Hypoglossal Schwannoma Masquerading as a Carotid Body Tumor: A Diagnostic Dilemma

Matthew K. Lee, MD; Douglas R. Sidell, MD; Abie H. Mendelsohn, MD; Keith E. Blackwell, MD
Department of Head and Neck Surgery
David Geffen School of Medicine at UCLA

ABSTRACT

EDUCATIONAL OBJECTIVE: At the conclusion of this presentation, the participants should be able to demonstrate an understanding of the presentation, evaluation, and treatment of hypoglossal schwannomas. An emphasis will be placed on distinguishing its clinical and radiologic diagnosis from that of the more common carotid body tumor.

STUDY OBJECTIVE: Describe the clinical presentation, evaluation, and treatment of a hypoglossal schwannoma.

STUDY DESIGN: Case Report.

METHODS: We report an unusual case of a hypoglossal schwannoma presenting as a pulsatile level II neck mass at the bifurcation of the external and internal carotid arteries, mimicking a carotid body tumor. Radiologic findings are reviewed in detail.

RESULTS: A 59-year-old female presented to a tertiary care medical center with complaints of a pulsatile right sided neck mass, progressively enlarging over the past 2 years. An MRA of the neck was obtained for further evaluation, demonstrating a 5 cm mass located at the carotid artery bifurcation and causing spaying of the internal and external carotid arteries. Subsequent presentation and imaging, a presumptive diagnosis of a carotid body tumor was conferred. Upon formal angiography, the mass was unexpectedly discovered to be hypovascular in nature and not amenable to embolization. Intraoperatively, the mass was noted to arise from the hypoglossal nerve, remaining independent of the carotid artery. Despite attempts at preservation of the hypoglossal nerve, transection was required for complete tumor excision. The nerve was repaired primarily. On final histopathologic diagnosis, the mass was determined to be consistent with hypoglossal schwannoma.

CONCLUSION: Though rare, the hypoglossal schwannoma should remain a consideration in the evaluation of a parapharyngeal space mass. As this report demonstrates, the clinical and radiologic presentation of a hypoglossal schwannoma may closely mimic that of the more common carotid body tumor. As such, a high level of suspicion must be maintained whenever an abnormality in the usual presentation of a carotid body tumor is encountered.

INTRODUCTION

Magnetic resonance imaging (MRI) and computed tomography (CT) scanning play a significant role in the diagnostic workup of masses located in the retrostyloid parapharyngeal space. Retrostyloid neoplasms are frequently of neural crest origin, and both paragangliomas and schwannomas are common considerations in the differential diagnosis of such masses. Although paragangliomas and schwannomas share several similarities on imaging, it is often the unique characteristics of each lesion that allow for a definitive radiologic diagnosis.

The carotid body tumor is the most frequently identified paraganglioma in the head and neck, and has a typical appearance on imaging that is rarely replicated by other lesions. Despite this distinctive appearance, other lesions have been known to imitate the appearance of the carotid body tumor, thereby resulting in an inaccurate diagnosis. In this report, we present a patient with a rare hypoglossal schwannoma mimicking a carotid body tumor. To our knowledge, this is the first report of such a lesion.

CASE REPORT

A 59-year-old female presented to a tertiary-care university medical center, with a two-year history of a progressively enlarging right-sided neck mass. Symptoms on presentation were limited to neck discomfort and mild dysphagia. The patient had an otherwise unremarkable past medical history. On physical examination, a large, mobile, pulsatile level II neck mass was identified. A comprehensive cranial nerve examination was normal. An MRI demonstrated a mass at the level of the carotid bifurcation, with post gadolinium enhancement, and heterogeneity consistent with internal flow voids. An MRA of the neck was subsequently obtained, demonstrating a 5.3 x 3.3 cm heterogeneous mass causing spaying of the internal and external carotid arteries (FIGURES 1 AND 2).

Given the radiographic characteristics of the mass and findings on physical examination, a preliminary diagnosis of a carotid body tumor was established. In anticipation of surgical resection, the patient was subsequently scheduled for formal angiography with preoperative embolization. Interestingly, the mass was discovered to be hypovascular during angiography, and as such it was not amenable to embolization. The patient was subsequently brought to the operating suite for a definitive surgical excision. Intraoperatively, a gray, gelatinous-appearing mass was identified at the carotid bifurcation. Upon further dissection, the tumor was unequivocally identified as originating from the proximal segment of hypoglossal nerve, independent of the carotid artery. The distal segment of the hypoglossal nerve was identified as it exited the mass (FIGURE 3). In light of the preoperative angiography and intraoperative appearance, the constellation of findings favored the diagnosis of hypoglossal schwannoma with inferior extension. Electric stimulation of the mass resulted in a contractile response in the distribution of the hypoglossal nerve, lending credence to this diagnosis.

Initial attempts at subcapsular dissection with preservation of the hypoglossal nerve proved unsuccessful, as fibers of the nerve became progressively attenuated and inseparable from the tumor. The decision was thus made to transect the hypoglossal nerve and excise the mass. After excision, a neurorrhaphy was performed via primary anastomosis using epineural 8-0 nylon sutures in simple interrupted fashion. Final surgical pathology confirmed the working diagnosis of a hypoglossal schwannoma.

DISCUSSION

Carotid body paragangliomas are uncommon parapharyngeal space neoplasms that arise from chemoreceptor and sustentacular cells at the carotid bifurcation. Similar to paragangliomas, head and neck schwannomas are also benign neoplasms with an insidious growth pattern and are found to be associated with the vestibular nerve in greater than 90% of cases.1-4 Although rare, solitary extracranial cervical schwannomas involving the facial, vagus, hypoglossal and lingual nerve have also been described. The differential diagnosis for masses located in the parapharyngeal space can be refined based on preoperative imaging characteristics and physical examination findings. Currently, a standard component of the diagnosis and workup for parapharyngeal space lesions includes MR or CT angiography.2,3

Despite the clinical and radiologic presentation, the lesion was indeed determined to be originating from the hypoglossal nerve intraoperatively. In retrospect, the heterogeneous appearance of the lesion misconstrued as vascular flow voids was likely secondary to variable density of the enlarging neoplasm, a phenomenon that is known to occur in some schwannomas. Considering the natural location of the hypoglossal nerve, inferior growth of this neoplasm also allowed for the internal and external carotid artery to spay as it further occupied the carotid bifurcation, thereby resulting in the “lyre sign” typical of a carotid body tumor. To our knowledge, this is the first hypoglossal schwannoma mimicking a carotid body paraganglioma to be described in the literature.

CONCLUSIONS

Though rare, the hypoglossal schwannoma should remain a consideration in the evaluation of a parapharyngeal space mass. As this report demonstrates, the clinical and radiologic presentation of a hypoglossal schwannoma may closely mimic that of the more common carotid body tumor. As such, a high level of suspicion must be maintained whenever an abnormality in the usual presentation of a carotid body tumor is encountered.

REFERENCES