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Giant recurrent glomus jugulotympanicum with intracranial, extracranial, and nasophayngeal extension: The imaging role in clinical management

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We present a 50-year-old male with a massive glomus jugulotympanicum that demonstrated extracranial extension breaking through the skin in the posterior auricular region, intracranial extension into the middle cranial fossa, and nasopharyngeal extension involving the entire length of the eustachian tube. Characteristic CT, MR, angiographic, and pathologic findings are demonstrated in this case. The difficult decision regarding treatment options must be made with careful consideration of the associated morbidities of slow tumor growth and recurrence with conservative management versus the surgical morbidities of aggressive surgical resection. Imaging is arguably the most important component of this decision-making process, with the radiologist contributing significantly to multiple aspects of the management of glomus tumors.

Introduction

Paragangliomas, also known as glomus tumors and chemodectomas, of the head and neck typically are slow-growing benign masses with a favorable prognosis. The four most common extra-adrenal sites are the carotid body at the common carotid artery (CCA) bifurcation, the jugular foramen, along the vagus nerve, and within the middle ear. Glomus tympanicum, the middle-ear variety, often exhibits recurrence. Glomus jugulare refers to paragangliomas arising from the adventitia of the jugular vein in the jugular fossa. The term glomus jugulotympanicum is used when tumor size precludes this distinction.

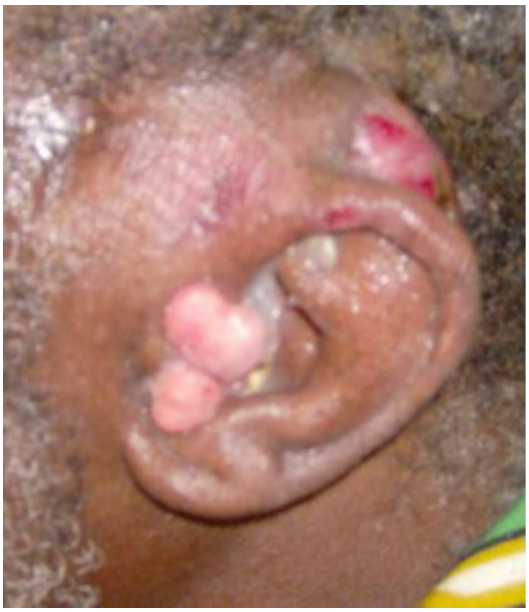

Figure 1. Photograph of left ear with pink, glistening tumor extending through the left EAC and skin of the posterior auricular surface. Bulging of the intact skin by the underlying mass can also be noted.

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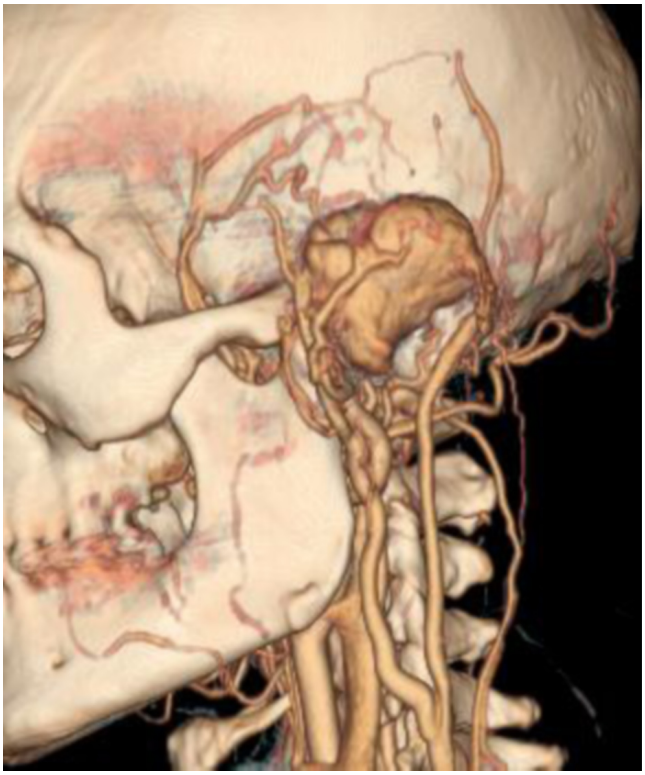

Figure 2A. 3D volumetric reconstructed image obtained from a CTA after IV contrast administration correlates well with the physical findings demonstrated in Fig. 1 and also demonstrates the extensive vascular supply of the tumor, primarily from branches of the external carotid artery.

