

Glomus tumor of the proximal trachea: A case report

Proksimal trakea yerleşimli glomus tümörü: Olgu sunumu

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Glomus tumors are uncommon benign neoplasms, which rarely occur in the trachea, usually originating from the posterior wall of the distal portion. Most of tracheal glomus tumors are located in the lower two thirds of the trachea. In this article, we present a 50-year-old male patient with a glomus tumor located on the upper third of the posterior wall of the trachea. A transoral endoscopic laser excision was performed. No finding suggesting tumoral regrowth at 36 months following surgery was found, as assessed by laryngoscopy and imaging studies.

Key Words: Endoscopic; glomus tumor; laser excision; trachea.

Glomus tümörleri, trakeada nadiren görülen ve genellikle distal kısmın arka duvarından köken alan sık görülmeyen benign tümörlerdir. Trakea yerleşimli glomus tümörlerinin birçoğu, trakeanın alt kısmının üçte ikisini tutar. Bu makalede trakeanın arka duvarının üstten üçüncü kısmında yer alan glomus tümörlü 50 yaşında bir erkek hasta sunuldu. Hastaya transoral endoskopik lazer eksizyon yapıldı. Laringoskopi ve görüntüleme çalışmaları ile değerlendirildiği üzere, ameliyattan sonra 36. ayda tümörün yeniden büyüdüğüne işaret eden herhangi bir bulguya rastlanmadı.

Anahtar Sözcükler: Endoskopik; glomus tümörü; lazer eksizyon; trakea.

Glomus tumors originate from the modified smooth muscle cells of the glomus body. With rare exceptions the tumors are benign and most commonly arise in the deep dermis and adjacent subcutaneous tissue, particularly in the subungual region of the fingers. Other common locations where glomus tumors originate include the palm, wrist, forearm, and foot. They develop in extracutaneous locations rarely, including the trachea.^[1-15] Masson first described this tumor in

the tactile regions in 1924.^[2] Since then varying types and locations of glomus tumors have been reported in the literature. Four distinctive subtypes of glomus tumor have been classified based on ultrastructural differences and on the relative proportions of glomus cells, vascular structures, and smooth muscle tissue in the tumor. Among these subtypes: 75% are classic glomus tumors, 20% are glomangiomas and rare examples of oncocyctic glomus tumors have been reported.^[12]

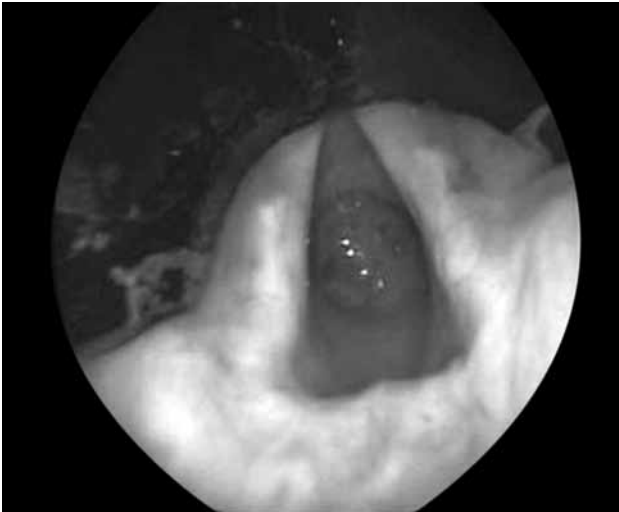


Figure 1. A mass which originated from the posterior wall of the trachea was observed with endoscopic examination.

Histopathologically, glomus tumors have fine reticulin fibers that surround individual small round cells. The cells react immunohistochemically with actin and desmin. Smooth muscle differentiation of basal lamina envelope, pinocytotic vesicles along the plasma membrane, and intracytoplasmic bundles of myofibrils with electron-dense plaques are seen by the electron microscopy.^[3-7] The trachea is an unusual site for both benign and malignant neoplasms and glomus tumor is very rarely seen in the trachea.

CASE REPORT

A 50-year-old male patient with longstanding chronic obstructive pulmonary disease and

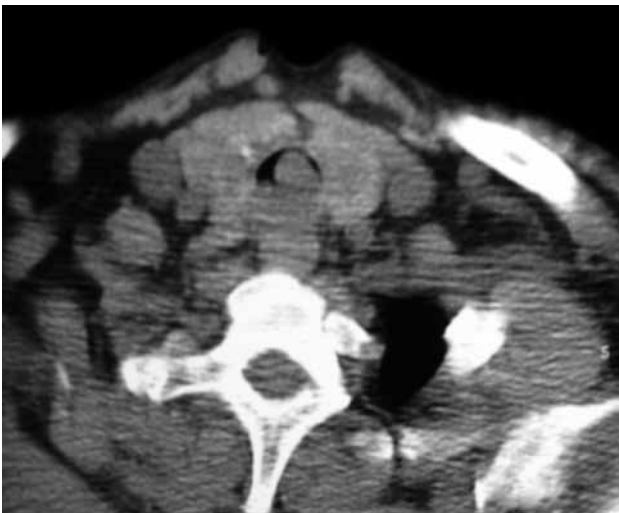


Figure 2. Preoperative computed tomography scan section.

