

GLOMUS JUGULARE TUMOR: A CLASSIC PRESENTATION AND REVIEW OF THE LITERATURE

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ABSTRACT

Background: Glomus jugulare tumor (GJT) is a slow-growing, benign neural crest tumor arising in the jugular foramen of the temporal bone. It is a type of paraganglioma. The incidence has been cited as 1 per 1.3 million people.

Aims: We aimed to present a classic case of a glomus jugulare tumor and a practical approach to identifying and managing this condition.

Methods: A chart review of a single case of GJT was performed along with a review of relevant literature.

Results: A GJT can be managed with observation, preoperative embolization, surgery, radiotherapy or radiosurgery, with traditional surgery being the definitive treatment. Radiation can be used as primary or adjunct treatment in patients who are elderly, have increased morbidity, or if the tumor is near significant arteries, nerves, or veins.

Conclusion: GJT's were once thought to be exceedingly rare, but now are increasing in incidence, perhaps due to the increased use of radiographic imaging. They are slow-growing tumors with subtle symptoms and will often go undiagnosed for a delayed period of time. Physician awareness should be increased on the presentation and treatment options in this setting to optimize patient care.

Keywords: Glomus jugulare; Paraganglioma; Gamma knife; Tumor control

INTRODUCTION

Glomus jugulare tumor (GJT) is a slow-growing, benign neural crest tumor arising in the jugular foramen of the temporal bone. As a type of paraganglioma, these tumors are highly vascular neuroendocrine neoplasms found in the autonomic nervous system. The most common paragangliomas in the head and neck are carotid body, glomus jugulare, glomus tympanicum, and glomus vagale tumors. GJT is the most common neoplasm of the middle ear, and the second most common of the temporal bone. The incidence of glomus jugulare tumor has been cited as 1 per 1.3 million people. They affect females more commonly than males at a 3: 1 ratio and present in the fifth and sixth decades of life [1]. These tumors are typically benign but locally invasive and can lead to erosion of the temporal bone, though 1-5% of cases are malignant [2]. One to three percent of glomus tumors are functioning paragangliomas and secrete catecholamines, causing hypertension, palpitations, and headaches [1]. Herein we present a classic case of a GJT and a practical approach to identifying and managing this condition.

CASE PRESENTATION

A 63-year-old female presented to the otolaryngology clinic with right-sided pulsatile tinnitus, hearing loss, and intermittent vertigo for several months. These symptoms were associated with aural fullness, intermittent racing heartbeat, and a positional globus sensation. Otolaryngoscopic examination revealed an intact tympanic membrane, but a retrotympanic cherry-red bulge was seen in the posterior-inferior quadrant. Her contralateral ear was unremarkable. Further cardiovascular maneuvers revealed improvement in tinnitus volume with compression of the right neck. Her cranial nerve exam was within normal limits.

An audiogram and radiographic images were ordered and highly suggestive of a GJT. The audiogram showed mixed conductive and sensorineural hearing loss of the right ear, with mild sensorineural hearing loss of the left ear. Computed tomography (CT) revealed an 8 x 12 x 23 mm homogenous mass in the right jugular foramen (Figure 1-2), confirmed by magnetic resonance imaging (MRI) to be a glomus jugulare tumor, extending from the jugular bulb, along the lower cranial nerves, invading the carotid canal, and surrounding the vertical segment of the petrous carotid artery. There was no extension into the posterior fossa, but there was extensive involvement of the lower cranial nerves, including IX, X, XI, and XII. The tumor was therefore staged as a Fisch type C2 glomus

jugulare tumor (Figure 3). The magnetic resonance angiography (MRA) revealed tortuous branches of the right external carotid artery feeding the tumor (Figure 4).

