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Case Report

Tympanojugular Paraganglioma Treated with Stereotactic Radiotherapy: A Case Report and Literature Review

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Abstract

Paragangliomas are rare neuroendocrine tumors arising from extra-adrenal paraganglionic cells of the autonomic nervous system. In the head and neck region, jugulotympanic paragangliomas are among the most common tumors of the temporal bone. Although typically benign and slow-growing, they may lead to significant morbidity due to local extension.

We report the case of a 62-year-old patient presenting with progressive left-sided hearing loss associated with pulsatile tinnitus. Magnetic resonance imaging revealed a tympanojugular paraganglioma measuring 32 x 13 x 12 mm with extension to adjacent structures. Surgical resection was contraindicated due to the highly vascular nature of the lesion. The patient was treated with fractionated stereotactic radiotherapy (30 Gy in 5 fractions). At three-month follow-up, clinical improvement was observed with minimal residual symptoms and no treatment-related toxicity. Imaging demonstrated a reduction in tumor size and disease stabilization.

Stereotactic radiotherapy represents a safe, effective, and minimally invasive therapeutic option for unresectable tympanojugular paragangliomas, offering favorable tumor control with low morbidity.

Keywords: *Tympanojugular Paraganglioma; Stereotactic Radiotherapy; Fractionated Radiotherapy; Temporal Bone Tumor; Pulsatile Tinnitus; Case Report*

Introduction

Paragangliomas are rare, usually benign neuroendocrine tumors arising from paraganglionic tissue derived from the neural crest. In the head and neck region, they most commonly develop at the level of the carotid body, the vagus nerve, or the jugulotympanic region [1].

Tympanojugular paragangliomas typically present with pulsatile tinnitus and conductive hearing loss. Magnetic resonance imaging (MRI) is the key imaging modality, usually demonstrating a hypervascular lesion with a characteristic salt-and-pepper appearance after contrast administration [1].

The diagnostic workup may include head and neck MRI, functional imaging when indicated, angiographic evaluation in selected cases, and biochemical assessment to rule out functional catecholamine secretion. Tumor extension is commonly described using the Fisch classification, which helps guide therapeutic decision-making [1,2].

Management remains individualized and depends on tumor size, anatomical extension, symptoms, patient comorbidities, and the expected morbidity of treatment. Although complete surgical excision, with or without preoperative embolization, has long been considered a standard approach, stereotactic radiotherapy is increasingly recognized as a less invasive alternative offering durable tumor control with low morbidity in selected patients [3].

This case report illustrates the role of fractionated stereotactic radiotherapy in the management of an unresectable tympanojugular paraganglioma and discusses relevant data from the literature.

Case Presentation

A 62-year-old patient with a history of well-controlled type 2 diabetes mellitus, treated with oral antidiabetic agents for four years, presented with progressive left-sided hearing loss and pulsatile tinnitus evolving over two years, with recent worsening. Otoscopic examination revealed a polypoid lesion arising from the middle ear and protruding into the external auditory canal.

A biopsy of the left middle ear was performed on 08/08/2024 because the initial otoscopic appearance suggested a polypoid external auditory canal/middle ear lesion. Macroscopic examination showed a small grayish-white friable tissue fragment measuring approximately 2 mm. Histological analysis demonstrated a cystic lesion lined by well-differentiated keratinizing squamous epithelium with keratin pearl formation, consistent with a keratinizing epithelial lesion of cholesteatoma type. No evidence of neuroendocrine tumor proliferation, Zellballen architecture, or features suggestive of paraganglioma was identified (Figure 1). However, subsequent MRI findings and intraoperative exploration were highly suggestive of a hypervascular tympanojugular paraganglioma, explaining why definitive surgical excision was not pursued.

