Operative Techniques in Otolaryngology (2016) 27, 7–14

ELSEVIER

Operative Techniques in Otolaryngology

Paraganglioma—Role of preoperative evaluation and endovascular embolization

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KEYWORDS
Paraganglioma;
Angiogram;
Balloon Test Occlusion;
Embolization

An understanding of the vascular architecture of paragangliomas can aid in both diagnosis and treatment. There is a growing role for vascular evaluation and intervention of these highly vascular tumors. This article addresses the specifics of the vasculature of paragangliomas, with focus on preoperative imaging, preoperative balloon test occlusion and endovascular embolization. The authors present both common and improvised techniques.

Introduction

Paragangliomas are rare neoplasms and account for 0.6% of all head and neck neoplasms and 0.03% of all neoplasms.[1] These tumors are neoplasms that originate from the neural crest progenitor cells that belong to the extra-adrenal chromaffin cell system. Currently they are sub-classified based on their location and neurovascular association. The 3 most common locations of this tumor are the carotid body (CP), (Figures 1 and 2) the jugular foramen (JP), (Figure 3) and the vagus nerve (VP).[2] The other site in the head and neck region includes Jacobson's nerve within the middle ear called tympanic paraganglioma (TP).

Paragangliomas are commonly detected in the age group of 40-50 years, more common in women than men and are more common in people living in high altitude and those with chronic pulmonary obstructive diseases.[3] Half of all paragangliomas are hereditary and may be associated with familial paraganglioma, neurofibromatosis type 1, von Hippel-Lindau disease, the Carney triad, and, rarely, with multiple endocrine neoplasia type 2. Multiplicity is common in hereditary forms, affecting up to one half of patients. Paragangliomas are primarily benign tumors, though 6% of them are malignant.[4]

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http://dx.doi.org/10.1016/j.otot.2015.12.004
1043-1810/

Evaluation

This article assumes that the diagnosis of a paraganglioma at any site has been entertained. In nearly every case, the diagnosis of a paraganglioma is made without the benefit of a tissue diagnosis hence imaging is quintessential in the work-up and sometimes treatment of these unusual tumors.

Imaging

Computed tomography scanning

Paragangliomas demonstrate a homogenous mass with intense enhancement after administration of intravenous

