The mimic of tracheal carcinoid in elderly female

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ABSTRACT

We present a case of a glomus tumor of trachea in an elderly female who presented with a mass originating from the posterior trachea. She underwent rigid bronchoscopy with tumor debulking combined with laser therapy. Frozen section initially suggested carcinoid tumor but later turned out to be a glomus tumor. She improved with additional laser therapy. We present her clinical course and a literature review on glomus tumor.

KEY WORDS: Carcinoid tumor, glomus tumor, tracheal tumor

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INTRODUCTION

Glomus tumors are rare benign neoplasms, first described in 1924, accounting for <2% of soft tissue tumors.^[1] They arise from the glomus body which is a cutaneous structure that regulates the temperature through arteriovenous shunting of blood. The common locations of glomus tumors are the subungual regions of digits or the deep dermis of the palm, wrist, and forearm.^[2] The trachea is a very uncommon site of this tumor.^[3] Less than fifty cases of tracheal glomus tumors have been reported.^[4]

CASE REPORT

A 79-year-old female presented to the emergency department for chest pain and shortness of breath. She underwent a computer tomography (CT) of the chest which showed a mass $(1.7 \text{ cm} \times 1.4 \text{ cm})$ arising from the posterior wall of trachea [Figure 1]. The mass had narrowed the tracheal lumen to about 50%. She underwent a flexible bronchoscopy, which showed a hypervascular endobronchial mass with a wide base

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arising from the posterior wall of the trachea [Figure 2]. Endobronchial biopsies were not performed because of hypervascularity. Instead, rigid bronchoscopy and yttrium-aluminum-garnet (YAG) laser were done for therapeutic and diagnostic purposes. Clear margins were not obtained because of the tumor extension into the posterior membranous trachea.

The intraoperative frozen section of the mass was suggestive of carcinoid tumor. On permanent histologic sections, the tumor consisted of groups of uniform, round cells with oval nuclei and pale eosinophilic cytoplasm surrounding dilated vessels without mitosis or necrosis. The immunohistochemical staining revealed that the tumor cells were positive for smooth muscle actin HHF-35 and negative for CD31, cytokeratins, and S100. The final pathological diagnosis was glomus tumor of tracheal origin [Figure 3].

The patient's symptoms improved postprocedure. The patient refused tracheal resection and opted instead to undergo repeated bronchoscopies with YAG laser, which she tolerated well.

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Figure 1: Computer tomography chest showing mass arising from posterior wall of trachea



Figure 2: Bronchoscopic view of mass arising from posterior wall of trachea



Figure 3: Tracheal glomus tumor histopathology. (a) The tumor consists of groups of uniform, round cells with oval nuclei and pale eosinophilic cytoplasm surrounding dilated vessels. Mitotic activity is absent (H and E, ×340). (b) The tumor cells are positive for smooth muscle actin. (HHF-35 immunohistochemical staining, ×340)

DISCUSSION

Tracheal glomus tumors arise from the posterior wall of the lower two-third of trachea which is rich in vasculature and mucus glands. Tumor is more common in males with a mean age at presentation being 51 years.^[4] The tumor size ranges from 1.2 to 4.6 cm.^[5] The most common presentation includes dyspnea, cough, hemoptysis and/or stridor.^[5,6] Tracheal glomus tumors are mostly benign; however, few have local invasion and malignant potential.^[7]

Chest CT is the modality of choice. However, the ultimate diagnosis depends on tissue sampling, histology, and immunohistochemistry. Glomus tumors typically have three components: glomus cells, vasculature, and smooth muscle cells. The characteristic stains include positivity for smooth muscle actin and negativity for CD31, cytokeratin stain, and S100.^[8] The results of immune histochemical staining in this report are consistent with this pattern. The differential diagnoses of tracheal glomus tumor include carcinoid tumors, small cell carcinoma, and paraganglioma. Carcinoid tumor has a less prominent vascular pattern, stains positively for neuroendocrine markers and cytokeratin, and does not express smooth muscle actin. Small cell carcinoma cells have high nuclear to cytoplasmic ratio, fine chromatin, indistinct nucleoli, nuclear moldings, and DNA streaking is often present. The tumor cells are positive for the above neuroendocrine markers, mitotic figure of usually above 90%. Paraganglioma has a unique zellballen or trabecular pattern of cells within a prominent vascular network. The tumor cells are negative for smooth muscle actin.^[9-12]

The treatment of choice is sleeve tracheal resection with tracheal end-to-end anastomosis. Bronchoscopic treatment modalities including rigid bronchoscopy with tumor debulking in combination with YAG laser has been reported in patients who are not surgical candidates.^[3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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