Malignant Paragangliomas of the Head and Neck: A Case Series and Literature Review

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Introduction

Head and neck paragangliomas (HNPG) are rare, slow-growing, hypervascular neoplasms that arise from extra-adrenal paraganglia cells located at different sites in head and neck. HNPGs can cause significant mass effect on adjacent structures and, very rarely, by catecholamine secretion with systemic effects. It is estimated that less than 10% of all HNPGs have malignant potential. Malignancy is commonly defined by neuroendocrine tissue occurring outside of the original tumor bed with isolated growth in another tissue or lymph node. Due to low incidence of malignant HNPG, the optimal treatment and prognosis are not well defined. Nonetheless, management of HNPG requires a systematic assessment through a well-coordinated, multi-disciplinary approach.

Purpose

The purpose of this study is to report two cases of malignant HNPG and to review the current literature relating to the malignant potential and the therapeutic strategies for these tumors.

Case Presentation

- A 47-year-old female presented with right-sided neck mass. Ct of the neck revealed multiple paragangliomas (PG): bilateral carotid bulbs and left skull base. She underwent staged surgical resection. Left glomus vagale tumor had one positive left level II lymph node consistent with metastatic jugular PG. (Fig 1) Follow-up MRI revealed residual disease in the region of left jugular foramen. She received post-op radiation to the left neck levels II-IV. Genetic testing was positive for hereditary PG syndrome type 1 (SDHD) gene mutation, which predisposes her for developing PGs, GIST, pheochromocytomas, pituitary and papillary thyroid tumors.

- A 35-year-old male presented with right neck mass. MRI of the neck revealed 3.0x3.1x4.6 cm enhancing right neck mass splaying the right external and internal carotid arteries (ICA) with enlarged adjacent lymph nodes. (Fig 2) Patient underwent surgical excision of the mass with partial resection of right ICA and subsequent reconstruction with interposition graft. On pathology, the surgical margin was positive with perineural invasion. One level II cervical lymph node was positive for metastatic PG. Octreotide scan was significant for 2 abdominal tumors for which he underwent resection. He received proton therapy as part of his overall treatment.

Imaging

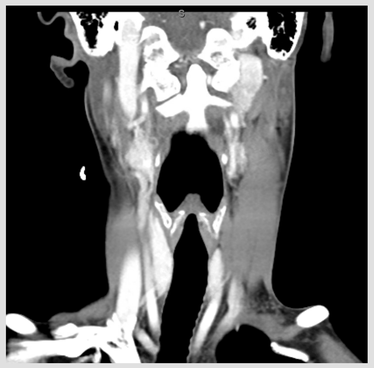

Figure 1

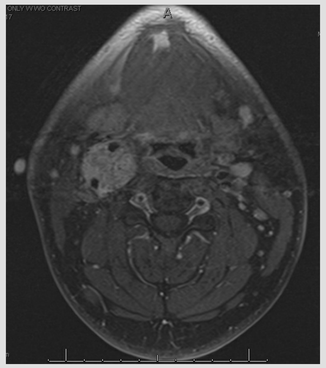

Figure 2

Figure 1. Neck CT, coronal view, an enhancing mass at the left skull base.

Figure 2. Neck MRI, axial view, showing an enhancing right carotid bifurcation mass.

Discussion

Currently, malignant PGs can only be diagnosed in the setting of local or distant metastatic disease. Optimal treatment of these rare tumors have not been well-documented within currently literature. Watchful waiting is an initial option that may be appropriate for elderly patients or select patients whose tumors remain asymptomatic. However, for those who require treatment, a multidisciplinary algorithmic approach should be utilized as it minimizes intra- and postoperative complications and aims to yield exceptional local tumor control. It is hypothesized that optimal treatment for malignant HPNGs is through complete surgical resection followed by adjuvant radiation.

Given it's high inheritance pattern, the cornerstone for management for these patients is to undergo genetic testing. This composition helps form the basis for diagnosis and leads to future follow-ups, as well as management for family members.

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