronchial epithelium with focal erosion. The tumor was composed of sheets of oval to cuboidal cells. Abundant vascular spaces were observed in the tumor, and some were surrounded by tumor cells (Fig. 2a). Blood vessels in the tumor were small to medium-sized, thin-walled, and some were dilated, resembling capillaries or venules. The tumor cells were uniformly monotonous with a centrally placed round nucleus and amphophilic to lightly eosinophilic cytoplasm (Fig. 2b). No necrosis, vascular invasion, obvious cellular atypia, or mitotic figures were observed. Immunohistochemically, the tumor cells exhibited cytoplasmic positivity for α-smooth muscle actin, calponin, and vimentin (Fig. 2c and d). Immunostaining was negative for desmin. The individual tumor cells were surrounded by positive staining for silver impregnation and anti-type IV collagen antibody, which showed an intricate chicken-wire pattern representing basement membrane material (Fig. 2e and f). No immunoreactivity was found for cytokeratin, CD34, CD31, S-100, CD56, chromogranin A, or synaptophysin (data not shown). Ki-67 immunolabeling was detected in approximately 3% of tumor cell nuclei. Histopathologically, the tumor was diagnosed as a

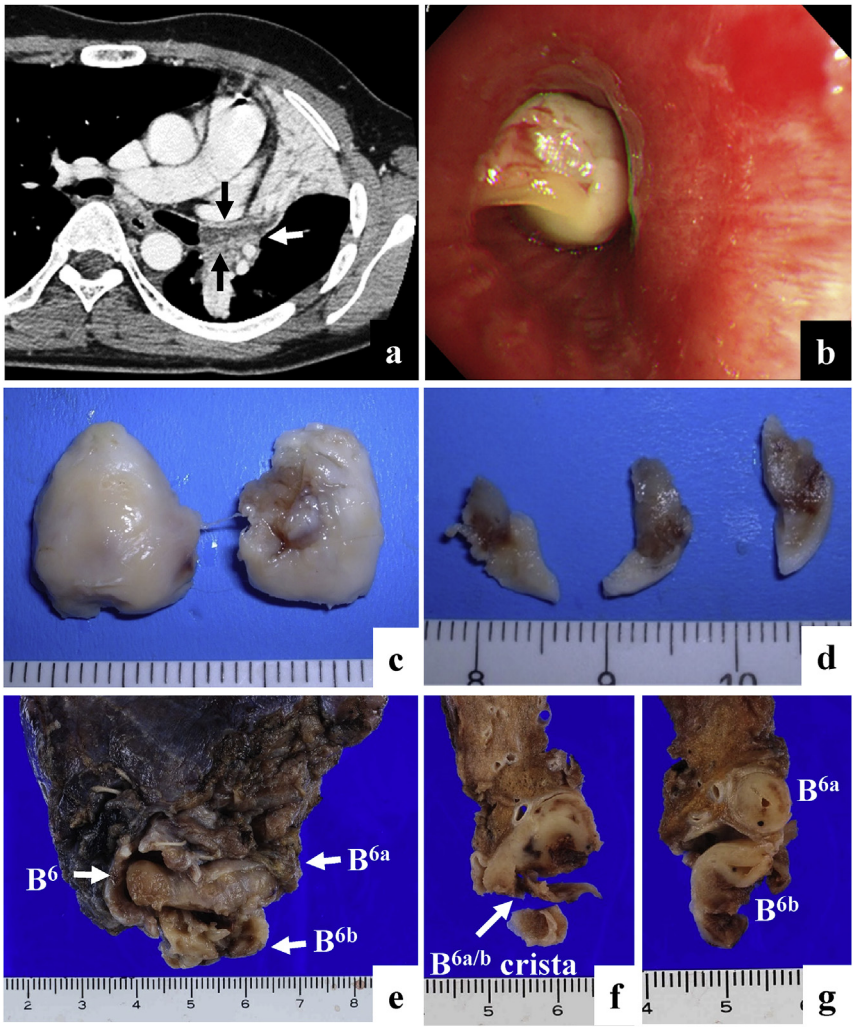

Fig. 1. Radiological, endoscopic, and macroscopic findings. (a) Contrast-enhanced chest CT reveals a water density mass in the left main bronchus (arrows). (b) Bronchoscopy shows a white polypoid mass filling the left main bronchial lumen. (c, d) The tip of the endobronchial tumor partially resected by endoscopic snaring procedure. The surface of the tumor is smooth, and the cut-surface is tan in color. (e–g) Formalin-fixed bronchopulmonary tissue obtained by left lower lobe segmental resection. A cylindrical mass arises from the superior segmental $B^{6a/b}$ crista and enlarged the lumen of B^{6}, B^{6a}, and B^{6b}.