Multidisciplinary Management of Skull Base Paragangliomas: A Case Report

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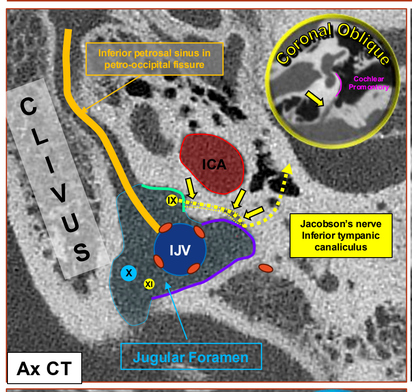
Paragangliomas

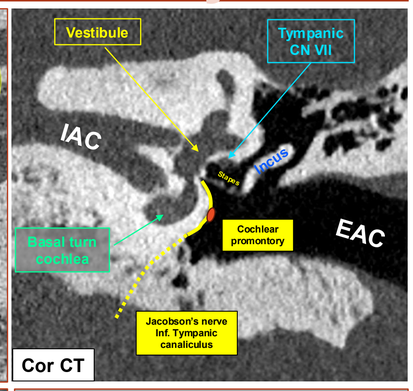
Paragangliomas are benign, highly vascular tumors that arise from paraganglia, which are a group of neuroendocrine cells classified as either sympathetic (chromaffin) or parasympathetic (nonchromaffin. Within the skull base, nonchromaffin parasympathetic paraganglia are found in distinct anatomic locations: the jugular bulb, the inferior tympanic canaliculus (Jacobson's nerve), the cochlear promontory, and the mastoid canaliculus (Arnold's nerve) and descending facial nerve canal. These sites give a clear anatomic roadmap to where paragangliomas occur, known as glomus jugulotympanicum (GJT) when arising in the skull base.

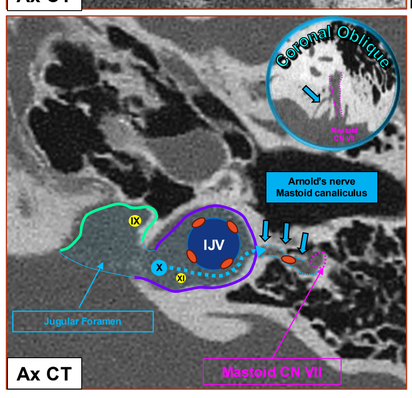
Although benign, GJT can exhibit locally aggressive behavior. Their highly vascular nature, propensity for osseous destruction, and proximity to critical neurovascular structures can complicate management. As a result, the diagnosis and management of GJT often require a coordinated, multidisciplinary approach.

To illustrate these challenges and underscore the importance of collaborative care, we present a rare case of GJT with extensive vascular invasion into the internal jugular vein (IJV).

Normal Anatomy







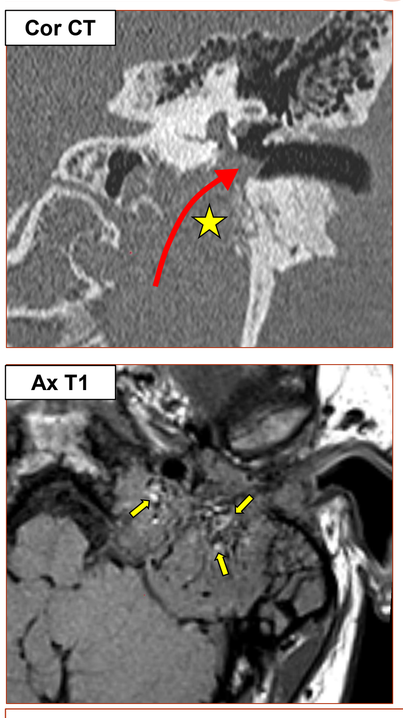
JUGULAR FORAMEN ANATOMY
- Pars Nervosa
  - CN IX
  - Inferior petrosal sinus
    - Drains cavernous sinus
    - Courses thru petro-occipital fissure into jugular bulb
- Pars Vascularis
  - CNs X, XI
  - Internal jugular vein

