Case Study

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Malignant glomus tumor of trachea: a case report with literature review

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Abstract

Glomus tumors of the trachea are extremely rare and generally benign, with very few cases of malignant transformation reported in literature. We present the case of a 74-year-old man explored for cough and dyspnea. Bronchoscopy showed a polypoid mass arising from the posterolateral tracheal wall. Computed tomography demonstrated a mid-tracheal tissular mass obliterating the tracheal lumen. Resection and anastomosis of the trachea were performed. The histological and immunohistochemical findings were consistent with malignant glomus tumor.

Keywords

Glomus tumor, Trachea, Tracheal neoplasms

Introduction

Glomus tumors are rare benign neoplasms that arise from glomus bodies, specialized smooth muscle cells located around arteriovenous anastomoses to regulate local vascular flow. Glomus tumors in the respiratory system are extremely rare and predominantly benign. Very few cases of malignant transformation have been reported. Herein we present an exceptional case of malignant tracheal glomus tumor.

Case report

A 74-year-old man was admitted with cough and progressive dyspnea for 3 months. On physical examination, he had an inspiratory wheeze. Bronchoscopy showed a polypoid mass arising from the posterolateral tracheal wall 4 cm below the vocal cord, occupying 80% of the tracheal lumen. Computed tomography demonstrated a mid-tracheal tissular mass of 29×23 mm, almost totally obliterating the tracheal lumen (Figure 1). A diagnosis of tracheal carcinoid was discussed. Surgery was performed via a median sternotomy and consisted of resection and anastomosis of the trachea. The surgical specimen was a tracheal segment comprising a sessile solid polypoid endoluminal tumor of 25 mm in diameter and blue-red in appearance (Figure 2). Histologically, the tumor occupying the submucosa of the trachea and was composed of round cells with eosinophilic cytoplasm and round nuclei with fine chromatin (Figure 3). The neoplastic cells exhibited moderate nuclear atypia and increased mitotic activity (4 mitoses per 10 high-power fields). These cells were arranged in trabeculae and sheets interspersed with vessels of various sizes. There was no necrosis or atypical mitotic figures. On immunohistochemistry, the tumor cells express vimentin, smooth muscle actin, and caldesmon, but they were negative for cytokeratin, desmin, CD31, CD34, PS100, and CD56. These findings were consistent with a glomus tumor. The high mitotic index, deep location, and the size of the lesion indicated that the tumor was malignant, warranting surveillance because of a metastatic risk. Postoperative bronchoscopy revealed a well-healed anastomosis. The postoperative course was uneventful, and the patient was asymptomatic at 30 months after the operation.

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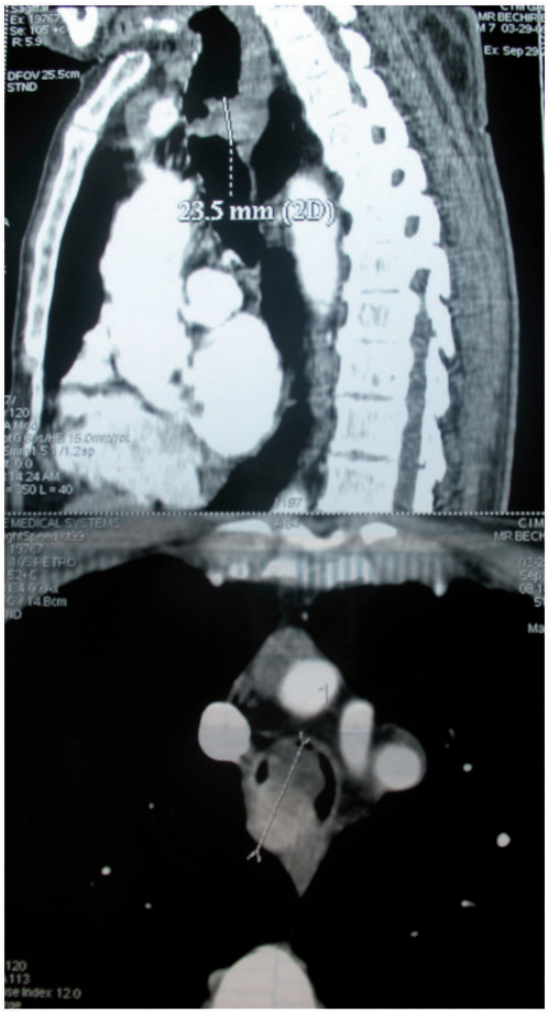

Figure 1. Computed tomography of the chest showing a tumor arising from the posterior wall of the trachea.