hypertension was admitted to our clinic with severe dyspnea and a tracheostomy was performed in the emergency room. He had used bronchodilator drugs for two years. He was a ten pack-year smoker. He showed symptoms of cough, brief hemoptysis, dyspnea, and stridor. On ear nose and throat (ENT) physical examination there was no abnormality observed. A mass which originated from the posterior wall of the trachea was observed with endoscopic examination (Figure 1). A computed tomography scan showed a polypoid mass arising from the posterior wall of the upper third of the trachea (Figure 2). No lymphadenopathy was detected. A transoral endoscopic laser excision was performed. During the operation, the mass which originated from the posterior wall of trachea (level of first and second rings) was observed. The mass had a smooth surface and almost the whole tracheal lumen was obliterated. Intraoperative frozen section revealed a submucosal mesenchymal tumor. The histopathologic sections of specimen revealed a tumor composed of small round cells and sheets surrounding thick-walled enlarged capillaries that were deeply seated beneath the mucosa. The overlying mucosa was intact (Figure 3). No necrosis or mitotic activity was noticed. Tumor cells showed positive staining with smooth muscle actin. Keratin and neuroendocrine markers were negative. Histopathologic diagnosis was glomus tumor. There was no evidence of recurrent disease in a three-year follow-up period (Figure 4, 5).

DISCUSSION

Glomus tumor was first described by Masson in 1924.^[2] The tumor is a distinctive type of vascular

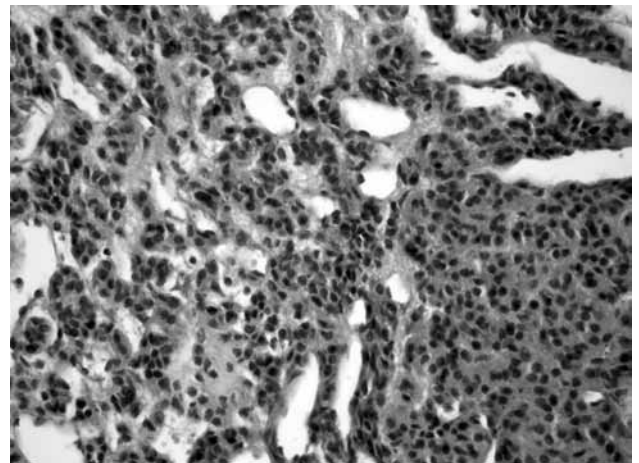


Figure 3. Polygonal and round cells with uniform, hyperchromatic nuclei forming microvascular spaces were seen in histopathologic sections (H-E x 20).

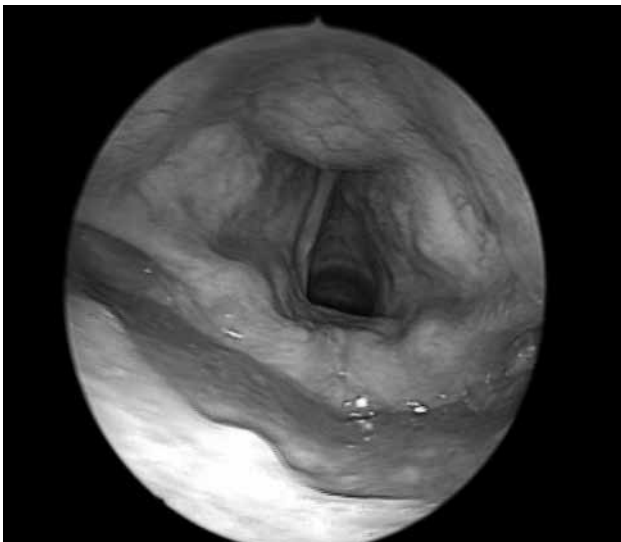


Figure 4. The end of three-year follow-up period endoscopic examination.

tumor and the cell type is a modified smooth muscle cell closely resembling the glomus body from which the name is derived.^[1-15] Glomus tumors are uncommon benign neoplasms which are commonly seen in the extremities particularly in the subungual region of the finger. The trachea is an unusual site for benign and malignant neoplasms and glomus tumors of the trachea are distinctly rare; only 29 reports have been published in the literature. The patients, including ours, ranged in age from 43 to 69 years and $2/3$ were males and $1/3$ were females.^[1-15]

All tumors reported in the literature arose from the posterior membranous wall of the trachea and most were in the lower third of the trachea.^[1-15] The current tumor was located on the upper third part of the trachea. Most of the published cases had symptoms related to obstruction of the tracheal lumen such as dyspnea, cough, and hemoptysis. Dyspnea with hemoptysis were the main complaints of this patient on admission to the hospital. Surgical resection of the tumor is definitive treatment.^[8-10] Segmental or sleeve tracheal resection or total tumor excision by different approaches such as rigid bronchoscopy, bronchofiberscopy, transoral laser endoscopy may be an option for patients who prefer this over surgical resection. In case of tumors that extend outside the trachea, tracheal resection be performed.^[9,11-13]

A transoral endoscopic laser excision was performed in our case. The main histologic



Figure 5. The end of three-year follow-up period computed tomography scan section.

differential diagnoses of glomus tumors are carcinoid tumor and hemangiopericytomas.^[8,11] These tumors also arise from the submucosa of the trachea and consist of sheets and nests of cells surrounding the numerous vascular spaces.^[12] The thick-walled blood vessels, the strong staining for smooth muscle actin and type 4 collagen, and the lack of staining for chromogranin and cytokeratin support the diagnosis of glomus tumor.^[16] Small and relatively uniform round cells surrounding the vessels with immunohistochemical expression (neither epithelial nor neuroendocrine markers) supported the histopathologic diagnosis in the current case. In conclusion, although the trachea is an unexpected location for the glomus tumor, it should still be kept in mind for correct intervention for those patients, even if they are located in the upper part of the trachea.

Declaration of conflicting interests

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