Following diagnosis and staging, radiation was recommended, either fractionated radiation therapy or radiosurgery, due to the size and location of the tumor adjacent to the carotid artery and internal jugular vein with high risk of morbidity from surgical intervention. Surgical resection of this tumor would have required, at least a Fisch type A infratemporal fossa approach and would have carried a high risk of lower cranial nerve injury, as well as facial nerve injury, in a patient with no preoperative lower cranial nerve deficits. Due to the small tumor volume, radiosurgery would be an appropriate primary treatment option to control tumor growth and improve clinical outcomes.

DISCUSSION

Glomus jugulare and glomus tympanicum tumors are now grouped into a single category, jugulotympanic tumors. While glomus tympanicum tumors originate in the middle ear space, glomus jugulare tumors originate from the paraganglia tissue around the jugular bulb, particularly the tympanic branch of CN IX (Jacobson's nerve) or auricular branch of CN X (Arnold's nerve) (3). Although only 1-5% of cases are malignant, these tumors are locally invasive and often adjacent to the skull base, jugular vein, carotid artery and cranial nerves [2]. They typically expand within the temporal bone via the path of least resistance, first eroding the jugular fossa, and then the posteroinferior petrous bone [4].

Jugulotympanic tumors are organized by the Fisch Classification system which stratifies tumors based on location and extension to local structures. Type A designates tumors limited to the middle ear, for instance glomus tympanicum tumors. Type B designates tumors limited to the tympanomastoid area. Type C designates tumors invading the infralabyrinthine compartment of the temporal bone. Type D designates tumors with intracranial extension. The Fisch Classification aids in identifying the appropriate surgical approach.

The most common presenting symptoms include pulsatile tinnitus (80%), hearing loss (77%) and aural fullness (70%) [5]. Patients may have dysfunction of cranial nerve 9 through 12 with symptoms such as vertigo, dysphagia, hoarseness, and even facial paresis. A small percentage of glomus tumors are functioning paragangliomas. Although rare, a thorough history should be taken and plasma and/or urine tested for catecholamine breakdown products, including metanephrine, normetanephrine, and vanillylmandelic acid preoperatively [1].

Imaging studies, with the combination of CT and MRI with contrast, are the preferred diagnostic regimen [3]. Angiography is supplementary and helps in treatment planning. It is also useful for pre-operative embolization. These modalities help identify the location, extent and neurovascular involvement of the tumor, which are all important in determining the preferred treatment method.

The ideal treatment method for a GJT is highly debated due to positioning near critical neurovascular structures. Treatment options include observation, preoperative embolization, surgery, radiotherapy or radiosurgery. Surgical approaches consist of post auricular hypotympanotomy or transcanal tympanotomy for smaller tumors restricted to the middle ear, Fisch type A. For larger tumors involving the tympanomastoid area, Fisch type B, atransmastoid resection is needed. Lastly, for tumors with either involvement of the carotid canal or intracranial extension, an extended facial recess with an infratemporal or transtemporal approach is necessary [6, 7]. Although preoperative embolization is controversial, it is commonly used to decrease vascularity leading to less operative time and blood loss [8]. While microsurgery is the definitive treatment, tumors in certain locations are considered high risk for surgical complications with the most common being cranial nerve deficits. Fayad et al. reported a series of 83 patients with jugular foramen tumors in which 81% of the cases achieved gross-total resection of the tumor, however 18.9% of the cases reported new cranial nerve deficits following resection [9]. Other surgical morbidities include cerebrospinal fluid leakage, catastrophic bleeding, residual tumor and mortality. Surgical resection is only recommended in medically operable patients with small tumors and minimal risk of neuropathy. GJT's can be a challenge to manage as surgeons must optimize their surgical approach while minimizing morbidity and mortality.

Radiation can be used as primary or adjunct treatment in patients who are elderly, have increased morbidity, or if the tumor is near significant arteries, nerves, or veins. However, considerable literature exists propagating fractionated external beam radiotherapy and radiosurgery to be used as primary treatment and as a substitute to surgery [10, 11]. The difference between fractionated radiotherapy and radiosurgery revolves around the intensity and duration of treatments. Radiation treatment prevents progression of the tumor by inducing fibrosis of the connective stromal tissue and thrombosis of small vessels [12]. Although a residual mass may remain after radiation, the local control of GJT's is defined as regression or the absence of tumor progression, which studies advocate to be equal to a cure [10, 11].