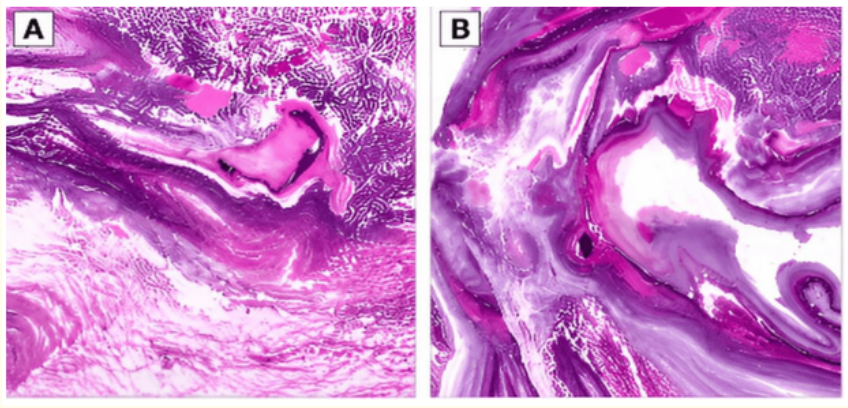

Figure 1: *Histology showing keratinizing squamous epithelium with keratin pearls.*

(A) Histological section demonstrating a cystic lesion lined by keratinizing squamous epithelium.

(B) Higher magnification showing keratin pearl formation within the epithelial component.

Brain and temporal bone MRI performed on 22/08/2024 revealed an expansive lesion measuring 32 x 13 x 12 mm centered on the tympanic cavity and left external auditory canal, suggestive of a tympanojugular paraganglioma. The lesion completely filled the tympanic cavity and extended laterally into the external auditory canal, which appeared widened. Medially, it extended toward the Eustachian tube with associated mastoid air cell opacification, while the inner ear structures were preserved and the lesion remained distant from

the carotid canal. Inferiorly and posteriorly, it reached the jugular bulb. Superiorly, bone lysis of the temporal bone was observed with extension into the mandibular fossa, causing anterior displacement of the mandibular condyle. No intradural or intracranial extension was identified (Figure 2).

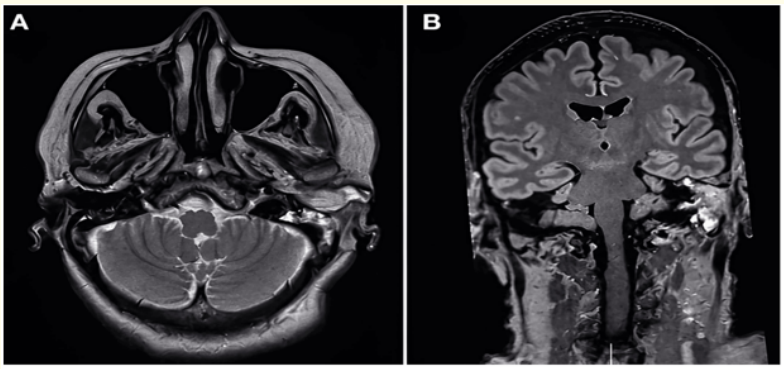

Figure 2: *Axial and coronal MRI showing a hyperintense lesion with contrast enhancement involving the tympanojugular region.*

(A) Axial T1-weighted post-contrast image demonstrating an enhancing lesion in the tympanojugular region.

(B) Coronal T1-weighted post-contrast image showing the extent of the lesion and its anatomical relationships.

Intraoperative exploration confirmed the highly vascular nature of the lesion, rendering surgical excision contraindicated. The patient was therefore referred for fractionated stereotactic radiotherapy. Treatment consisted of a total dose of 30 Gy delivered in five fractions of 6 Gy on alternate days. Radiotherapy was delivered between October 30, 2024, and November 11, 2024, following CT-based simulation. The gross tumor volume (GTV) corresponded to the visible tumor on imaging, expanded by a 5-mm margin to define the planning target volume (PTV). Organs at risk included the cochlea, brainstem, optic apparatus, temporomandibular joint, spinal cord, oral cavity, and larynx. Dosimetric planning demonstrated optimal conformality with adequate tumor coverage and preservation of adjacent critical structures (Figure 3).

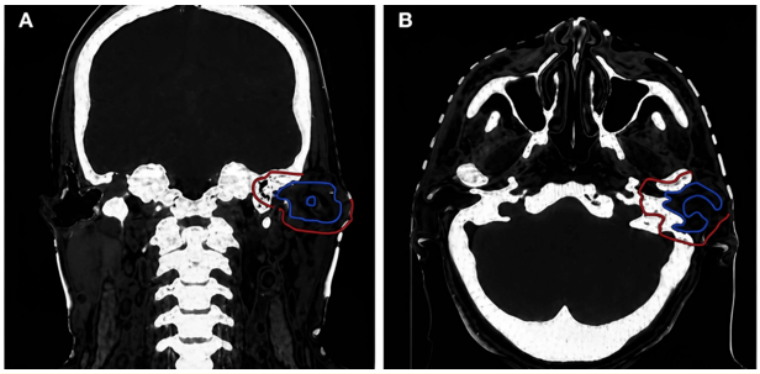

Figure 3: *Treatment planning showing the GTV in blue and the PTV in red.*

(A) Coronal view showing the delineation of the gross tumor volume (GTV) in blue.

