Pitfalls in imaging: Differentiating intravagal and carotid body paragangliomas

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Abstract

We report a case of an intravagal paraganglioma and a case of a carotid body tumor to illustrate a variable presentation of the former in which it mimicked the latter on preoperative imaging. The atypical imaging features of the intravagal paraganglioma included inferior extension to the level of the carotid bifurcation and splaying of the internal and external carotid arteries, features that are similar to those seen in a case of carotid body paraganglioma. Proper differentiation of these lesions permits more appropriate preoperative counseling and surgical preparation. When using magnetic resonance angiography rather than catheter angiography, we advocate the inclusion of gadolinium contrast and three-dimensional time-of-flight techniques to better demonstrate the position of the tumor relative to the carotid bifurcation.

Introduction

Magnetic resonance imaging (MRI) has greatly simplified the preoperative diagnosis of paragangliomas, revealing their distinctive salt-and-pepper pattern of multiple flow voids superimposed on a background of intense contrast enhancement. Differentiation among carotid body, intravagal, and jugulotympanic paragangliomas depends on proper localization of the mass in relation to the carotid bifurcation, the internal carotid artery, the internal jugular vein, and the skull base. This distinction is crucial because of the differences in patient preparation, surgical approach, the incidence of surgical complications, and the utility of adjunctive therapies required to treat these lesions.

We report a case of an intravagal paraganglioma and a case of a carotid body tumor to illustrate a variable presentation of the former in which it mimicked the latter on preoperative imaging.

Case reports

Patient 1. A 46-year-old woman was referred to our otolaryngology clinic with a 3-month history of right neck swelling. Physical examination revealed the presence of a 2-cm mass posterior to the angle of the mandible; this tumor was initially thought to be located within the parotid gland. No evidence of any cranial nerve deficit was noted.

MRI demonstrated an oval, sharply circumscribed, 3.1 × 1.8-cm mass in the right carotid space. The tumor had displaced the proximal internal carotid artery and the internal jugular vein anteriorly. The mass demonstrated a homogenous soft-tissue signal on T1- and T2-weighted imaging; multiple small, punctate foci of hypointensity suggested the presence of flow voids within the mass. Following intravenous administration of gadolinium contrast, the mass enhanced intensely and homogenously. The tumor extended inferiorly to the level of the carotid bifurcation and splayed the internal and external carotid arteries, which draped over the mass's anteromedial and posterolateral surfaces (figure 1, A and B). Magnetic resonance angiography (MRA) (figure 1, C) and duplex Doppler ultrasonography better demonstrated the splayed appearance of the carotid branches, and the appearance was thought to be most consistent with a carotid body paraganglioma.

At surgery, a 4 × 3-cm mass was found to intimately involve the vagus nerve, requiring sacrifice of a major portion of the nerve (figure 1, D); some posterior vagal fibers were preserved. Histopathologic examination revealed a characteristic cellular Zellballen pattern. Synaptophysin and chromogranin stains were positive, supporting the diagnosis of an intravagal paraganglioma.

Following surgery, the patient demonstrated vagal and hypoglossal nerve deficits. Although the hypoglossal deficit abated over 3 months, the patient continued to...
experience mild inspiratory dyspnea on exertion. Pulmonary function tests demonstrated an extrathoracic inspiratory obstruction consistent with paralysis of the right true vocal fold. The paramidline position of the fold identified on nasopharyngoscopy may have been the result of the vagus fibers spared at the time of surgery.

**Patient 2.** A 37-year-old man was referred to our otolaryngology clinic for evaluation of a 20-month history of a left neck mass. Physical examination identified the tumor as a 4-cm left lateral cervical mass. Findings on the remainder of the head and neck examination, including an assessment of cranial nerve function, were unremarkable.

Contrast-enhanced computed tomography (CT) of the neck demonstrated a left carotid sheath mass centered at the carotid bifurcation. There was a marked degree of diffuse enhancement following contrast administration. The internal carotid artery was displaced posteriorly, the external carotid artery was displaced anteriorly, and the jugular vein was displaced posterolaterally (figure 2, A). The mass measured 3.9 × 4.4 cm axially and extended 6 cm in the cranial caudal plane. Color flow imaging with ultrasonography of the left neck demonstrated extensive vascularization of the mass. Carotid catheter angiography demonstrated the typical splaying of the carotid vessels (lyre sign) (figure 2, B). Given the large size of the tumor and the adequate size of the feeder vessels, preoperative embolization of the external carotid artery was performed.

At surgery, the highly vascular lesion was removed. The mass had been positioned within the carotid bifurcation (figure 2, C). Histopathologic examination confirmed that it was a carotid body paraganglioma. Postoperatively, the patient experienced transient weakness of the left true vocal fold. At the 8-month follow-up, he had recovered completely.

**Discussion**

Intravagal paragangliomas can usually be differentiated from carotid body tumors by the site of origin. Both arise in the carotid space, but the former are centered 2 to 3 cm below the base of the skull, whereas carotid body tumors arise at the carotid bifurcation 7 to 8 cm below the skull base. Occasionally, intravagal paragangliomas extend inferiorly toward the carotid bifurcation and mimic carotid body tumors. Angiography, as well as CT and MRI performed after injection of contrast, are useful in differentiating the
two lesions. Intravagal paragangliomas displace the carotid vessels anteriorly—the internal artery medially and the external artery laterally; angiography shows a V-shaped splaying of the two vessels. Carotid body tumors displace the internal carotid artery posterolaterally and the external carotid artery anteriorly; angiography demonstrates the typical lyre sign.

The case of intravagal paraganglioma described in this article represents an unusual presentation in that it mimicked a carotid body tumor on imaging, which is the method of diagnosing these tumors. We advocate the use of noninvasive MRA with contrast enhancement to appropriately differentiate intravagal paragangliomas from carotid body tumors.

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