Fractionated external beam radiotherapy is an established treatment option for GJT's which consists of lower intensity radiation delivered to a wider field over multiple treatments. Hinerman et al. reported local control

with no severe complications in 95% of 121 paraganglioma lesions in the head and neck over a 10.5-year period with fractionated radiotherapy [10]. Local tumor control rates fluctuate from 80% to 100% in other studies using fractionated radiotherapy [13-15]. Although fractionated radiotherapy is a recognized treatment option, the calculated treatment field is always larger than the size of the tumor causing bordering tissue and neurovascular structures to receive a radiation dose [16]. Radiotherapy is given over multiple treatments to decrease the risk of neuropathy and neoplasia and enable healing of normal tissue. However, there is still a 2% risk of neoplasia succeeding fractionated radiotherapy [16-18].

Radiosurgery, particularly gamma-knife radiosurgery (GKS), is a more recent management option which consists of a single-high dose radiation treatment to a small field. GKS has been recognized to successfully treat arteriovenous malformations and other vascular tumors in the brain, providing GJT's to be a suitable candidate for GKS (19). For small volume tumors, several studies show radiosurgery to be less invasive, provide cranial nerve protection, limit complications and control tumor growth in comparison to surgery and radiotherapy [11, 16, 19]. In reported literature, neuropathy seems to be infrequent following GKS at margin doses of 12-15 Gy [11, 16]. Sheehan et al., achieved overall tumor control with GKS in 93% of one hundred thirty-four patients and improvement of pulsatile tinnitus in 49% of patients (16). In concordance, Hafez et al. observed tumor size control with primary GKS in 97.5% of patients at 3 years, 97% at 5 years and 92% at 10 years of follow-up [19]. Lastly, Liscak et al. treated 46 patients with GKS and achieved improvement of neurological deficits in 42% of patients, while worsening deficits in only 4% [20]. Shortcomings of GKS comprise a marginal miss of a tumor in comparison to fractionated radiotherapy. However, there have been no reported cases of neoplasia following GKS [16-18].

Lastly, glomus jugulare tumors have a slow growth rate of 0.8mm/yr and symptoms typically remain stable. Many, especially asymptomatic patients, are choosing to observe these tumors over time with serial imaging [21]. The optimal choice of treatment is not clear, and depends greatly on the patient's demographics, symptoms, comorbidities, as well as the volume and location of the tumor. For patients in which surgery is not feasible, radiotherapy and radiosurgery are safe and effective management options with low morbidity.

CONCLUSION

GJT were once thought to be exceedingly rare, but now are increasing in incidence, perhaps due to the increased availability of CT and MRI. They are slow-growing with subtle symptoms and will often go undiagnosed

for years. Physicians must be aware of this entity in the differential diagnosis of hearing loss, tinnitus, and cranial nerve deficit. Additionally, understanding of various treatment options helps streamline the care pathway to improve patient outcomes and quality of life.

