Int J Clin Exp Pathol 2015;8(8):9723-9726
www.ijcep.com /ISSN:1936-2625/IJCEP0011682

Case Report

Glomus tumor of the trachea: a rare case report

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Received June 20, 2015; Accepted July 26, 2015; Epub August 1, 2015; Published August 15, 2015

Abstract: A tracheal glomus tumor is extremely rare. There were approximately 18 reported cases before in China. Here we report a 48-year-old male with glomus tumor of the trachea. The computed tomography (CT) scan of the chest showed a nodulein the basalsegmentof right lower lobe, focal uplift in the left-posterior wall of the trachea, measured 1.5×1.2×1.0 cm. Microscopically, the tumor tissue was rich with vessels, shaped flake-nest, and the tumor consisted of a sheet of uniform cells surrounding the vascular spaces. Immumohistochemical staining were positive for SMA, vimentin, collagen IV, CD34, the Ki-67 proliferation activity was low (<1%), and were negative for Syn, CgA, S-100, AE1/AE3 and EMA. PSA staining showing clear cell borders. With a review of the literature, the clinical, pathological characteristics and treatment modalities of this rare tumor are discussed.

Keywords: Glomus tumor, trachea, immunohistochemistry, differential diagnosis

Introduction

Glomus tumors are derived form specialized cells surrounding arteriovenous anastomosis that usually occur in the hands or feet but rarely have been described to appear in the tracheobronchial tree [1, 2]. Glomus tumor of the trachea present with symptoms including cough, dyspnea, and wheezing that may be mistaken for other pulmonary disorders [3]. So pathology is necessary to differentiate glomus tumors from other airway tumors. There were approximately 18 reported cases before in China, but all reports are more or less lack of favorable immunohistochemical markers and differential diagnosis. We herein report a typical case of glomus tumor arising from the trachea.

Case report

On November 7, 2014, a 48-year-old male smoker presented to the Third Affiliated Hospital of Soochow University suffering from cough and sputum with blood about one month. The patient was recurrent cough, sputum, phlegm within blood with dark-red, and the phlegm within blood was about 4 to 5 times a day. Physical examination showed the temperature was 37.5°C, smooth breathing, breathe sounds clear, symmetry, negative dry and wet rales of trachea. The computed tomography (CT) scan of the chest showed a nodulein the basalsegmentof right lower lobe, focal uplift in the left-posterior wall of the trachea. The lesion was about 1.5×1.2×1.0 cm. Pathological examination showed the surgical specimen consisted of a segment of trachea, 2.0 cm in length, 2.5 cm outer diameter and 2.0 cm inner diameter, with a off-red, dish-brown tumor mass, 1.5×1.2×1 cm in size, protruding into the lumen as a polypoid mass. Dissection of the specimen revealed a well-circumscribed and off-red tumor.

Microscopy

The tumor was located in the tracheal mucous membrane, no envelope and well-circumscribed. Tumor tissue was rich in vessels, shaped flake-nest. The tumor cells was uneven-thickness, composed of round to polygonal cells with uniform round to ovoid nuclei, punched-out nuclei and slightly eosinophilic to cytoplasm. The tumor cells were not obvious atypia, pathological fission, and tumor stroma was transparent or myxoid (**Figure 1A**, **1B**).

Immunohistochemistry

Immumohistochemical staining were positive for SMA (**Figure 2A**), Vimentin (**Figure 2B**),