Case report

A 50-year-old male was initially transferred to our institution after hemorrhage from his left ear, which required transfusion. Although the surgical history could not be validated from outside hospital records, the patient did report a left middle-ear tumor resection approximately 10 years prior. Multiple attempts at obtaining these medical records were unsuccessful.

Physical examination revealed a large pink, glistening tumor emerging from and occluding the left external auditory canal, with lateral displacement of the pinna. The mass also eroded through the mastoid sinus, skin, and posterior auricular sulcus, where the pink tumor could also be visualized by inspection (Fig. 1). Left cranial nerve palsies of VII, VIII, and XII were elicited.

Initially, CT angiography demonstrated a large, avidly enhancing vascular mass with extensive collateral vessels centered in the left middle ear extending through the EAC, pinna, and soft tissues of the periauricular region with evidence of skin erosion (Fig. 2, A-C). Gadolinium-enhanced MRI was subsequently performed for further characterization. The tumor extended to involve the length of the left eustachian tube. Bony invasion was demonstrated, with the mass opacifying the mastoid sinus and petrous apex. Intracranially, there was extension into the left middle cranial

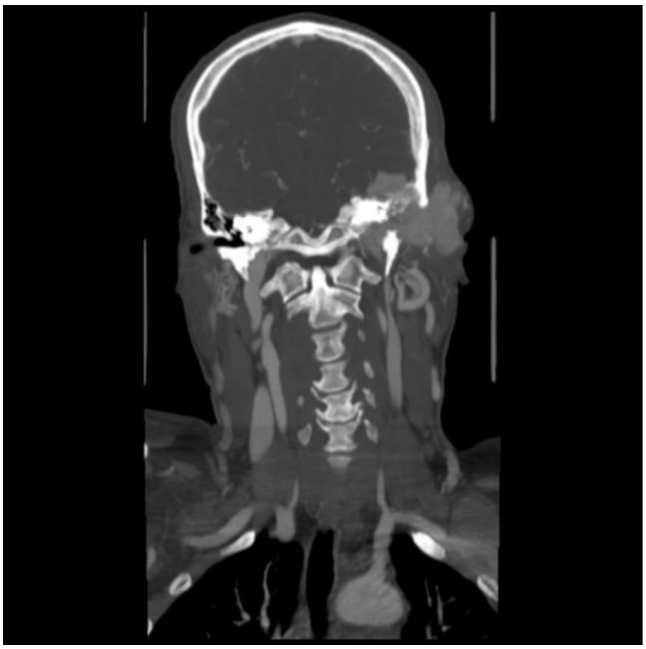

Figure 2B. Coronal CT angiogram. Reformatted image after contrast administration demonstrates enhancing mass centered in the left middle ear, extending intracranially to the middle cranial fossa. The tumor can also be seen surrounding the vertical segment of the left petrous internal carotid artery with extension into the left peri-auricular region.

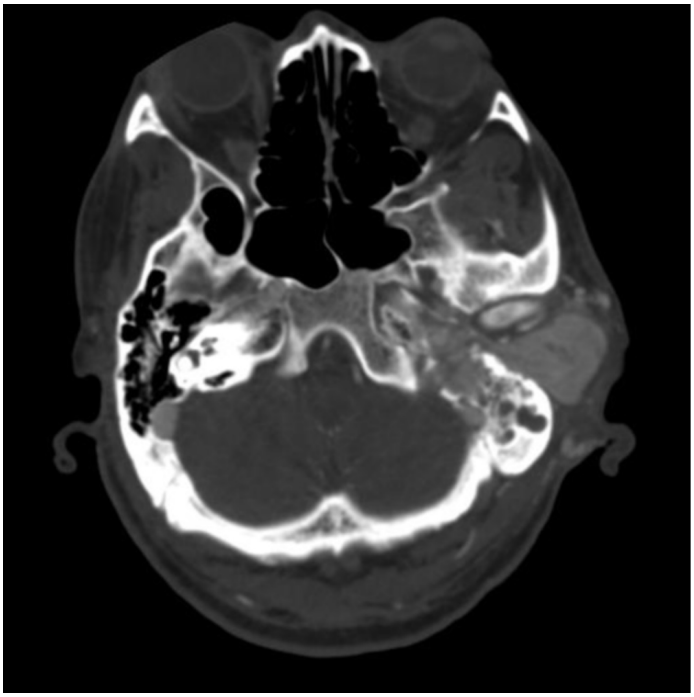

Figure 2C. Axial CT images after administration of contrast with soft tissue algorithm demonstrated extensive bony erosion of the jugular bulb region with opacification of left mastoid sinus.