(B) Axial view showing the planning target volume (PTV) in red encompassing the GTV with appropriate margins.

The stereotactic dosimetry of this paraganglioma demonstrates highly precise treatment planning aimed at optimizing tumor control while minimizing toxicity to the surrounding healthy tissues. The axial, coronal, and sagittal sections show excellent isodose conformity around the target volume, with concentration of the maximum dose within the tumor. The rapid fall-off of isodoses at the periphery reflects good sparing of adjacent critical structures, particularly the neighboring neurovascular elements. Analysis of the dose-volume histogram (DVH) confirms adequate coverage of the tumor volume by the prescribed dose while respecting the tolerance constraints of the organs at risk. This homogeneous and conformal dose distribution highlights the value of stereotactic radiotherapy in the management of paragangliomas, allowing effective, targeted, and safe irradiation (Figure 4).

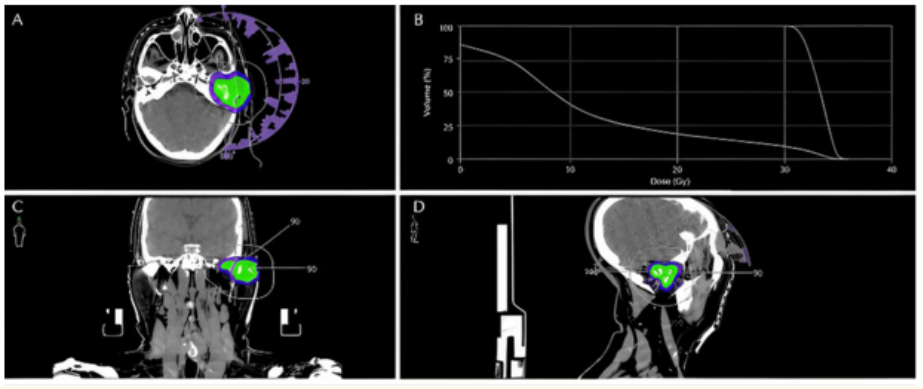

Figure 4: *Dosimetric planning of stereotactic radiotherapy for a cranial paraganglioma.*

(A) Axial CT slice showing the target volume delineation with dose distribution overlay.

(B) Dose-volume histogram (DVH) illustrating target coverage and dose fall-off to surrounding structures.

(C) Coronal CT reconstruction demonstrating the spatial relationship of the lesion to adjacent anatomical structures with isodose lines.

(D) Sagittal CT reconstruction showing the three-dimensional conformal dose distribution and target coverage.

At three months post-treatment, the patient was in good general condition with an ECOG performance status of 0. Clinically, only mild intermittent tinnitus persisted without radiotherapy-related toxicity. MRI of the temporal bone and brain performed on 12/02/2025 showed a stable appearance of the soft tissue lesion of the left petrous pyramid, centered on the tympanic cavity and the external auditory canal, measuring 22 x 9 x 12 mm (T x AP x H). The tumor remained confined to the tympanic cavity. Laterally, it extended into the external auditory canal, which appeared widened compared to the contralateral side. Medially, it extended toward the Eustachian tube, associated with opacification of the mastoid air cells; the inner ear structures were preserved, and the lesion remained distant from the carotid canal. Superiorly, it caused lysis of the tympanic bone and extended into the mandibular fossa, resulting in anterior dislocation of the mandibular condyle. Inferiorly and posteriorly, it reached the jugular bulb. No cervical lymphadenopathy was observed (Figure 5).

At 12 months follow-up, the patient maintained excellent clinical status (ECOG 0), with marked improvement of pulsatile tinnitus, now intermittent and minimally symptomatic. Hearing loss remained stable without deterioration, and no new cranial nerve deficits were reported. No late radiation-induced toxicity, including radionecrosis or cochlear damage, was observed.

