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Jugular vein invasion rate in surgically operated paragangliomas: a multimodality retrospective study

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ABSTRACT

Purpose: To assess the rate of jugular vein (JV) invasion by paragangliomas and imaging's ability to predict invasion.
Methods: Imaging studies of patients with paragangliomas were evaluated for JV invasion. Rates of invasion by different paragangliomas and each modality's accuracy were determined.
Results: JV invasion occurred in 11/11 glomus jugulares (GJs), 3/5 glomus vagales, and 2/8 carotid body tumors. Accuracy for magnetic resonance imaging (92.3%), computed tomography (92.3%), and digital subtraction angiography (94.1%) was comparable.
Conclusions: JV invasion is characteristic of but not specific to GJs. All modalities show high accuracy in detecting invasion.

1. Introduction

Paragangliomas are tumors that arise from clusters of extraadrenal neuroendocrine cells of neural crest origin called paraganglia. Parasympathetic paraganglia are collections of cellular elements called glomus cells that primarily function as chemoceptors and do not normally show endocrine activity. Paragangliomas may be classified according to their location and endocrine functionality [1,2] and involve the head and neck region in 3% of cases [3]. They represent 0.6% of tumors in this area [4].

Within the head and neck, these tumors arise in four main locations: the carotid space as carotid body tumors (CBTs), the middle ear as glomus tympanicum, the jugular foramen as glomus jugulare (GJ), and along the vagus nerve as glomus vagale.

All of these pathologic entities, with the exception of the localized glomus tympanicums, possess a close anatomic relationship with the jugular vein (JV) and the internal carotid artery (ICA) [4–8]. Treatment options comprise surgery, radiotherapy, and watchful waiting with imaging follow-up in selected cases. Surgery is the mainstay if complete cure is desired, and it often requires careful control of the ICA and JV during vessel skeletonization from the tumor and/or the surrounding tissues [3,5]. Presurgical knowledge of JV invasion by these tumors can be important to reduce the risk of intraoperative bleeding, including tumors in which it is not expected (CBTs and glomus vagales) [9–12]. The aim of this paper is to determine whether JV invasion is specific for GJ and to learn the specificity (Sp), sensitivity (Se), positive predictive value (PPV), negative predictive value (NPV), and accuracy (Acc) of various diagnostic imaging modalities in the evaluation of JV invasion so that we would be able to recommend the most appropriate studies for presurgical evaluation. To our knowledge, this is the first study specifically addressing the matter. The spatial relationship of paragangliomas with the JV and the ICA were also assessed. We hypothesized that only GJ tumors, of all the paragangliomas, would invade the JV and be specific for this finding.

2. Materials and methods

This retrospective study was Health Insurance Portability and Accountability Act (HIPAA) compliant and approved by the institutional review board. The study included all patients surgically treated

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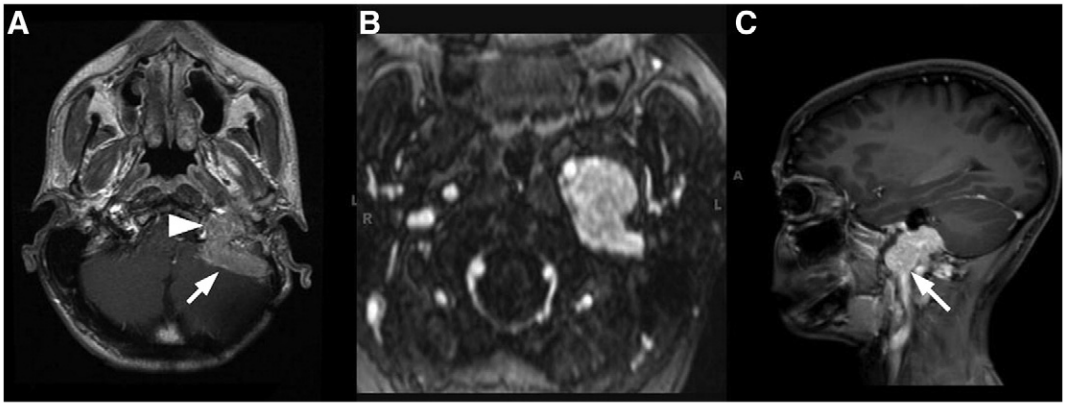

Fig. 1. (A) MRI (T1WI with contrast) of a jugular foramen paraganglioma (arrowhead) with JV invasion up to and including the sigmoid sinus (arrows). (B) Glomus vagale with surgical proof of JV invasion. (C) Contrast-enhanced sagittal T1WI shows growth of a GJ into the JV (arrow).

at our institution in the past 10 years for a head and neck paraganglioma that had at least one preoperative neuroradiological study. The imaging modalities which were taken into account, carried out following our standard institutional protocols, were as follows: (1) contrast-enhanced (100 cc of Omnipaque 350) computed tomography (CT) of the neck with acquisition at 80 mAs and (2) magnetic resonance (MR) with the following sequences: axial and sagittal T2WI, FLAIR, and T1 fat-sat postcontrast using gadolinium at a dose of 0.1 mmol/kg, time-of-flight MR angiogram, diffusion-weighted imaging, and susceptibility-weighted imaging; digital subtraction angiography (DSA) images were part of the preoperative embolization procedure and lasted until the venous phase. The patients were identified via a radiology information system (RIS) keyword search. Only patients who had pathologic proof of diagnosis and/or intraoperative assessment of the JV status were included among the RIS identified cases.