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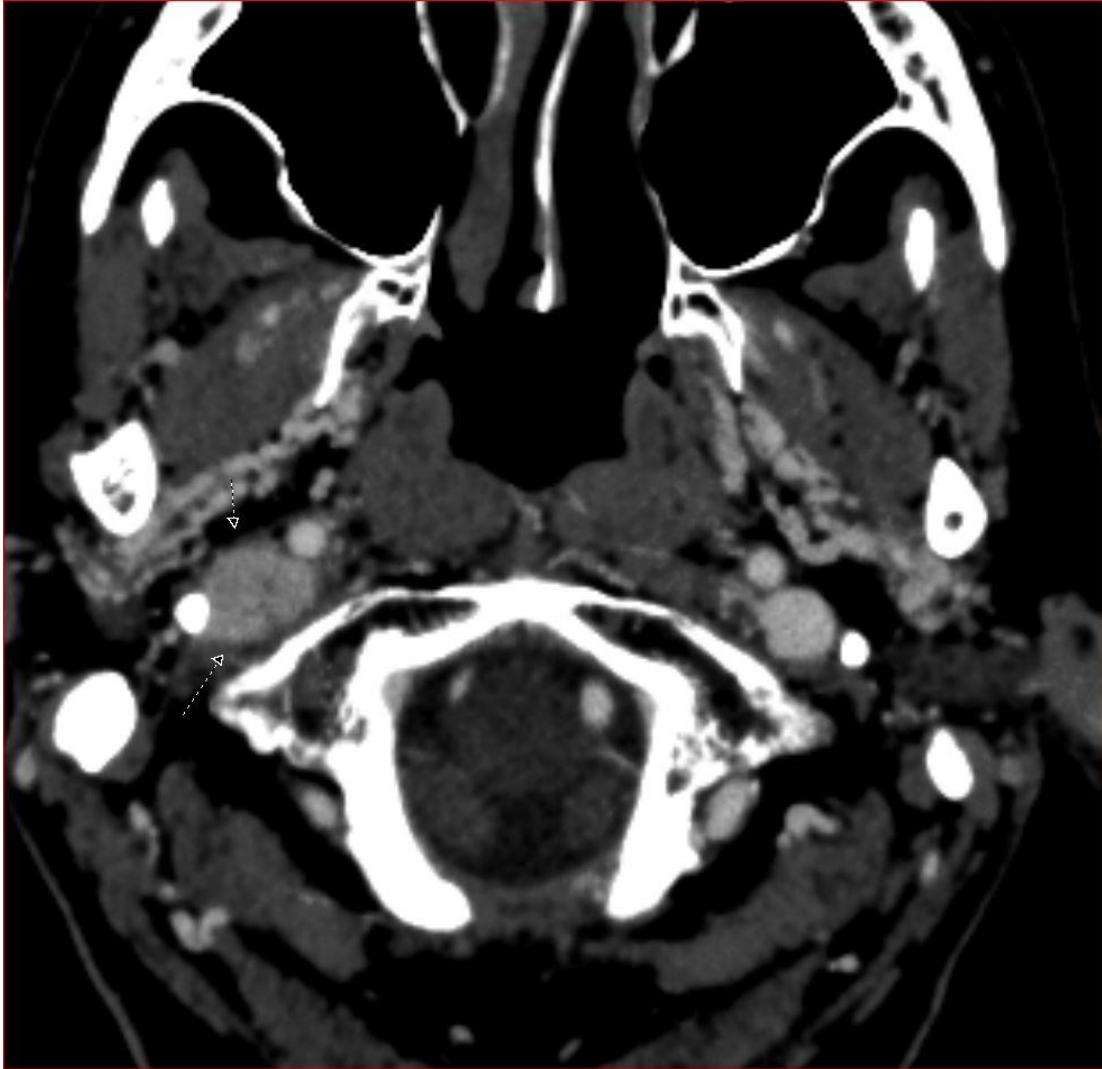


Figure 1: Axial contrast enhanced CT of the neck revealed homogeneously enhancing mass extending inferiorly along the course of the jugular bulb, proximal internal jugular vein (IJV), and inferiorly medial to the styloid process.



Figure 2: Coronal CT temporal bone revealed 8 x 12x23mm (AP x TV x CC) mass in the right jugular fossa with erosions of the mastoid temporal bone, medial wall of tympanic cavity. and jugular fossa. The mass extends into the hypotympanum and laterally along the posterior inferior wall of the osseous external auditory canal.