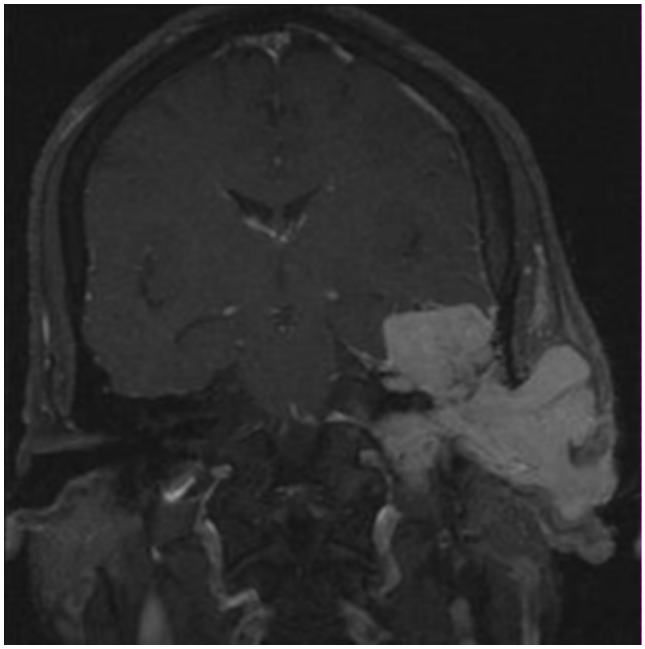

Figure 3A. Coronal gadolinium-enhanced T1 MRI with fat suppression demonstrates the avidly enhancing mass. Extension through the EAC into the peri-auricular region is seen, with the enhancing mass involving the middle cranial fossa. Note linear dural enhancement on the left side and the "salt-and-pepper" appearance of the left jugular foramen mass.

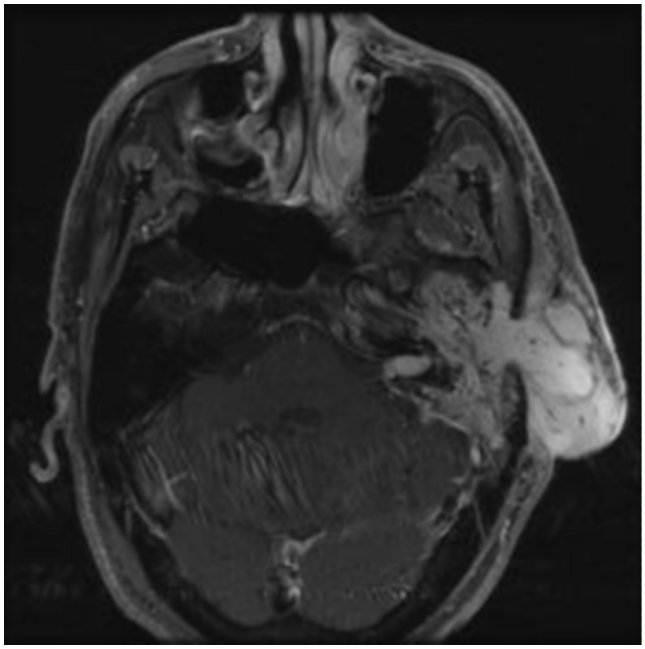

Figure 3B. Axial gadolinium-enhanced T1 MRI Image with fat saturation demonstrating large enhancing tumor involving the temporal bone, middle ear, and peri-auricular region. The tumor is also seen extending into the left IAC.

fossa and into the left internal auditory canal. Multiple small flow voids were seen on MRI, giving the tumor the characteristic "salt and pepper" appearance and suggesting a glomus tumor (Fig. 3, A-C). Based on the clinical history, this tumor was thought to have originated in the middle ear as a glomus tympanicum. However, given the size, this tumor was referred to as a glomus jugulotympanicum.