Each cross-sectional imaging study was blindly reviewed by a subspecialty trained neuroradiologist with 25 years of skull base imaging experience for characteristics of the tumor regarding the neck vessels, including whether the JV was invaded, if it was displaced and in which direction, and the relationship of the mass with the ICA. Tumors were distinguished according to their location: the carotid bifurcation for the CBT, the cervical ICA for the glomus vagale, and the jugular foramen for the GJ. If a patient was studied with different modalities, he/she was blindly reviewed at different times as separate exams spaced at least 2 weeks apart by the same diagnostic neuroradiologist. An attending interventional neuroradiologist with 18 years of experience reviewed the DSA studies to detect signs of intravascular invasion of the JV. Regarding JV invasion, with cross-sectional imaging modalities, the finding was considered positive if there was enhancing tissue growth inside the vessel and/or if there was a narrowing of the lumen by the tumor without identifiable pathology-free vessel wall. DSAs were considered positive for JV invasion in the case of minimal or absent flow in the vessel during the venous phase with or without evidence of aberrant venous drainage through collaterals or the contralateral venous circulation. The cranial boundary of the JV was defined as the jugular bulb, without considering the sigmoid sinus.

The definitive proof of JV invasion was obtained by a blinded review of the surgical and pathological notes of the patients included in the database by a research fellow in neuroradiology. The surgeons' description of tumor removal from the lumen or inner wall of the JV was considered as the definitive proof of JV invasion. The diagnosis of paraganglioma was based on the review of the pathological reports of the surgical specimens.

The obtained data were processed and analyzed to determine the rate of JV invasion for each subtype of tumor and to assess the Sp, Se, NPV, PPV, and Acc of each imaging modality in predicting such invasion. Descriptive statistics of the spatial relationships of these tumors with the ICA and the JV were also obtained.

3. Results

The study group consisted of 22 patients, 12 males (54.5%) and 10 females (45.5%) with an average age of 47.9 years (range: 26–68 years, SD: 13.2). Twenty-four tumors were identified in the patient group. Eleven tumors (45.8%) were GJ, 8 (33.3%) were CBT, and 5 (20.8%) were glomus vagale. The mass was on the right side in 15/24 cases (62.5%) and on the left side in 9/24 cases (37.5%). Two tumors were multiple; in both cases, the association was between a CBT and a glomus vagale, in one case on the same side and in the other case contralateral. All these tumors were pathologically proven paragangliomas with chromogranin, synaptophysin, and/or S100 protein positivity in the pathological specimens.

Multimodality imaging studies identified JV invasion (intraluminal growth or wall invasion from a neighboring tumor) following the above-mentioned criteria in 16 tumors (66.7%). In all cases, JV invasion was surgically confirmed by the review of the surgical note. Of these 16 tumors, 11 (68.8%) were GJ, 3 (18.7%) were glomus vagale, and 2 (12.5%) were CBTs. Thus, JV invasion was present in 100% of the GJ tumors, in 60% (3/5) of the glomus vagales, and in 25% (2/8) of the CBTs. GJs showed invasion of the jugular bulb with growth of pathologic tissue inside the lumen of the vessel in a caudal direction (Fig. 1). Cranial growth into the sigmoid sinus was also noted in 9/11 (81.8%) of these tumors. CBTs and glomus vagales that involved the vein invaded the vessel from the outside, hence involving the wall in their growth with subsequent narrowing the lumen. Four GJs showed invasion of the posterior cranial fossa, in 2 cases compressing the cerebellum, and in 2 cases compressing the cerebello-pontine angle.

MR imaging (MRI) was performed in 13 lesions and gave 10 positive results. Of these 10 positive exams, 1 was a false-positive case of a CBT that was encasing the JV but was not found to invade it during surgery. There were no false negatives. Data analysis showed an Se of 100%, an Sp of 75%, a PPV of 90%, and an NPV of 100%. Acc for MRI in detecting JV invasion was 92.3%.

CT was performed in 13 tumors with 9 cases in which the JV was read as invaded. There was one false-positive case of a glomus vagale in which the lumen was almost completely narrowed at the imaging but in which the surgical note did not confirm invasion (Fig. 2). CT did not reveal any false-negative studies, showing an Se of 100%, an Sp of 80.0%, a PPV of 88.9%, and an NPV of 100%. Acc was 92.3%. Disagreement between the interpretation of findings in MRI and CT was present in the case of the right CBT that the CT correctly classified