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Glomus tumor of trachea in an adult male

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CASE REPORT

Glomus Tumor of Trachea in an Adult Male

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ABSTRACT

Glomus tumors most commonly occur in distal extremities, particularly in the subungual region. The origin of these tumors is modified smooth muscle cells of glomus bodies. Trachea is one of the rare sites where normal glomus bodies may be sparse or absent, so occurrence of glomus tumor in trachea is very rare. Only 30 cases have been reported in the literature so far. In this case report, the authors describe a very rare and in fact first case of glomus tumor of trachea in a 45-year male Pakistani patient.

Key Words: *Glomus tumor. Trachea. Dyspnea.*

INTRODUCTION

Glomus tumors are benign neoplasms. They arise from the modified smooth muscle cells of the glomus body, which is a specialized arteriovenous anastomosis involved in thermoregulation. Glomus tumors occur most commonly in the dermis and subcutaneous tissue. They uncommonly occur in visceral organs making extra cutaneous presentation a rare finding. Normal glomus bodies may be sparse or even absent in the trachea. Only 30 cases of tracheal glomus tumors have been reported so far.[1,2]

We report a tracheal glomus tumor in an adult which was surgically resected.

CASE REPORT

A 45-year man, a known diabetic, presented with a 4-year history of progressively increasing dyspnea. On examination, inspiratory stridor was observed. CT scan showed a polypoidal mass protruding into the tracheal lumen approximately 1 - 2 cms below the vocal cord (Figure 1A). He underwent surgery and the tumor was resected along with the second and third tracheal rings. The patient's symptoms were relieved and he was discharged after surgery without any complications.

The specimen contained 2 tracheal rings measuring 4 x 3 cm. The inner surface showed a polypoidal growth, protruding into the lumen measuring 2.5 x 1.5 cm. Cut surface of the growth was greyish white, it was firm in consistency with few myxoidy areas (Figure 1B). Microscopically, the tumor was composed of multiple, branching vascular channels surrounded by nests of cells. These cells were monotonous, round to oval in shape, had moderate eosinophilic cytoplasm, rounded hyperchromatic nuclei and indistinct nucleoli (Figure 2A-C). There was no evidence of nuclear pleomorphism, increased mitotic activity or necrosis. On immunohistochemistry, tumor cells showed positivity for immunohistochemical stain anti-smooth muscle actin (ASMA, Figure 2D) and were negative for CD31, CKAE1/AE3, and chromogranin. Based on morphology and immunoprofile, a diagnosis of glomus tumor was rendered. All surgical margins were negative.

Follow-up bronchoscopy was performed 6 months later. After 20 months of follow-up, he has had no complications.

DISCUSSION

Glomus tumors are uncommon benign soft tissue tumors with an incidence of 1 - 6%.[3] Most glomus tumors are diagnosed in young adults (20 - 40 years of age). No gender predilection has been reported. According to the World Health Organization classification of soft tissue tumors, it can be divided into 3 broad groups on the basis of their biologic potential: glomus tumors, glomus tumors of uncertain malignant potential, and malignant glomus tumors.[4] The benign form accounts for more than 95% of the cases.[5]

The single-most common site is the subungual region of the finger; other common sites include palm, wrist, forearm and foot. Thus glomus tumors mainly occur in sites where glomus bodies are usually present. However, these tumors may also uncommonly develop in sites where glomus bodies are sparse or even absent. Such unusual locations include bone, gastrointestinal tract, ovary, cervix, mediastinum and respiratory tract.[6]

Classic glomus tumors are typically solitary. Characteristically, these lesions are up to 1 cm in size and complete excision is the treatment of choice. Trachea is an extremely rare site for glomus tumors. Only 30 cases of tracheal glomus tumors have been reported.[1,2] A tumor in the trachea usually protrudes into the lumen like a polypoid mass, and causes partial obstruction resulting in progressive dyspnea, so intervention is unavoidable.

