Paraganglioma of the Hypoglossal Nerve

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ABSTRACT

Objectives: To report a case of paraganglioma arising from the hypoglossal nerve and review the anatomy, clinical features, and literature to date.

Study Design: Case report and review of the literature.

Methods: Case records including paper and electronic chart and imaging reports were reviewed and summarized for the index case. A literature search was performed using PubMed keywords paraganglioma, hypoglossal, chemodectoma, carotid body, and glomus tumor. The available literature on the topic was reviewed and summarized.

Results: Paragangliomas associated with the hypoglossal nerve were reported in 4 cases over the past 47 years. Our index case presented with similar clinical features compared to those reported in the literature. Imaging with computed tomography and angiography showed a hypervascular mass at the carotid bifurcation, splying the internal and external carotid arteries. As in previously reported cases, the source of the paraganglioma was only identified intraoperatively. The current case differs from prior reported literature in that the tumor was dissected from the associated hypoglossal nerve which was preserved. The patient clinically had no deficits in articulation or deglutition following excision of the lesion and was able to return to a normal diet within 24 hours of surgery.

Conclusions: Hypoglossal paraganglioma is a neck mass that may not be distinguishable from more common carotid body or vagus tumors despite the use of multiple imaging modalities. Although XIIth nerve sacrifice may be required in some instances, nerve preserving surgery, when possible, allows for complete recovery without functional deficits.

INTRODUCTION

- Paraganglionomas are rare neuroendocrine tumors from extra-adrenal paraganglia of the autonomic nervous system.
- The most common sites include the carotid body, vagus nerve, jugular and tympanic ganglia. More rare sites include along Arnold’s nerve, Jacobson’s, nerve, or in the laryngeal, orbital, or nasal tissues.
- Rarely, paragangliomata can originate in relation to the hypoglossal nerve with four previously reported cases.
- Previous resections have required sacrifice of XIIth nerve with functional consequences.
- We present a case of complete surgical removal of a hypoglossal paraganglioma with preservation of the XIIth nerve. A review of the literature is provided as well as features relevant to this nerve preserving procedure.

FIGURE 1

- 74 year-old woman presented with an asymptomatic mass in the upper right neck.
- Physical exam revealed a non-tender 3 x 3 cm neck mass at the level of the carotid bifurcation anterior to the sternocleidomastoid muscle.
- All cranial nerves were functional and symmetric. She had no carotid bruits. The remainder of her head and neck exam was unremarkable.
- CT angiogram revealed hypervascular lesion splying the internal and external carotid arteries measuring approximately 3 cm. (Figure 2)

FIGURE 2

- An octreotide scan revealed a focal area of increased uptake in the right neck with no other abnormal areas of uptake in the body.
- Cerebral angiography revealed branches from the ascending pharyngeal and occipital arteries supplying the tumor. They were selectively embolized.
- Surgical exploration by joint otolaryngology and vascular surgery teams revealed hypoglossal (12th) nerve was noted to be hypervascular and intimately involved with the tumor (Figure 3)

FIGURE 3

- Although easily mobilized from the carotid bifurcation, the mass was adherent to the 12th nerve and appeared to derive the majority of its blood supply from perineural vessels.
- The nerve was dissected from the mass with ligation of arterial feeding vessels using 8-0 nylon suture. The mass was removed and the 12th nerve was intact to stimulation at the end of the case.
- Post-operatively the patient had no alterations in tongue mobility and no other cranial nerve deficits. The patient was able to eat a regular diet.

CONCLUSIONS

- Hypoglossal paraganglioma is a neck mass that may not be distinguishable from more common carotid body or vagus tumors pre-operatively despite the use of multiple imaging modalities.
- Although XIIth nerve sacrifice may be required in some instances, nerve preserving surgery, when possible, is diagnostic and allows for complete recovery without functional deficits.

REFERENCES