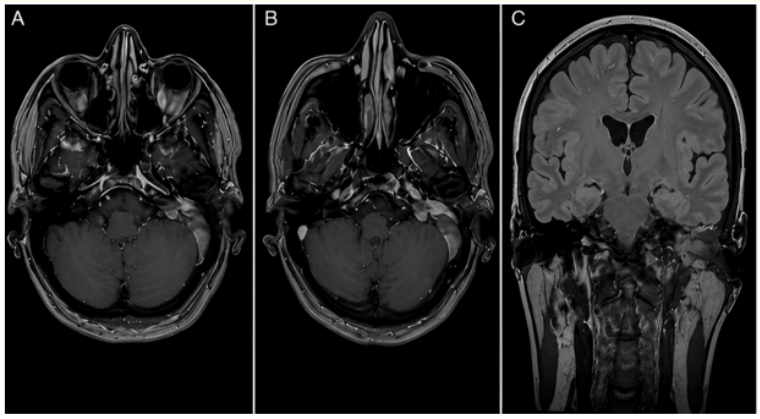

Figure 5: *Brain and temporal bone MRI (12/02/2025) showing a stable soft tissue lesion of the left petrous pyramid with locoregional extension.*

(A) Axial contrast-enhanced T1-weighted MRI demonstrating a lesion involving the left petrous pyramid.

(B) Axial contrast-enhanced T1-weighted MRI further delineating the extent of the lesion without significant progression.

(C) Coronal FLAIR MRI showing locoregional extension of the lesion with no evidence of interval progression compared with prior imaging.

Radiological evaluation at 12 months was performed as part of routine surveillance. Although the original imaging studies were not available for direct review, the official radiological report was retrieved from the medical record and confirmed overall disease stability, with the lesion demonstrating global stability and a slight volumetric reduction compared with both the initial imaging and the 3-month follow-up, measuring 22 x 9 x 12 mm initially and decreasing to approximately 20 x 8 x 10 mm. No locoregional progression or intracranial extension was observed. Post-contrast MRI sequences showed persistent heterogeneous enhancement with a slight reduction in hypervascularity and attenuation of the typical salt-and-pepper appearance. The lesion remained stable in all compartments, including the tympanic cavity, external auditory canal, Eustachian tube, mastoid cells, jugular bulb, and mandibular fossa, without invasion of the carotid canal or adjacent neurovascular structures. No cervical lymphadenopathy or post-radiotherapy complications such as edema or radionecrosis were identified.

Discussion

Paraganglia are composed of neuroendocrine cells distributed along the major vascular axes of the head and neck, as well as along the thoracic and lumbar spine. Embryologically, these cells derive from the neural crest and are associated with the autonomic nervous system - parasympathetic in the cervicofacial region and sympathetic in the thoracoabdominal region [4]. The adrenal medulla contains the largest concentration of paraganglia, which regulate homeostasis through catecholamine secretion and may give rise to pheochromocytomas [4].

Temporal bone paraganglia are located on the promontory of the middle ear along Jacobson's nerve and within the adventitia of the jugular bulb. They are closely related to the ninth and tenth cranial nerves and are supplied by branches of the ascending pharyngeal artery.

In 1840-1841, Valentin described a cluster of highly vascularized cells surrounding Jacobson's nerve, which he termed the glomus tympanicum [4]. A century later, in 1941, Guild described paraganglia within the adventitia of the jugular bulb (glomus jugulare), reporting

an average of three paraganglia per temporal bone across 88 specimens [5]. In 1945, Rosenwasser provided the first description of a paraganglioma of the middle ear [6]. The distinction between tympanic and jugular origins of temporal bone paragangliomas was later clarified by Alford and Guilford [7].

Paragangliomas, the tumors arising from these paraganglia, typically appear as well-defined reddish or brownish masses of firm to elastic consistency. They are highly vascular, pulsatile, and prone to bleeding upon contact. Microscopically, they display a uniform architecture composed of Zellballen clusters of chief (type I) cells containing neurosecretory granules, surrounded by elongated sustentacular (type II) cells and fibrovascular trabeculae [8].

Paragangliomas are part of the diffuse neuroendocrine system. Excluding pheochromocytomas, only 1 - 3% of cases are functionally active [9,10], secreting primarily catecholamines. In the presence of tachycardia, headache, or excessive sweating, measurement of plasma catecholamines and urinary metabolites is indicated. Rarely, paragangliomas secrete serotonin, leading to a carcinoid syndrome.