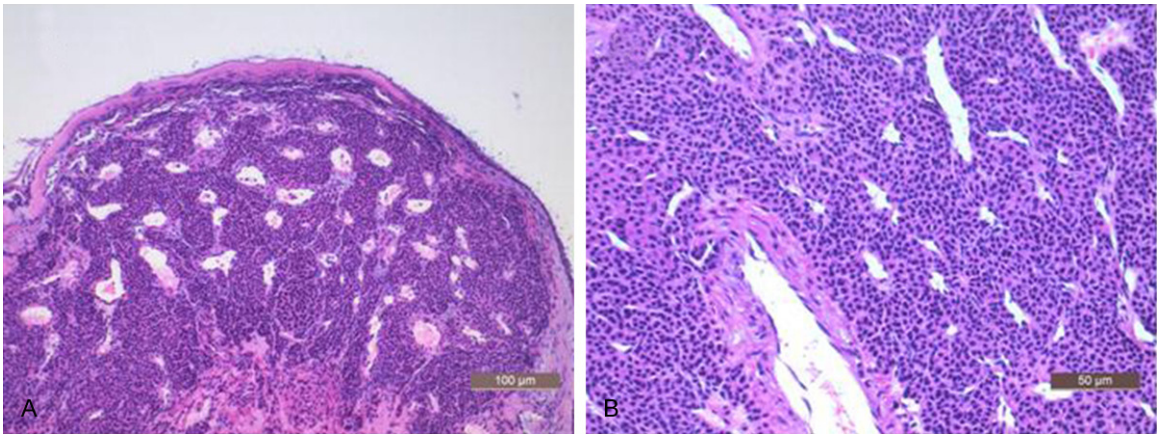

Figure 1. A: The glomus tumor showing a hypervascular tumor composed of branching, dilated, thick-walled, vascular channels and thin-walled, no envelope and well circumscribed, and tumor tissue was rich in vessels, shaped flake-nest. (magnification, ×100); B: The tumor showing lobular arrangements of oval- to-spindle-shaped cells and slightly eosinophilic to cytoplasm surrounded thin-walled blood vessels, tumor stroma was transparent or myxoid (magnification, ×200).

Collagen IV (**Figure 2C**), PSA staining showing clear cell borders (**Figure 2D**), and CD34 (**Figure 2F**), and the Ki-67 proliferation activity was low (less than 1%) (**Figure 2E**), and were negative for Syn, CgA, S-100, AE1/AE3 and EMA.

Discussion

Glomus tumor is a benign lesion that arises from the vessel in the arterial and venous anastomosis sphere [4]. The glomus body located in the anastomosis tube of Sucquct-Hoyer in the small arteriovenous anastomosis is a modified smooth muscle cells, distributed throughout the whole-body [5]. Glomus tumors are most commonly in limb, especially in the dermis or subcutis [6]. Extracutaneous presentations occur but are rare, especially in the trachea. Glomus tumor of the trachea is located in the tracheal mucous membrane, and present with symptoms including cough, dyspnea, and wheezing that may be mistaken for other pulmonary disorders, such as chronic bronchitis, bronchial asthma, and is easily to misdiagnose as carcinoid [7]. Histological features: the tumor tissues showing a hypervascular tumor composed of branching, dilated, thick-walled, vascular channels and thin-walled, no envelope and well circumscribed, and tumor tissue was rich in blood vessels, shaped flake-nest. The tumor cells is uneventhickness, composed of round to polygonal cells with uniform round to ovoid nuclei, punched-out nuclei and slightly eosinophilic cytoplasm. The tumor cells were not obvious atypia, pathological fission, and tumor stroma was transparent or myxoid and can see some mastocyte [8]. According to the different proportion of oncocyte, vascular structures and smooth muscle cells, the tumor divided into three histological patterns: (1) classic glomus tumor, 75%, well-demarcated, consists of capillary small vessels and tumor cells that grew around the vessel [9]; (2) glomangioma, 20%, poor-demarcated, most of the tumor vessels were the expansion of cavernous vessels, little and thinning of cell clusters around vessels, and can see thrombus and angiolith in the intracavitary vessel lumen [10]; (3) glomangiomyoma, less than 10%, the tumor tissue contained smooth muscle bundle besides rules-circular glomus cell, and presented transition phenomenon between glomus cell and smooth muscle cells [2]. The case was classic glomus tumor.

Differential diagnosis: (1) carcinoid, the size of tumor vessels were uniform and cells were round, oval, or slightly irregular nuclei and finely-dispersed chromatin, punched-out nuclei and eosinophilic cytoplasm, all of the features were similar to glomus tumor, while the carcinoid cells usually arranged in funicular cord, nests, and glandular structures. Immumohistochemical staining were positive for NSE, Syn and CgA, but were negative for SMA and vimenin [11]; (2) small cell carcinoma, the tumor cells usually arranged in diffuse patchy, were small cells, round or ovoid, similar to the

