Laryngeal and Cervical Glomangiopericytomas: First Case Report

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ABSTRACT

Objectives: 1) To describe the first reported laryngeal, retropharyngeal, and cervical glomangiopericytomas. 2) To review the presentation, histopathology, immunohistochemistry, and treatment of glomangiopericytoma.

Methods: We present a case report and brief literature review of head and neck glomangiopericytoma.

Results: A 64-year-old female presented for evaluation of glottic and retropharyngeal masses found incidentally on CT angiography during a syncopal workup. Flexible laryngoscopy demonstrated submucosal fullness of the right false vocal fold with intact true vocal fold mobility and effacement of the ipsilateral pyriform sinus. MRI with contrast revealed contrast enhancing lesions that were T1 hypo-to-isointense and T2 hyperintense. After direct laryngoscopy and biopsy, pathology showed a neoplasm with spindle to small round cells with numerous branching thin-walled vessels. No nuclear pleomorphism, increased mitotic activity or necrosis was noted. Immunohistochemistry was positive for smooth muscle actin (SMA) and desmin, and negative for CD34, pan-cytokeratin (PCK), S100, and CD99. Two independent pathologists confirmed the diagnosis of glomangiopericytoma. The patient subsequently underwent transcervical surgical excision of the laryngeal and retropharyngeal masses. Intraoperatively, an additional level 3 mass was noted. Pathology confirmed that there were in fact four separate glomangiopericytomas (laryngeal, two retropharyngeal, and right level 3 mass). PubMed review revealed no reports of laryngeal, retropharyngeal, or cervical glomangiopericytomas.

Conclusion: Glomangiopericytoma is a rare tumor previously described in the sinonasal cavity but not the neck. This tumor is characterized by a perivascular myoid phenotype, and immunohistochemical stains are used to support the diagnosis. Glomangiopericytoma has a high propensity for local recurrence and requires definitive surgical resection.

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INTRODUCTION

Glomangiopericytoma is a rare vascular soft tissue tumor within the non-specific category of hemangiopericytoma-like tumors. Previously, these tumors were named sinonasal-type hemangiopericytomas, but the World Health Organization transitioned to glomangiopericytoma in 2005 to reflect the similarity to glomus tumors. These tumors are classified as borderline or low-malignancy tumors and predominantly appear in the sinonasal region, representing 0.5% of all sinus tumors.2 We present a patient with multifocal laryngeal, cervical, and retropharyngeal glomangiopericytomas. While there are less than a dozen reported laryngeal hemangiopericytomas,2 this is the first reported case of a cervical glomangiopericytoma and one of the first non-sinosal glomangiopericytomas described in the literature. Otolaryngologists and pathologists should be aware of the possible extrasinonasal location of glomangiopericytomas.

CASE REPORT

A 64-year-old female was evaluated for laryngeal and retropharyngeal masses found incidentally on CT angiography during an evaluation for syncope. She had no hemoptysis, voice or swallowing complaints, and no history of prior alcohol or tobacco use. Flexible laryngoscopy demonstrated a submucosal fullness of the right false vocal fold with effacement of the ipsilateral pyriform sinus. Vocal fold mobility was intact. CT angiography demonstrated a 3.7 x 2.6-cm right glottic mass and a 2.4 x 1.6-cm right retropharyngeal mass.

Pathology revealed a vascular neoplasm with numerous thin-walled vessels, some branching, and perivascular hyalinization. There were also spindle-shaped cells and glomus-like cells. No nuclear pleomorphism, increased mitotic activity or necrosis was seen. Prior to excision, an angiogram demonstrated complex moderately vascular right neck masses supplied by terminal branches of the superior thyroid artery. Embolization was performed, and the patient subsequently underwent definitive transcervical surgical excision. Final pathology provided the diagnosis of glomangiopericytoma, which was confirmed by an outside expert head and neck pathologist. Pathology confirmed that there were in fact four separate glomangiopericytomas and one benign lymph node.

DISCUSSION

While cervical glomangiopericytomas have not been previously described, glomangiopericytomas of the sinonasal cavity have been described fairly extensively. For tumors arising in the sinonasal cavity, presentation is usually with unilateral nasal obstruction and epistaxis, and a red polypoid mass on exam.

REFERENCES