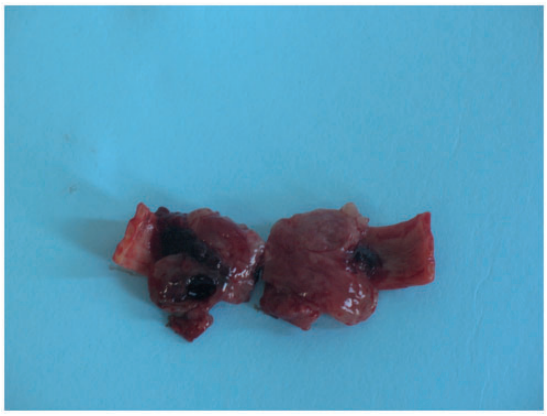

Figure 2. The resected tracheal specimen showing a protruding polypoid mass arising from the wall of the trachea.

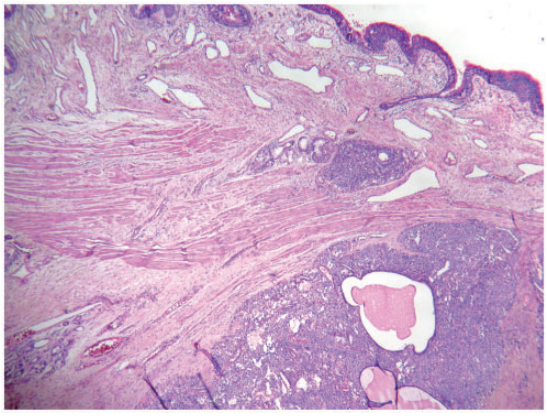

Figure 3. The tumor that occupied the submucosa of the trachea. Hematoxylin and eosin stain, original magnification × 100.

Discussion

Glomus tumors were first described in 1924. They are uncommon neoplasms, representing less than 2% of all soft-tissue tumors.[1] They can be solitary or multicentric, and are most commonly found in the subungual area. Unusual sites include muscle, tendons, ligaments, periosteum, and visceral organs such as the stomach, vagina, penis, mediastinum, and heart.[2–4] Glomus tumors of the trachea are extremely rare and normal glomus bodies may be few or even absent.[1] The first case was reported in 1950. To our knowledge, 40 cases of tracheal glomus tumor have been described so far, most commonly arising from the posterior wall of the lower two-thirds of the trachea where mucus glands and vessels are numerous.[2,3,5–7] Glomus tumor of the trachea is more common in males than females. The mean age of the patients was found to be 51 years with a range of 10–83 years. Common presenting symptoms include hemoptysis, dyspnea, and cough. Less frequently, chest pain, stridor, and hoarseness have been described. The tumor usually protrudes into the lumen of the trachea, like a polypoid mass, causing partial or complete obstruction.[4] Tumor sizes range from 1.2 to 4.5 cm. The vast majority of cases were surgically treated, and some underwent endoscopic resection, with no evidence of recurrence at follow-up ranging from one month to 6 years.[2–7] Histologically, glomus tumor cells are medium sized with round regular nuclei and eosinophilic cytoplasm, and are arranged in sheets that form collars around capillary-sized vessels. Immunohistochemically, they stain for smooth-muscle actin and vimentin, but not for cytokeratin, chromogranin, or neuroendocrine markers.[1]

Glomus tumors are most often benign, although atypical and malignant features have been described in a few cases, with only one in the trachea.[6] The World Health Organization's classification divides glomus tumors into benign glomus tumors, glomus tumors of uncertain malignant potential, and

malignant glomus tumors. The histologic criteria of malignancy are deep location, size 2 cm, atypical mitotic figures or a combination of moderate to high nuclear grade and 5 mitotic figures per 50 high-power fields.[8] Our case is the second reported case of malignant glomus tumor originating in the trachea, based on a high mitotic index, deep location, and size. The main differential diagnoses of tracheal glomus tumor are carcinoid tumor and hemangiopericytoma. Carcinoid tumor has a less prominent vascular pattern, stains positively for neuroendocrine markers and cytokeratin, and does not express smooth-muscle actin. A hemangiopericytoma consists of spindle-shaped cells with elongated nuclei, the vascular channels display a ''staghorn-like'' pattern, and the cells stain positive for vimentin with or without CD34. Smooth-muscle differentiation is infrequent.[2–4,7] Sleeve resection with primary reconstruction of the trachea is the treatment of choice for a tracheal glomus tumor. Complete surgical resection is curative, does not require adjuvant treatment, and has an excellent prognosis. Endoscopic intervention alone may be indicated if the lesion is strictly confined to the airway lumen without extension into the airway wall.[2–4,7]

Declaration of conflicting interests

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