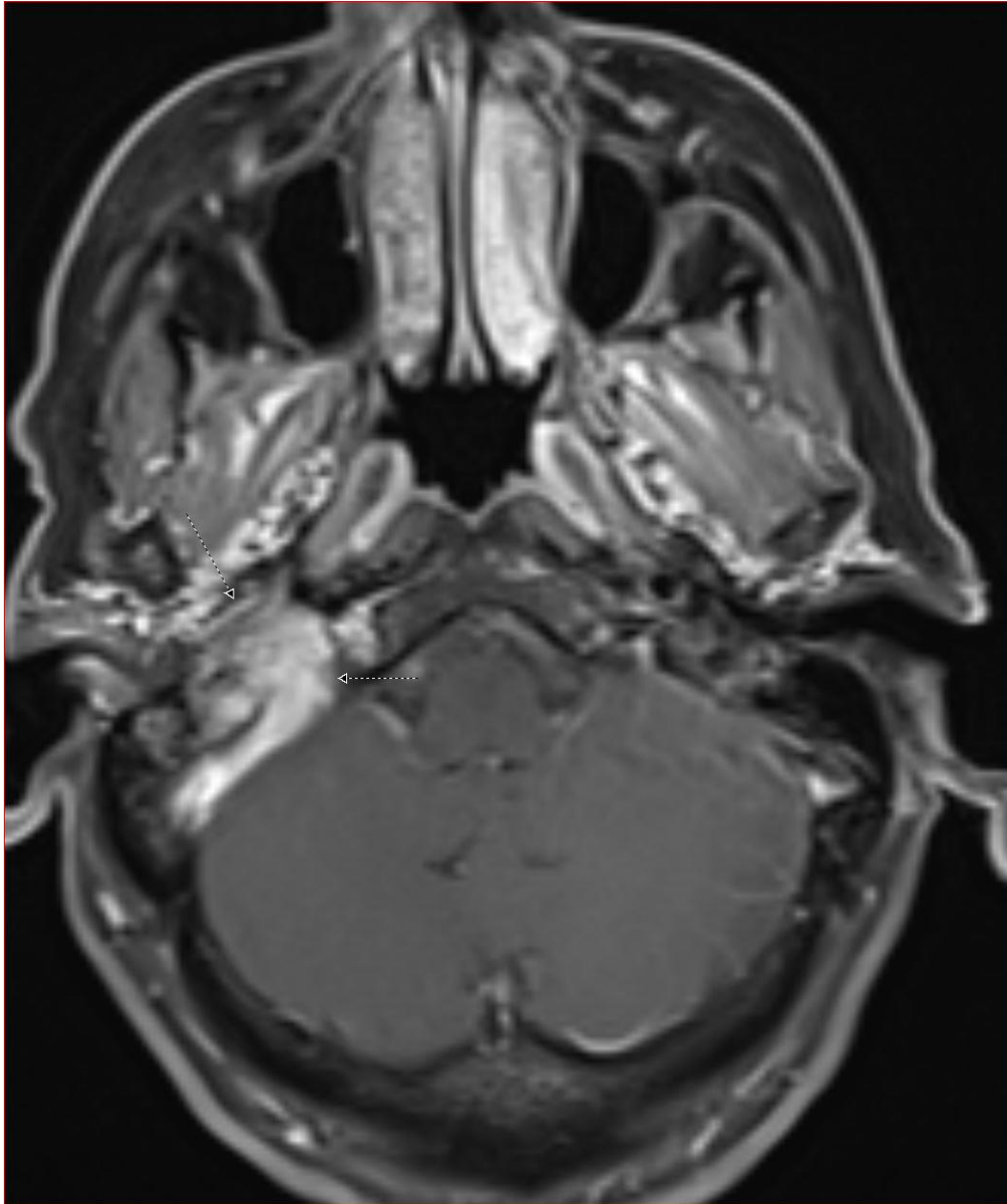


Figure 3: Axial contrast enhanced T1 weighted 3T MRI revealed right jugular fossa enhancing mass extending along the jugular bulb. Adjacent IJV is compressed medially.