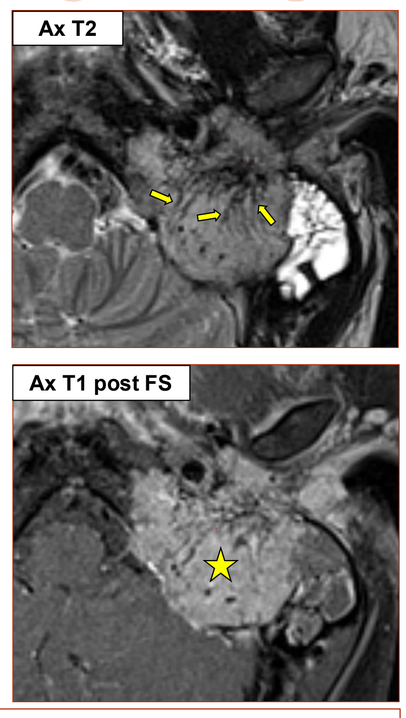
GLOMUS BODY LOCATIONS
- Jugular bulb
  - Around the adventitia

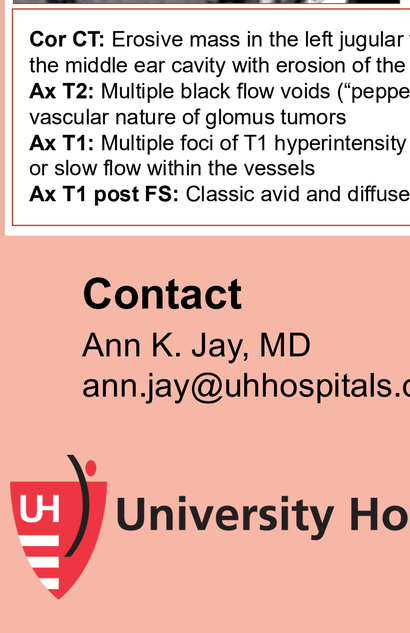
Note the close proximity of the glomus body locations to critical neurovascular structures!!

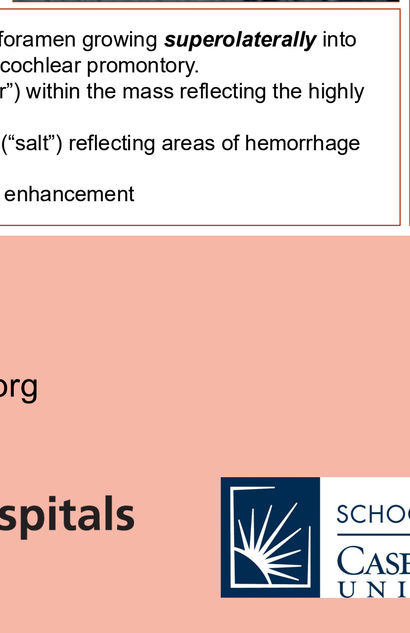
- **Jacobson's nerve** (tympanic br of CN IX)
  - Inferior tympanic canaliculus
    - From pars nervosa, courses between the ICA and jugular foramen
    - Into middle ear cavity to innervate the tympanic plexus on the cochlear promontory
- **Arnold's nerve** (auricular br of CN X)
  - Mastoid canaliculus
    - From pars vascularis to distal mastoid CN VII just superior to the stylomastoid foramen