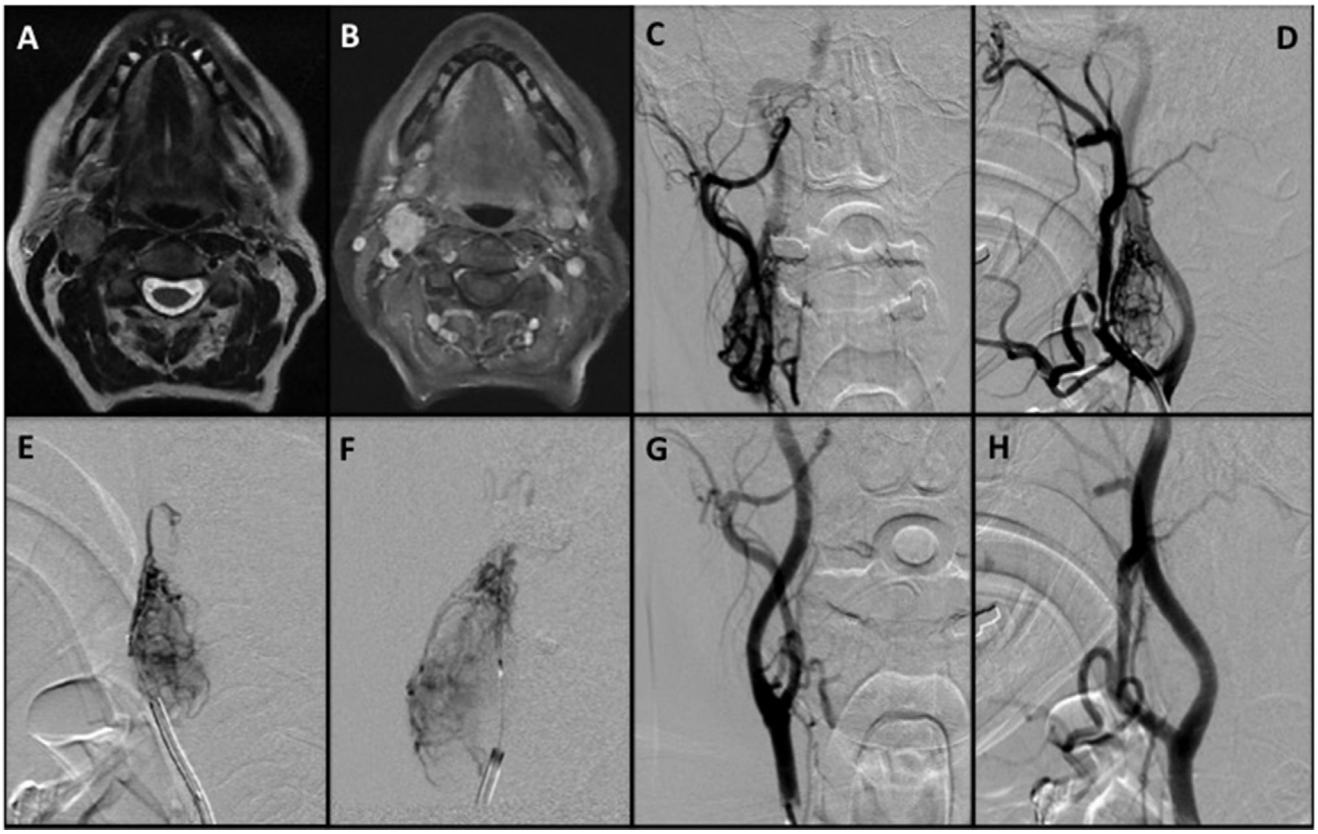

Figure 1 Right-sided carotid body tumor is shown in MRI T2W images (A) and postcontrast MRI (B). DSA images of right common carotid artery show the carotid body tumor located at the carotid bifurcation with splaying of the internal and external carotid arteries. (C and D). The arterial feeders are from the ascending pharyngeal artery. Super selective angiogram of the arterial feeders. (E and F). Postembolization angiogram show near complete devascularization of the tumor. (G and H).

contrast material.[5] The typical location of each paraganglioma is the key contributor to the specific diagnosis. CPs are seen as a well-defined homogenous mass within the carotid space of the infrahyoid neck, at the bifurcation of common carotid artery (CCA) and cause splaying of the carotid bifurcation.[5] Large lesions can extend into the suprahyoid neck and may demonstrate an inhomogeneous mass. In these instances, the internal carotid artery (ICA) is deflected posteriorly, distinguishing it from vagal paragangliomas.

VPs are usually confined to the parapharyngeal space, but large tumors may extend higher up through the JP into the posterior fossa or extend down toward the carotid bifurcation and cause displacement of the ICA anteriorly.[6] On high-resolution thin section bone window computed tomography (CT) of the temporal bones, JPs usually show a moth-eaten pattern of erosion and enlargement of the JP, which is result of irregular demineralization of the cortical bone (Figure 3A). Tumor expansion can occur superiorly and subsequently into the tympanic cavity, causing destruction of the ossicular chain.[7] Further enlargement of the tumor may lead to invasion of the bony canal of the facial nerve, with possible infiltration of the nerve. Intracranial posterior fossa extension can also occur.[5] TPs are often seen as a small, circumscribed mass on the cochlear promontory and as they enlarge they may encase the ossicles, extend into the mastoid and anteriorly into the Eustachian canal.

Magnetic resonance imaging

Magnetic resonance imaging (MRI) is the noninvasive imaging study of choice for paragangliomas as it better delineates soft-tissue contrast, whereas CT scan is better for looking at bony anatomy. In MRI, paragangliomas appear isointense to predominantly hypointense on T1-weighted images and hyperintense on T2-weighted images (Figure 1A and B, Figure 2A, Figure 3B and C). The most characteristic MR finding of paragangliomas is the presence of multiple serpentine and punctate areas of signal void within the tumor matrix. These areas of signal void are believed to be caused by high velocity flow of the intratumoral vessels. The adjacent areas of high-signal and low-signal intensity, give a "salt and pepper appearance," due to flow voids from enlarged intratumoral vessels.[8] With intravenous contrast injection, rapid and intense homogenous enhancement is visualized. The diagnosis is based on a combination of imaging features with the typical localization, associated with typical vessel displacement, with enlarged feeding vessels, and intratumoral flow signal makes the diagnosis of a paraganglioma highly likely.[9] Most tumor feeders also are identified when using 3-dimensional Time Of Flight MR angiography. The sensitivity of 3-dimensional Time Of Flight MR angiography, however, is too low to show the specific feeding vessels in