Given the clinical presentation of hemorrhage, the large size of the mass, and the extensive vascular supply of the tumor, conventional angiography was performed to achieve hemostasis and to reduce the potential risk of bleeding

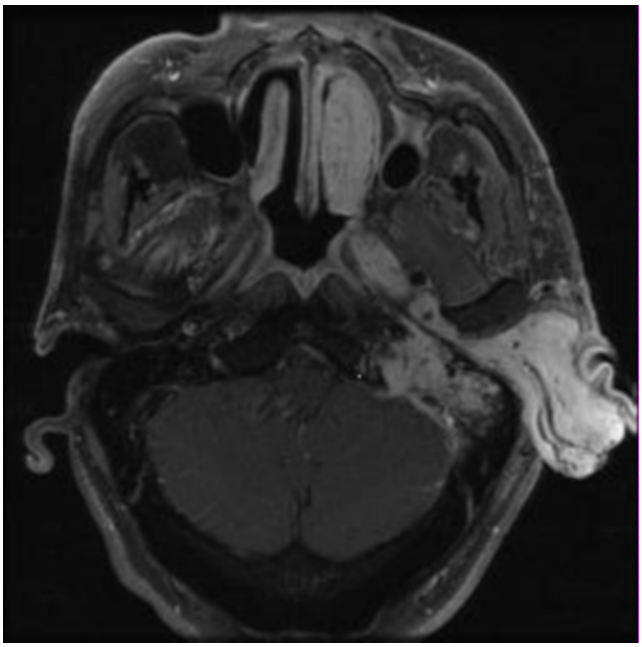

Figure 3C. Axial gadolinium-enhanced T1 MRI with fat saturation demonstrating tumor along the length of the eustachian tube with extension into the left nasopharyngeal region.

complications of tissue biopsy. Angiography demonstrated feeder branches to the tumor from the petrous and cavernous left internal carotid artery, and extensive tumoral blush originating from multiple external carotid artery branches (Fig. 4, A-B). Feeder branches from the superficial and deep temporal arteries were embolized using 100- to 300-micron embolization spheres. The embolization procedure was initially successful in controlling hemorrhage.

However, subsequent attempts at biopsy were abandoned after significant hemorrhage, despite the preoperative embolization procedure. This delayed plans for a possible combined otolaryngology and neurosurgical approach resection, as the expected diagnosis of paraganglioma was not confirmed. Unfortunately, the patient was lost to followup but presented again approximately two years later with reports of a recurrent episode of hemorrhage that required blood transfusion at an outside institution.

A repeat MRI with gadolinium showed enlargement of the tumor, and a successful, uncomplicated biopsy confirmed the diagnosis of paraganglioma. Given the patient's overall high functionality and the likely associated morbidity of surgery, radiation therapy was recommended in attempts at slowing tumor growth and reducing vascularity. No additional embolization procedures have been required at the time of this report.

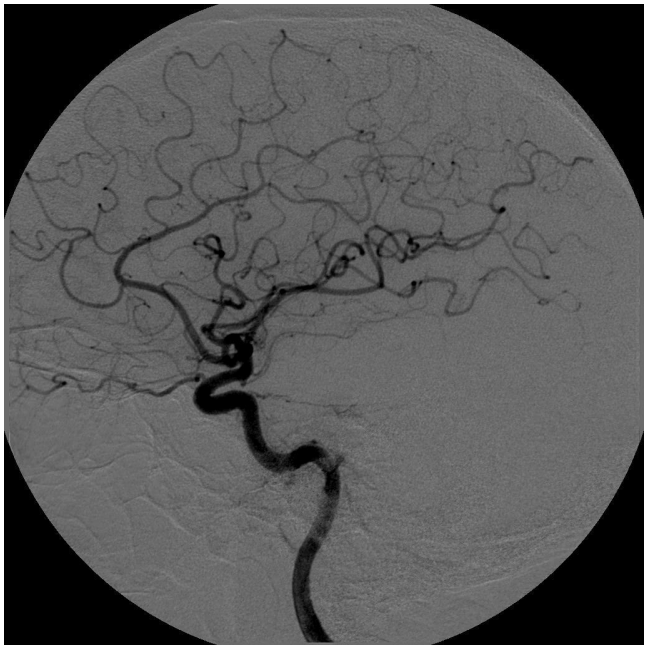

Figure 4A. Lateral view of left carotid injection angiogram in arterial phase image demonstrating tentorial meningeal branch supplying the tumor from the petrous internal carotid artery.