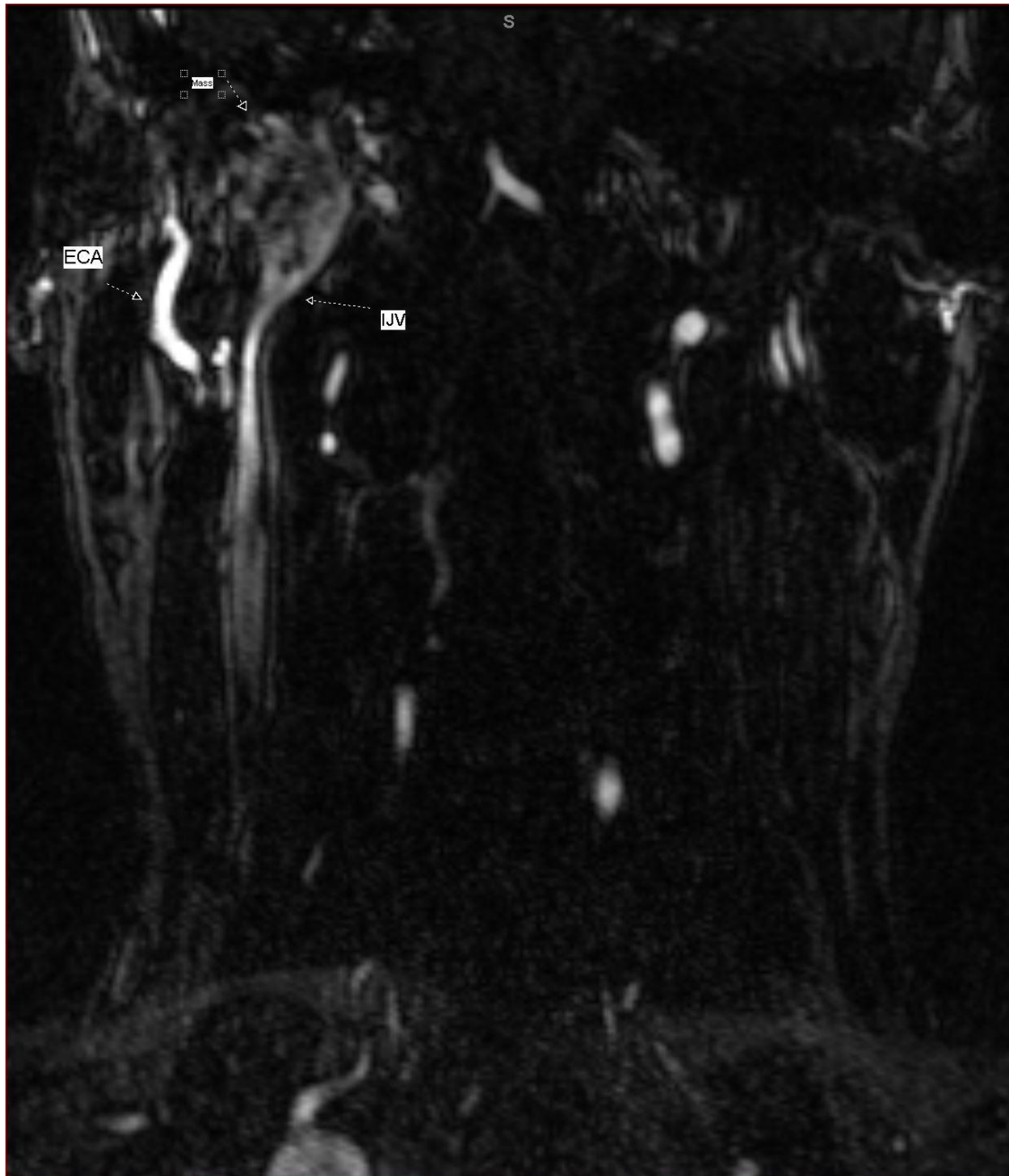


Figure 4: Coronal contrast enhanced neck MRA revealed vascular mass along the right jugular bulb and proximal IJV with arterial feeders from the external carotid artery.