The incidence of symptomatic paragangliomas, including pheochromocytomas, is estimated at 1 in 30,000 in the Caucasian population [10]. Cervicofacial paragangliomas account for approximately 0.6% of all head and neck tumors [10] and only 0.3% of all paragangliomas [11]. Jugular and tympanic paragangliomas are the most frequent tumors of the middle ear [12], representing 18 - 36% of head and neck paragangliomas; 60 - 80% are carotid body tumors, and 3 - 4% arise from the vagus nerve [12,13]. Exceptionally, paragangliomas may occur in the nasopharynx, nasal cavities, paranasal sinuses, larynx, thyroid gland, or orbit [14].

Most cervicofacial paragangliomas are benign and slow-growing, although they may exhibit locally aggressive behavior, invading bone and surrounding soft tissues. Malignant transformation has been observed in fewer than 5% of jugulotympanic and carotid paragangliomas, and in up to 20% of vagal paragangliomas [15]. The criteria for malignancy remain debated. According to Lack, the diagnosis requires at least two of the following three features: central necrosis, vascular or lymphatic invasion, and atypical mitoses [16]. Other authors, however, consider malignancy proven only in the presence of distant metastases [17].

Jugular and tympanic paragangliomas usually present with similar clinical features, although symptom onset tends to occur later in jugular forms [18]. The most frequent presenting symptom is pulsatile tinnitus, reported in about 80% of patients. Hearing loss, conductive or sensorineural depending on tumor extension, is found in approximately 60% of cases [18]. Involvement of cranial nerves, particularly the IXth to XIIth pairs, or the sympathetic trunk, may result in dysphagia, shoulder weakness, or Horner's syndrome. Facial nerve paresis generally appears at advanced stages.

When symptoms suggest a secretory form, measurement of 24-hour urinary catecholamine metabolites (metanephrine or vanillylmandelic acid) is essential. The presence of carcinoid syndrome warrants assessment of serotonin and its metabolites in blood and urine.

Otoscopy typically reveals a reddish retrotympanic mass. The pulsatile nature of the tumor is not always apparent. Occasionally, auscultation may detect a bruit synchronous with the pulse. The presence of bony erosion of the deep portion of the external auditory canal suggests a jugular origin of the lesion. Because of the tumor's marked hypervascularity, biopsy is strongly contraindicated [17].

Computed tomography (CT) and magnetic resonance imaging (MRI) are the imaging modalities of choice. MRI typically shows a salt-and-pepper appearance with intense enhancement after gadolinium injection [1,17]. MRI also evaluates intracranial extension, while MR angiography identifies feeding vessels. Conventional angiography has limited indications today but may be used for preoperative embolization or carotid occlusion testing [19].

Scintigraphy using octreotide or MIBG is useful for detecting multifocal disease [20]. The most commonly used classification is the Fisch classification [21] (Table 1).

Type	Description
Type A	Tumors originating along the tympanic plexus on the promontory; limited to the middle ear.
Type B	Tumors originating in the tympanic canal, extending into the hypotympanum and mastoid.
Type C	Tumors originating in the dome of the jugular bulb, invading the petrous bone and pyramid. C1-C4 are defined according to erosion of the carotid canal between the carotid foramen and the cavernous sinus.
Type D	Tumors with intracranial extension into the posterior fossa; subdivided as extradural (De1-De2) or intradural (Di1-Di3) according to depth of invasion.

Table 1: *Classification of temporal bone paragangliomas (Fisch classification).*

Table adapted and reformulated from the Fisch classification described in previous publications [21].

Conclusion

Tympanojugular paragangliomas are rare, slow-growing, and usually benign tumors, but their complex skull base location and marked vascularity can make surgical management difficult and potentially morbid. This case highlights the value of fractionated stereotactic radiotherapy as a safe and effective therapeutic option for unresectable or high-risk tympanojugular paragangliomas. Treatment with 30 Gy in five fractions achieved clinical improvement, radiological stability with slight volumetric reduction, and no treatment-related toxicity during follow-up. Careful multidisciplinary evaluation remains essential to individualize treatment and balance tumor control against functional morbidity.