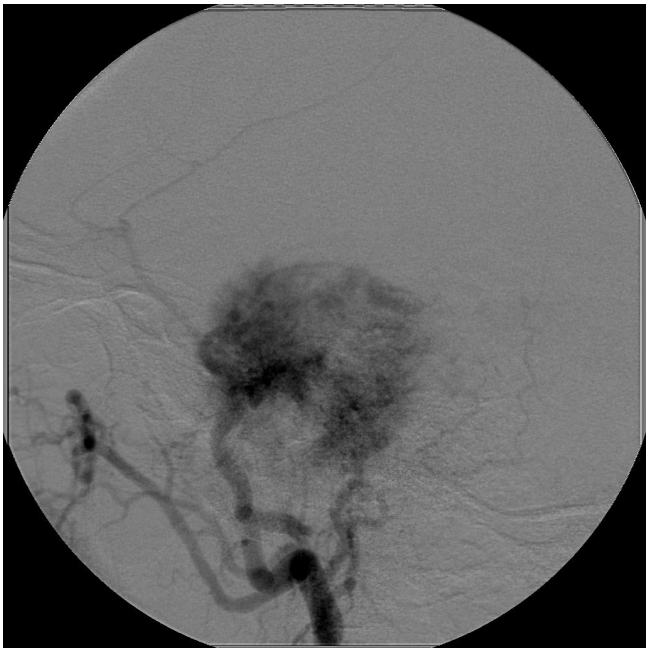

Figure 4B. Angiogram image demonstrating the extensive vascular supply from multiple external carotid artery branches with diffuse tumoral blush.

Discussion

Paragangliomas are neoplasms arising from glomus bodies, which are widely dispersed throughout the body and composed of cells of neural crest origin. The classic, pheochromocytoma, is a catecholamine-secreting paraganglioma arising from the adrenal medulla. Although the vast majority of these tumors are nonfunctional when extra-adrenal, catecholamine-secreting head and neck paragangliomas have been reported (1). Paragangliomas are second only to schwannomas in frequency of temporal bone tumors (2).

Marchand reported the first paraganglioma (of the carotid body) in 1891 (3). The term glomus tumor was used to describe the rich presence of vascular channels and nerves seen in these masses (4). Mulligan proposed the term chemodectoma to reflect the chemoreceptor tissue of origin. Glenner and Grimley coined the term paraganglioma, which is currently accepted and most widely used in the modern medical lexicon to describe these lesions (5, 6). To classify paragangliomas anatomically, Glenner and Grimley also divided the extra-adrenal paraganglion into several anatomic groups. The branchiomeric paraganglia arise in association with arterial vessels and cranial nerves of the head and neck region. Intravagal paraganglia are located within the perineurium of the vagus nerve, usually at the jugular or nodose ganglion (7). Paraganglia rests in the temporal bone occur with three discrete bodies, which are closely related to the auricular branch of the vagus nerve (Arnold nerve) and tympanic branch of the glossopharyngeal nerve (Jacobson nerve). About 50-55% of tumors in these sites are seen in the dome of the jugular bulb or along the paths of these two nerves. About 25% occur along the Jacobson nerve in the mucosa of the cochlear promontory (8, 9). These small lesions are confined to the middle ear or may extend to the mastoid air cells posteriorly.

Paragangliomas are named based on the site of origin. An intact jugular bulb is important in distinguishing glomus tympanicum from the more common glomus jugulare (11, 12). Glomus jugulare tumors arise from the jugular foramen; their spread typically follows the path of least resistance, including mastoid air cells, vascular channels, eustachian tube, and neural foramen (13). Glomus tympanicum, confined to the middle ear, has also been reported to demonstrate nasopharnyngeal spread (13, 14). Vagal paragangliomas, also called glomus vagale tumors, arising from the superior ganglion can appear "dumbbell"-shaped, with superior extension into the posterior fossa and inferior extension into the infratemporal space. More commonly, glomus vagale tumors arise from the inferior ganglion and appear spindle-shaped, with compression of the internal jugular vein. Splaying of the internal and external carotid

arteries at the common carotid artery bifurcation is characteristic of carotid body paragangliomas (1).