Classic Imaging Findings



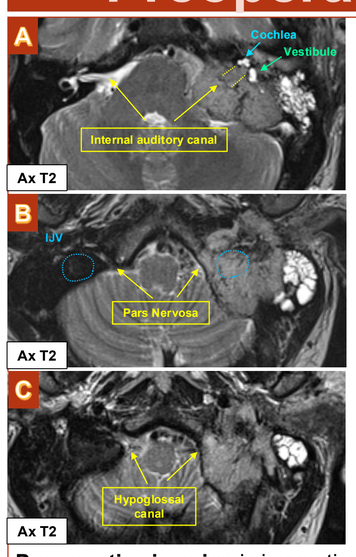


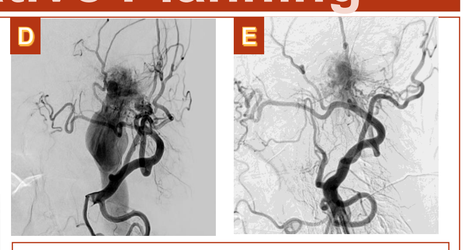




Cor CT: Erosive mass in the left jugular foramen growing ***superolaterally*** into the middle ear cavity with erosion of the cochlear promontory.
Ax T2: Multiple black flow voids ("pepper") within the mass reflecting the highly vascular nature of glomus tumors
Ax T1: Multiple foci of T1 hyperintensity ("salt") reflecting areas of hemorrhage or slow flow within the vessels
Ax T1 post FS: Classic avid and diffuse enhancement

Preoperative Planning





Axial T2 weighted images through the posterior skull base, show the left glomus JT extending into the (A) internal auditory canal and cochlear aperture (CN VII, VIII) (B) both pars nervosa and pars vascularis of the jugular foramen (CNs IX, X, XI, inferior petrosal sinus and IJV) and (C) into the hypoglossal canal (CN XII). (D) Selective left ECA injection shows prominent tumor blush with arterial supply from an enlarged posterior auricular artery and early filling of the IJV. (E) Postembolization angiogram shoes significant reduction in the tumor blush

Preoperative imaging is imperative to guide management. A combination of CT, MRI and conventional angiogram are used to determine the extent of disease, involvement of critical neurovascular structures, and understanding feeding vessels and relative anatomy. Dedicated vascular imaging is obtained to assess the degree of patency of involved dural venous sinuses, IJV, and ICA and for the presence of collateral flow. Given the highly vascular nature of the tumors, pre-operative embolization is often performed to reduce intraoperative blood loss.

Patient selection is critical and involves multiple factors, including age and health of the patient, and the presence and severity of cranial nerve deficits.

Patient counseling is important for setting realistic expectations regarding the possible extent of surgical resection while preserving cranial nerve function and the likelihood of adjuvant radiation therapy.

Staging with FISCH Classification based on extent of disease helps guide surgical approach and management.

TYPE A: Limited to the middle ear
TYPE B: Contained to tympanomastoid area; no infralabyrinthine extension
TYPE C(1-4): Extends beyond the tympanomastoid cavity; Subtypes based on extent of carotid canal involvement
TYPE D(1-2): Intracranial extension <2cm or >2cm in diameter

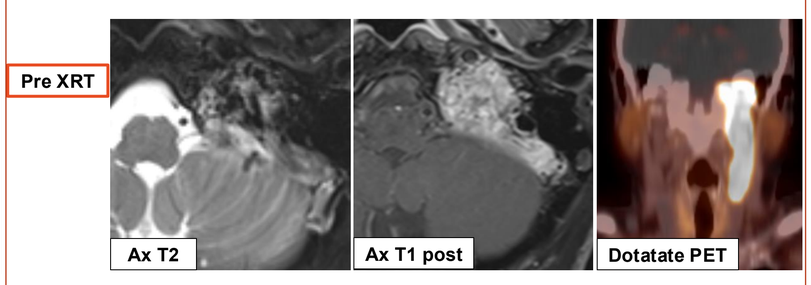
Class A and B tumors are typically managed via standard otologic approaches with high rates of complete resection.

While the Fisch Type A approach remains the surgical gold standard for **Class C and D** tumors, it necessitates facial nerve transposition. Given the high risk of permanent lower cranial nerve deficits, radiotherapy is often preferred, offering comparable local control with a significantly lower complication profile.

