**Hypopharyngeal Paraganglioma: A Case Report and Review of the Literature**

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**INTRODUCTION**

Paraganglioma are rare, highly vascular neoplasms that are most commonly occur in the head and neck and account for only 0.012% of all neoplasms. They arise from the paraganglia of neural crest origin and usually develop on the carotid body, jugulotympanic or vagal paraganglia. Rare cases of laryngeal paragangliomas have been reported. This paper presents a case of a hypopharyngeal paraganglioma and a review of the literature concerning laryngopharyngeal paragangliomas.

**CASE REPORT**

A 39 year old female complained of progressively increasing odynophagia, hoarseness, dysphagia and neck pain for two months. A right, mucosalized, supraglottic mass was noted emanating from the right pyriform sinus. Computed tomography (CT) scan of the neck with contrast demonstrated a 2 cm briskly enhancing mass in the right pyriform sinus extending to the right aryepiglottic fold. (Figure 1) Laterally, the mass bulged into the right thyrohyoid membrane. Magnetic resonance imaging of the neck with gadolinium confirmed the mass, bright on T2 weighted images, compressing the true vocal fold laterally and posteriorly. Following development of stridor, the patient underwent a tracheostomy and biopsy of the mass at an outside institution. Initial pathology was suggestive of a paraganglioma. Family history of paragangliomas was negative. An attempt was made at excision using a combination of endoscopic and external approaches, however the surgery was aborted due to excessive bleeding. She subsequently underwent arteriography and embolization and her care was transferred to the senior author.

Labs, including serology and 24 hour urine vanillylmandelic acid (VMA), metanephrine and normetanephrine were within normal limits. The patient underwent a right lateral thyrosectomy with excision of the mass. The supralateral one third of the right supralateral thyroid cartilage was excised along with a small portion of the pyriform sinus mucosa, violated during the previous procedure. The mucosal defect was closed primarily. Intraoperatively, the neoplasm originated from within the pyriform sinus without involvement of the false vocal folds. The specimen consisted of a 3.5 x 2.5 x 1.5 cm irregular, tan-pink mucosal lesion with an attached horn of thyroid cartilage. Pathologic examination confirmed the diagnosis of a paraganglioma with positive immunostains for synaptophysin, chromogranin and S100. Light microscopy demonstrated clusters of chief cells surrounded by sustentacular cells (Figure 2). The Ki-67 proliferative index was 5-10%. The patient was discharged on post-operative day #5 following successful decannulation and return to a regular diet.

**DISCUSSION**

Laryngopharyngeal tumors of neural-crest origin are unusual, distinct tumors that fall into two broad categories: epithelial and neural. Epithelial tumors are further subdivided into the 2005 WHO classification (see table 1: typical and atypical carcinoids, and small cell neuroendocrine tumors). The only tumors that are identified as “neural” are paragangliomas.

The histopathologic identification of these tumors is crucial for guiding treatment, both primary and adjuvant, since their biologic aggressiveness can vary widely. For epithelial neuroendocrine tumors, survival and histopathologic differentiation have a linear relationship: survival decreases as the tumors become less differentiated. Laryngopharyngeal paragangliomas, however, are consistently benign. The majority of malignant ones can be attributed to misclassification of atypical carcinoids and other tumors. Overall, the documented rate of malignant paragangliomas is 2%.

Grossly, paragangliomas are subepithelial, highly vascular, blue or red lesions without ulceration of the overlying mucosa. Two cell types are histologically noted: chief cells (“zellballen”) and sustentacular cells. The identification of a laryngopharyngeal neoplasm as neuroendocrine in origin requires a combination of light microscopy and either the use of particular immunohistochemical stains or electron microscopy. Utilizing specific antibody stains further subdivides the tumor into the neural or epithelial subgroups (Table 1). The presence of neuroendocrine markers helps differentiate from the non-neural crest origin tumors, while the absence of the epithelial and calcitonin staining segregates the paragangliomas from the laryngeal carcinoids and small-cell carcinomas. In our case, the neoplasm stained positive for S-100 in the sustentacular cells, chromogranin and synaptophysin thereby excluding all other diagnoses.

Laryngopharyngeal paragangliomas are rare, highly vascular and predominately benign neoplasms. The proper histopathologic identification of these tumors is tantamount to guiding treatment, as they can easily be confused with more aggressive neoplasms. The preferred operative approach is a lateral thyrosectomy to minimize patient morbidity. We present the third documented case of a hypopharyngeal paraganglioma and the first in the English literature.

**REFERENCES**