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

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Drafting of the manuscript: Lahlou Imane, Chakib Fatima Zahra, Karima Nouni, Amine Lachgar, Hanane Elkacemi, Tayeb Kebdani, Khalid Hassouni.

Critical review of the manuscript for important intellectual content: Lahlou Imane, Chakib Fatima Zahra, Karima Nouni, Amine Lachgar, Hanane Elkacemi, Tayeb Kebdani, Khalid Hassouni.

Supervision: Lahlou Imane, Chakib Fatima Zahra, Karima Nouni, Amine Lachgar, Hanane Elkacemi, Tayeb Kebdani, Khalid Hassouni.

Human Subjects

Informed consent for treatment and open access publication was obtained or waived by all participants in this study.

Conflicts of Interest

In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work.

Financial Relationships

All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

Other Relationships

All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

Bibliography

1. Williams MD. "Paragangliomas of the head and neck: an overview from diagnosis to genetics". *Head and Neck Pathology* 11 (2017): 278-287.
2. Fisch U. "Infratemporal fossa approach for glomus tumors of the temporal bone". *Annals of Otology, Rhinology, and Laryngology* 91 (1982): 474-479.
3. Offergeld C., *et al*. "Head and neck paragangliomas: clinical and molecular genetic classification". *Clinics (Sao Paulo)* 67.1 (2012): 19-28.
4. Valentin G. "Ueber eine gangliöse Anschwellung im Bereiche des Nervus glossopharyngeus und seiner Äste". Archiv für Anatomie, Physiologie und wissenschaftliche Medicin (1841): 287-290.
5. Guild SR. "The glomus jugulare, a nonchromaffin paraganglion, in man". *Annals of Otology, Rhinology, and Laryngology* 62.4 (1953): 1045-1071.
6. Rosenwasser H. "Carotid body tumor of the middle ear and mastoid". *Archives of Otolaryngology* 41.1 (1945): 64-67.
7. Alford BR and Guilford FR. "A comprehensive study of tumors of the glomus jugulare". *Laryngoscope* 72.6 (1962): 765-805.
8. Lack EE. "Tumors of the adrenal gland and extra-adrenal paraganglia. Atlas of tumor pathology, 3rd Series, Fascicle 19". Armed Forces Institute of Pathology, Washington, DC (1997).
9. Borba LA and Al-Mefty O. "Intravagal paragangliomas: report of four cases". *Neurosurgery* 38.3 (1996): 569-575.
10. Stewart KL. "Paragangliomas of the temporal bone". *American Journal of Otolaryngology* 14.4 (1993): 219-226.
11. van Baars FM., *et al*. "Familial non-chromaffin paragangliomas (glomus tumors): clinical and genetic aspects". *Acta Oto-Laryngologica* 91.1-6 (1981): 589-593.
12. Pellitteri PK., *et al*. "Paragangliomas of the head and neck". *Oral Oncology* 40.6 (2004): 563-575.
13. Manolidis S., *et al*. "Malignant glomus tumors". *Laryngoscope* 109.1 (1999): 30-34.
14. Lack EE., *et al*. "Paragangliomas of the head and neck region: a pathologic study of tumors from 71 patients". *Human Pathology* 10.2 (1979): 191-218.
15. Lee JH., *et al*. "National cancer data base report on malignant paragangliomas of the head and neck". *Cancer* 94.3 (2002): 730-737.
16. Boedeker CC., *et al*. "Paragangliomas of the head and neck: diagnosis and treatment". *Familial Cancer* 4 (2005): 55-59.

17. Remley KB., *et al*. "Pulsatile tinnitus and the vascular tympanic membrane: CT, MR, and angiographic findings". *Radiology* 174.2 (1990): 383-389.

18. Forest JA 3rd., *et al*. "Long-term control of surgically treated glomus tympanicum tumors". *Otology and Neurotology* 22.2 (2001): 232-236.

19. Gerosa M., *et al*. "Glomus jugulare tumors: the option of gamma knife radiosurgery". *Neurosurgery* 59.3 (2006): 561-569.

20. Shulkin BL., *et al*. "Pheochromocytomas and paragangliomas: imaging with 111In-pentetreotide". *Radiology* 188 (1993): 35-40.

21. Fisch U and Mattox DE. "Microsurgery of the skull base". Thieme Medical Publishers, New York (1988).

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