Case Presentation

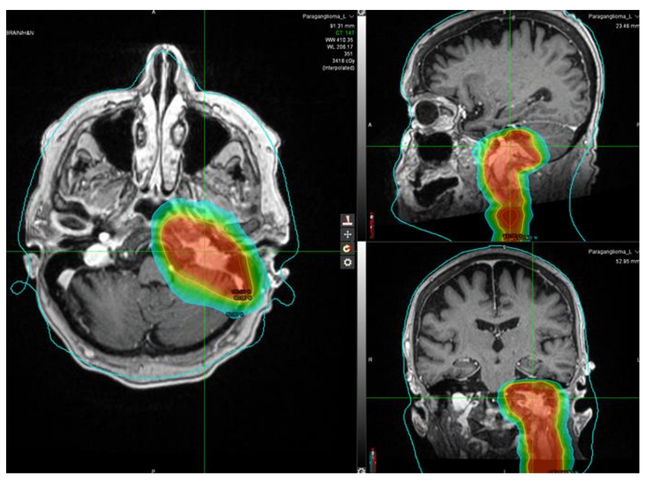
66-year-old female originally diagnosed with a left-sided GJT in 2009 underwent treatment consisting of surgical resection followed by adjuvant Gamma Knife radiosurgery.

In June 2023, the patient presented with new-onset left-sided pulsatile tinnitus and headaches. Diagnostic workup revealed a significant recurrence with invasion into the IJV.



Ax T2: Pre-treatment imaging demonstrates characteristic prominent flow voids (black serpiginous areas) within the mass.
Ax T1 post: Significant enhancement of the recurrent mass with extension into the sigmoid sinus and IJV.
Dotatate PET: Intense radiotracer uptake in the left jugular foramen and IJV, confirming somatostatin receptor-positive tumor recurrence and clarifying the extent of vascular invasion.

The case was reviewed at the multidisciplinary tumor board. While surgical intervention was considered, the team recommended against re-section to prioritize hearing preservation and avoid the morbidity of cranial nerve deficits in a previously irradiated field. Consensus was to proceed with stereotactic body radiation therapy. The patient received SBRT (25 Gy in 5 fractions) in March 2024. Follow-up imaging and clinical evaluation demonstrated a highly favorable response to salvage radiation.



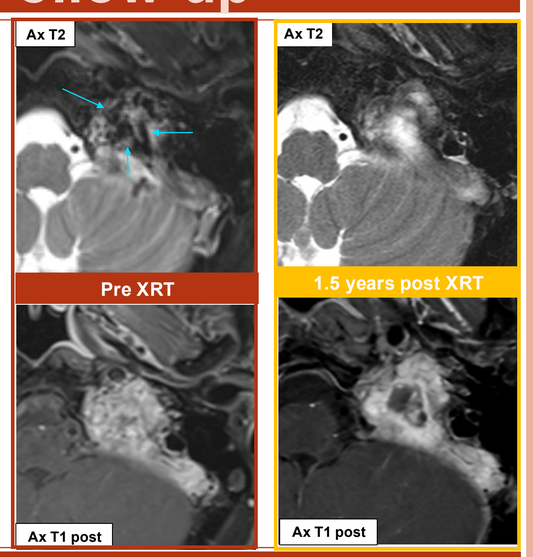
Multiplanar post-contrast T1-weighted MRI demonstrating radiation treatment planning with overlaid dose distribution. The gross tumor volume (GTV) is contoured, with color-wash representing isodose lines illustrating conformal coverage of the target while minimizing dose to adjacent critical structures.

Follow-up

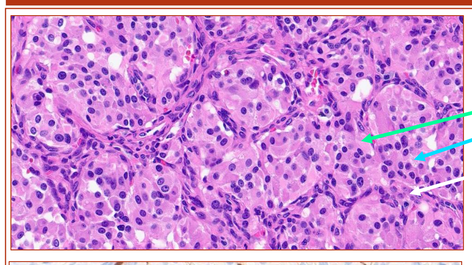
Follow-up MRI 1.5 years after radiation therapy

Ax T2: Resolution of the previously noted flow voids, indicating decreased vascularity within the tumor bed.