The most characteristic location of glomus tympanicum tumors is lateral to the cochlear promontory. CT with bone windows is an excellent tool to show an intact jugular fossa and caroticotympanic spine. However, if the tumor is large and involves the jugular bulb, it cannot be distinguished from a glomus jugulare tumor, and the term glomus jugulotympanicum applies. Our case demonstrates an exaggerated example of this dilemma. Another, rarer, paraganglioma of the facial nerve, glomus faciale, has been reported in a predominately extracranial location, and this could be a consideration in our case (1). However, the tumor in our case was centered in the middle ear, and the surgical history of middle-ear tumor resection led us to believe that this tumor originated as a glomus tympanicum.

Typically described as benign, slow-growing lesions, glomus tumors can be seen in both children and the elderly but usually present in middle-aged adults with a peak incidence in the 5th and 6th decades. A distinctly higher incidence is reported in females, with male-to-female ratios reported up to 1:5 (1, 2). Glomus tympanicum is the most common primary tumor of the middle ear, and usually presents with pulsatile tinnitus followed by hearing loss and otalgia (11, 13, 14). Bleeding is an uncommon presentation despite the tumor typically being highly vascular. Additionally, glomus tympanicum tumors often recur after resection, which is a key consideration in management decisions. Surgical resection is traditionally the treatment of choice for glomus tympanicum, but radiotherapy is often used in conjunction for large tumors or in an attempt to slow growth in unresectable tumors (12, 14). Conservative approaches with combined partial resection and radiotherapy have been advocated in the elderly (14).

One of the most debated issues regarding these tumors is the decision about conservative management versus aggressive surgical treatment. Regarding glomus tympanicum, total resection is usually the goal, as recurrence is not uncommon. Preoperative evaluation with CT, high-field MRI, magnetic resonance angiography, and digital subtraction angiography has been proposed as essential for optimal planning (3, 16). Careful evaluation of the hypotympanum and jugular bulb is important, as involvement alters the surgical approach (15, 16). Other important areas to evaluate on imaging include the inner ear, occipital condyle, and vertebral artery, which may need to be sacrificed if total resection is attempted. Internal carotid-artery involvement has also been reported, and preoperative interventional techniques have been proposed if more than 270 degrees of the internal carotid artery are engulfed on MRI. Interestingly, preoperative stenting of the internal carotid artery may decrease tumor vascularity if the tumor infiltrates the wall of the artery. Bony infiltration occurs frequently at the spongy bone of the petrous apex and requires extensive drilling for removal (16). Obviously, more extensive tumor creates complicated surgical cases, and despite advancing surgical techniques, associated morbidities also increase significantly.

On the other hand, the growth rate of paragangliomas is typically slow and has been reported at approximately 1.0 mm per year, with a doubling time of 4 years. Conservative approaches with radiation with or without partial tumor resection have been proposed for elderly patients in order to minimize the morbidities of both tumor growth and surgery. Radiation techniques have advanced and are approximately 90% successful at inhibiting tumor growth (8). However, radiation also entails risks of osteoradionecrosis, radiation-induced malignancy, and xerostomia. A further complication is that a malignant variant of paraganglioma exists and is diagnosed by metastatic spread, as the histology is very similar to the more common nonmalignant counterpart. Younger patients have a longer life expectancy, and unfortunately tend to have higher tumor growth rates, guiding management decisions toward aggressive surgery.

Preoperative techniques, which involve percutaneous injection of ethylene-vinyl alcohol copolymer to devascularize paragangliomas before surgical resection, have been described (17, 18). Wanke et al reported a series of four patients with six paragangliomas at the bifurcation of the common carotid artery, which were treated with preoperative percutaneous embolization (18). Their results suggest that this technique may provide better devascularization than traditional embolization techniques, although additional studies are needed. If hemostasis could not have been achieved in our case, this technique could have been considered, given the superficial access of the tumor. However, careful precautions to prevent distal embolization would be imperative (18).