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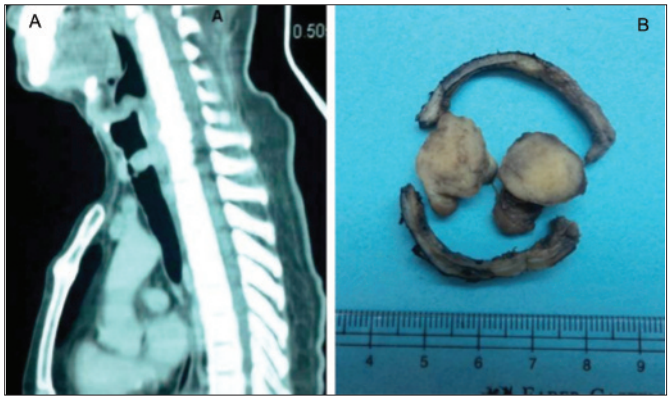

Figure 1: (A) CT scan sagittal image shows a polypoid lesion in the lumen of trachea arising from the posterior wall. **(B)** Gross appearance of the tumor as polypoid, pale firm cut surface. The 2 tracheal rings removed are also seen.

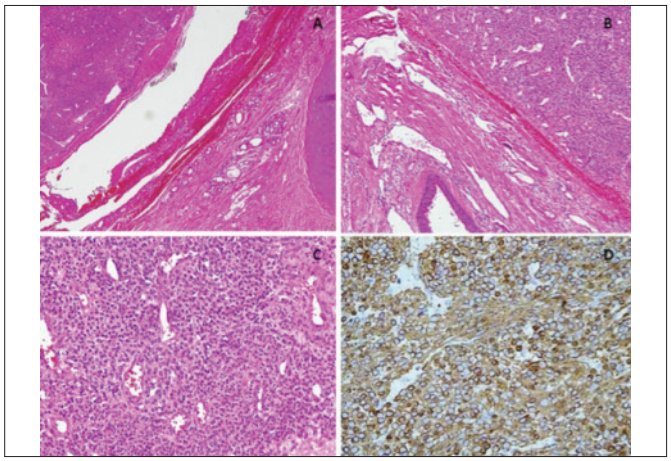

Figure 2: (A) Tumor protruding into lumen with tracheal cartilage in wall (H & E, 40x magnification). **(B)** Tumor in the tracheal wall showing overlying pseudostratified ciliated columnar epithelium (H & E, 40x magnification). **(C)** Monotonous proliferation of oval cells with clear cytoplasm around dilated vessels (H & E, 100x magnification). **(D)** Diffuse positivity of tumor cells for ASMA immuno-histochemical stain (100x magnification).

According to previous reports of tracheal glomus tumors, the most common symptoms are shortness of breath, cough, stridor due to obstruction, and above all, hemoptysis, which is the result of the rich vascularity of these tumors.

The size of the reported cases ranged from 1.2 to 4.5 cm and most were located in the posterior wall.[7] Histologically, glomus tumors are composed of rounded cells exhibiting punched out nuclei, pale cytoplasm and a lacework of basement membrane-like material. These cells are closely spaced and often interdigitate with each other along their short knobby processes. Rarely these cells may show oncocytic change.[3] Glomus tumors of trachea can histologically mimic carcinoid tumors and hemangiopericytomas. Like glomus tumors, tracheal carcinoids are also rare. They are composed of small monomorphic cells arranged in nests, trabeculae, and ribbons. However, the nuclei of tracheal carcinoids have the characteristic stippled chromatin of neuroendocrine tumors. They do not show the prominent vascular channels seen in a glomus tumor. Other differentiating features include immunohistochemical positivity for neuroendocrine markers, such as chromogranin and synaptophysin, and the presence of dense-core neurosecretory granules on electron microscopy.[8] Like glomus tumors, hemangiopericytomas are also composed of tumor cells that surround branching capillaries and vascular channels. However, the vessels within hemangiopericytomas are usually narrow and exhibit a characteristic staghorn pattern. In contrast to glomus tumors, cells of hemangiopericytoma are more spindly and do not demonstrate smooth muscle differentiation.[9] Other histological differentials include carcinoma and hemangioma, which can be excluded by immunohistochemical stains CKAE1/AE3 and CD31, respectively.

Almost all glomus tumors reported in the trachea behaved in a benign manner, although recurrences seen in cases where tumor resection was incomplete.[10] Complete surgical removal of the tumor with primary reconstruction of trachea is considered as the first choice for treatment for tracheal glomus tumors. The long-term prognosis is very good and the chances for recurrence are very low, if surgical margins are negative.

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