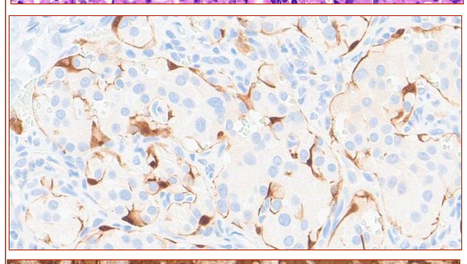
Ax T1 post: New central necrosis within the left jugular foramen consistent with a robust post-radiation therapeutic response



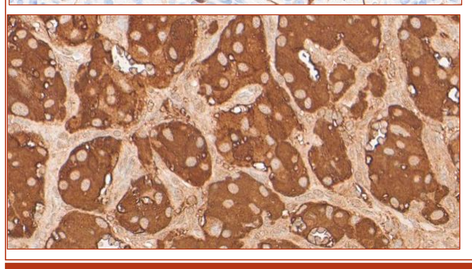
Pathology



H&E STAIN
- Classic ***Zellballen*** pattern
- Epithelioid chief cells
  - Granular cytoplasm (pink)
  - Uniform round to oval nuclei
- Cells form nests or "balls"
- Surrounding fibrovascular stroma or network of thin-walled capillaries



S100 STAIN
Positive for sustentacular cells (brown) surrounding the nests to provide support



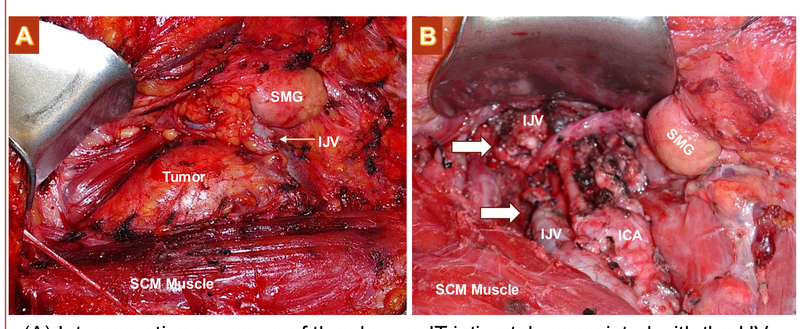
CHROMOGRANIN STAIN
Neuroendocrine marker. Positive staining highlights the formation of nests seen in the Zellballen pattern

Radiation Oncology

- Stereotactic radiosurgery (SRS) and conventional fractionated external beam radiation therapy (EBRT) achieve local control rates of 92-99% at 5-10 years.
- Single-fraction SRS is most effective for tumors <3 cm in maximum diameter, while hypofractionated SRS (3-5 fractions, 18-35Gy) is preferred for larger tumors or those with extensive vascular involvement
- Radiation therapy can be used as primary treatment, adjuvant therapy 8-12 weeks after subtotal resection, or salvage treatment for progressive/recurrent disease

Surgical considerations

- Management of GJT with cervical extension often necessitates a **combined transmastoid and transcervical approach** (e.g., Fisch Type A or B infratemporal fossa approach). This provides the exposure required for proximal and distal vascular control. If tumor invasion necessitates **IJV sacrifice**, cervical dissection must prioritize the identification and mobilization of the carotid artery and vagus nerve prior to vessel ligation.
- The facial nerve can be fully drilled out and transposed to avoid injury while allowing surgical access to the tumor. Vagus nerve preservation is critical as it travels into the neck through the carotid sheath. Other at-risk nerves include CN VIII (hearing), CN IX-XII (lower cranial nerves), and the sympathetic chain.
- IJV sacrifice is generally well tolerated, though patients may experience **facial/neck edema** requiring physical therapy. ICU monitoring is standard.
- Vagal injury results in **voice and swallowing dysfunction** requiring augmentation procedures, swallow therapy, or enteral nutrition. Facial nerve injury necessitates reanimation procedures and protective interventions (eyelid weights). Other complications include Horner syndrome (sympathetic involvement) and hearing loss (CN VIII injury)



(A) Intraoperative exposure of the glomus JT intimately associated with the IJV. (B) Tumor has been resected with IJV sacrifice. Control of vascular structures is imperative during surgical resection.

Conclusion

1. Skull base paragangliomas, specifically GJT tumors, are diagnostic and therapeutic challenges due to their vascularity and location
2. Successful outcomes rely on early integration of neuroradiology, radiation oncology, pathology, ENT, and neurosurgery.

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