Although our case is a massive tumor that would require extensive surgery with unavoidable morbidities, surgical resection was considered. However, after much discussion of the potential outcomes and consideration of the patient's functional status, the patient elected for conservative radiation therapy aimed at slowing tumor growth and decreasing tumor vascularity. If bleeding continues to be a clinical complication, further embolization procedures may be pursued. In addition to the impressive extracranial presentation, this patient provided an excellent demonstration of the characteristic imaging features of glomus tumors and an opportunity to evaluate the role of the radiologist in the extreme consequences of management decisions.

References

1. Mafee MF, Raofi B, Kumar A, Muscato C. Glomus faciale, glomus jugulare, glomus tympanicum, glomus vagale, carotid body tumors and stimulating lesions: Role of MR imaging. Radiol Clin North Am 2000;38:1059-76. [PubMed]
2. Rao AB, Koeller KK, Adair CF. Paragangliomas of the head and neck: Radiologic-pathologic correlation. Radiographics 1999;19:1605-1632. [PubMed]
3. Lack EE. Tumors of the adrenal gland and extra-adrenal paraganglia. In: Rosai J, eds. Atlas of tumor pathology. Ser 3, fasc 19. Washington, DC: Armed Forces Institute of Pathology, 1997; 303-409.

4. Guild SR. The glomus jugulare, a nonchromaffin paraganglion in man. Ann Otol Rhinol Laryngol 1953; 62:1045-1071. [PubMed]
5. Mulligan RM. Chemodectoma in the dog. Am J Pathol 1950; 26:680-681.
6. Glenner GG, Grimley PM. Tumors of the extra-adrenal paraganglion system (including chemoreceptors). In: Firminger HI, eds. Atlas of tumor pathology. Ser 2, fasc 9. Washington, DC: Armed Forces Institute of Pathology, 1974; 1-90.
7. Neves F, Huwart LH, Jourdan G, et al, Head and Neck Paragangliomas: Value of Contrast-Enhanced 3D MR Angiography. Am J Neuroradiol 2008;29:883-89. [PubMed]
8. O'Leary MJ, Shelton C, Giddings NA, Kwartler J, Brackmann DE. Glomus tympanicum tumors: a clinical perspective. Laryngoscope 1991;101:1038–1043 [PubMed]
9. Phelp PD, Stansbie JM. Glomus jugulare or tympanicum? The role of CT and MR Imaging with Gadolinium DTPA. The Journal of Laryngology and Otology 1988;102:766-776. [PubMed]
10. Connor SE, Gleeson MJ, Odell E. Extracranial glomus faciale tumour. The Journal of Laryngology & Otology 2008;122:986-989. [PubMed]
11. Alaani A, Chavda SV, Irving RM. The crucial role of imaging in determining the approach to glomus tympanicum tumours. Eur Arch Otorhinolarngol Oct. 8. [Epub ahead of print] [PubMed]
12. Lum C, Keller AM, Kassell E, et al. Unusual Eustachian Tube Mass: Glomus Tympanicum. Am J Neurorad 2001;22:508-509. [PubMed]
13. Weissman JL, Hirsch BE. Beyond the Promontory: The Multifocal Origin of Glomus Tympanicum Tumors. Am J Neurorad 1998;19:119-122. [PubMed]
14. Cosetti M, Linstrom C, Alexiades G. Glomus Tumors in Patients of Advanced Age: A Conservative Approach. The Laryngoscope 2008;118:270-274. [PubMed]
15. Li G, Chang S, Adler JR, et al. Irradiation of glomus jugulare tumors: a historical perspective. Neurosurg Focus 2007;23:E13 [PubMed]
16. Sanna M, De Donato G, Piazza P, Falcioni M. Revision Glomus Tumor Surgery. Otolaryngol Clin N Am 2006;39:763-782. [PubMed]
17. Quadros RS, Gallas S, Delcourt C, et al. Preoperative Embolization of a Cervicodorsal Paraganglioma by Direct Percutaneous Injection of Onyx and Endovascular Delivery of Particle. AJNR Am J Neuroradiol 2006;27:1907-1909. [PubMed]
18. Wanke I, Jackel MC, Goericke S, et al. Percutaneous Embolization of Carotid Paragangliomas Using Solely Onyx. AJNR Am J Neuroradiol 2009; www.ajnr.org – Epublished ahead of print. [PubMed]