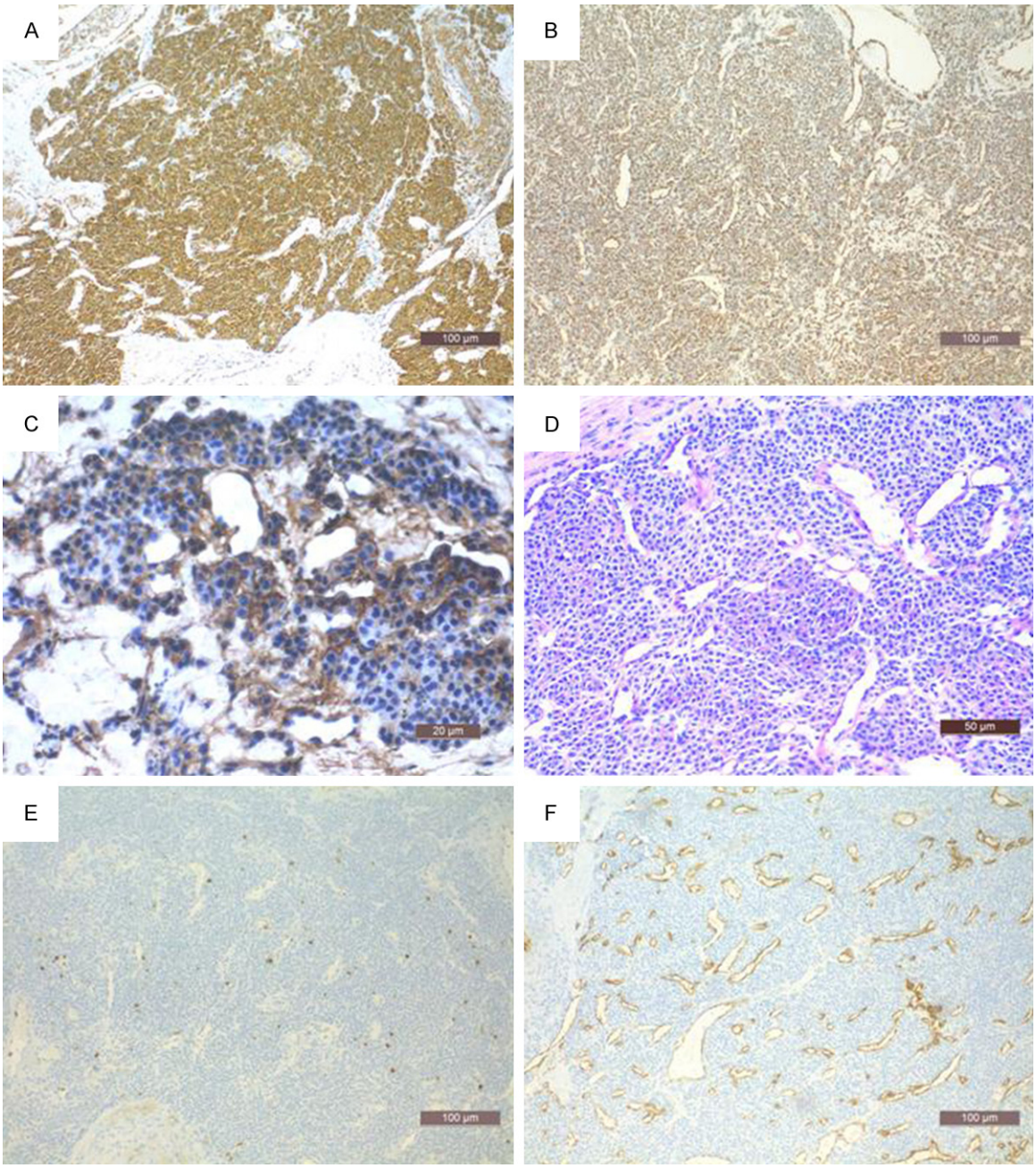

Figure 2. A: Immuno-staining showing tumor cells strongly positive SMA (magnification, ×100); B: Positive for vimentin (magnification, ×100); C: Type IV collagen was positive in basilar membrane (magnification, ×400); D: PSA staining showing clear cell borders ((magnification, ×200); E: Ki-67 proliferation activity was less than 1% (magnification, ×100); F: Positive for CD34 (magnification, ×100).

glomus tumor, but the cytoplasm of small cell carcinoma cells was little, and tumor cells were poor-demarcated, obvious allotype, pathological fission was visible, occasionally accompanied with obvious necrosis. Immunostaining of the tumor cells were positive for NSE, Syn, CgA, the Ki-67 proliferation activity was usually greater than 50% [12]; (3) poorly differentiated adenocarcinoma, the tumor cells arranged in diffuse patchy comparable to glomus tumor, but the tumor cells were obvious allotype, pathological fission was visible, usually accompanied with obvious necrosis. Immumohistochemical staining was positive for AE1/AE3 and EMA; (4) hemangiopericytoma, the tumor had the characteristic of the antler-like branch

vessels, and the complete basilar membrane wrapped the vessels, the tumor cells were intensive arrangement outside the basilar membrane, showed oval, shortshuttle-like, poor-demarcated. A mass of argyrophilicfibrils-distributed around the blood vessels and tumor cells. Immumohistochemical staining was positive for CD34 and Vimentin [13]; (5) paraganglioma, tumor epithelial cells contained master cell arranged in organoid or heter-cell that was oval or polygon and spindle sustentacular cell around the master cell. Some of the tumor had the acinar or adenoid structure, the cytoplasm of master cells was eosinophilic, slightly granular, usually had large vesicular nuclei. Immumohistochemical staining was positive for NSE, Syn, CgA, S-100 (spindle sustentacular cells) [14].

Glomus tumor of the trachea was usually a benign tumor, most of the treatment used local excision. The tumor had a favorable prognosis; the case has no recurrence and metastasis so far.

Acknowledgements

This research project was supported by National Natural Science Foundation of China (No. 81171653, 30972703).

Disclosure of conflict of interest

None.

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