

# **Tympanic Paraganglioma (Fisch Type B) Mimicking a Tympanojugular Tumor: Intraoperative Reclassification Revealing Jugular Bulb Dehiscence - A Case Report**

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## **A B S T R A C T**

**Background:** Tympanic paragangliomas are the most common benign tumors of the middle ear. Despite advances in temporal bone imaging, preoperative staging can be misleading when anatomical variants of the jugular fossa coexist with the tumor, leading to overclassification and unnecessarily aggressive surgical planning.

**Case presentation:** We report a 48-year-old woman with a three-year history of progressive left-sided conductive hearing loss and intermittent pulsatile tinnitus. Otoscopy revealed a pulsatile red retrotympanic mass with a positive Brown's sign. Audiometry showed a moderate-to-severe conductive hearing loss (air-bone gap ~60–70 dB). Both clinical findings and multimodal imaging - MRI and angio-CT showing a strongly enhancing mass with erosion of the anterior jugular foramen wall - converged toward a Fisch type C tympanojugular paraganglioma. Intraoperative exploration, however, revealed hypotympanic extension without jugular infiltration. A jugular bulb dehiscence, responsible for the radiological overclassification, was identified and preserved. The lesion was reclassified intraoperatively as Fisch type B. Complete resection was achieved without complication.

**Conclusion:** Jugular bulb dehiscence can simulate jugular invasion on CT even when clinical and imaging data jointly suggest a tympanojugular lesion. Intraoperative confirmation of tumor staging is essential before committing to a high-morbidity infratemporal fossa approach.

**Keywords:** Tympanic paraganglioma; Glomus tympanicum; Fisch type B; Jugular bulb dehiscence; Intraoperative reclassification; Preoperative imaging discordance; Pulsatile tinnitus

## **Introduction**

Paragangliomas of the middle ear, historically referred to as glomus tumors, are highly vascularized benign neoplasms arising from paraganglion cells associated with the tympanic

branch of the glossopharyngeal nerve (Jacobson's nerve, CN IX) or the auricular branch of the vagus nerve (Arnold's nerve, CN X)<sup>1,2</sup>. They represent the most common primary neoplasm of the middle ear and the second most frequent tumor of the

temporal bone overall<sup>3</sup>. Their classic presentation consists of pulsatile tinnitus, progressive conductive hearing loss, and a reddish pulsatile retrotympenic mass on otoscopy<sup>4</sup>.

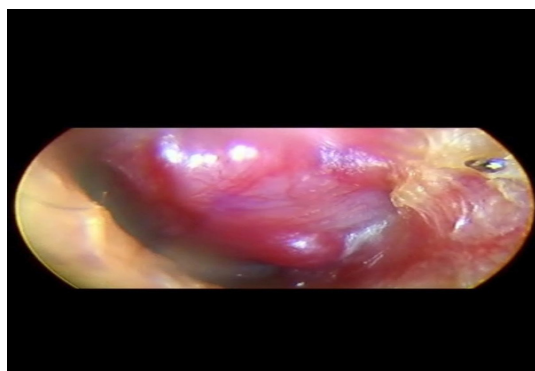
Preoperative staging follows the Fisch and Mattox classification<sup>5</sup>: type A tumors are confined to the middle ear; type B involve the hypotympanum without jugular extension; types C and D extend into the jugular canal, carotid canal, and potentially the intracranial compartment. This classification directly dictates surgical strategy - type A/B lesions are approached via transmeatal tympanotomy, while type C/D lesions require infra-temporal fossa approaches carrying substantially greater risks of facial nerve palsy, lower cranial nerve deficits, and major vascular complications<sup>6</sup>.

Anatomical variants of the jugular fossa - particularly jugular bulb dehiscence - can generate CT appearances indistinguishable from jugular invasion, leading to tumor overclassification<sup>7</sup>. We report a case in which both clinical presentation and multi-modal imaging converged toward a Fisch type C tympanojugular paraganglioma, while intraoperative exploration established the correct diagnosis of a Fisch type B tympanic paraganglioma associated with jugular bulb dehiscence, fundamentally altering the surgical strategy.

## Case Report

### Clinical history

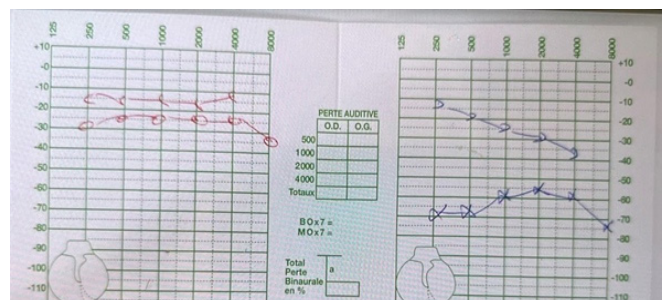
A 48-year-old woman presented with a three-year history of progressive left-sided hearing loss associated with intermittent pulsatile tinnitus. She denied vertigo, dysphagia, dysphonia, facial palsy, or neurological deficits. Her past medical history was notable for a laparoscopic cholecystectomy performed in 2022, with no familial history of paraganglioma. Otoscopic examination of the right ear was unremarkable, while the left ear demonstrated a pulsatile dark red mass bulging behind an intact tympanic membrane, confined to the posteroinferior quadrant - a presentation consistent with the classical “sunrise sign” of a retrotympenic vascular lesion (**Figure 1**). Brown’s sign was positive. No facial nerve palsy, spontaneous nystagmus, or cervical lymphadenopathy was noted.



**Figure 1:** Left ear otoscopy showing a dark red pulsatile mass occupying the posteroinferior quadrant -the sunrise sign of tympanic paraganglioma

Audiological assessment revealed a mild hearing loss in the right ear on pure-tone audiometry, with an average threshold of around 20 dB HL (**Figure 2**). The left ear exhibited a moderate-to-severe conductive hearing loss, characterized by an air-bone gap of approximately 60–70 dB HL with preserved bone conduction thresholds. Tuning fork tests were consistent with a

left-sided conductive mechanism, as Weber’s test lateralized to the left and Rinne’s test was negative on the left side.



**Figure 2:** Pure-tone audiogram showing mild right-sided hearing loss (~20 dB) and moderate-to-severe left conductive hearing loss with a 60–70 dB air-bone gap and preserved bone conduction. Weber lateralized to the left

Preoperative MRI of the temporal bones, including axial T1, T2/FLAIR, diffusion-weighted, submillimetric 3D T2 SPACE, and 3D T1 post-gadolinium sequences, revealed a soft-tissue mass occupying nearly the entire left tympanic cavity, measuring approximately 15 mm in height. The lesion demonstrated mild T1 hypersignal, T2 hyposignal, weak gadolinium enhancement, and no restricted diffusion. Ossicular erosion involving the malleus, incus, tegmen tympani, and the second portion of the facial canal was noted. No jugulo-carotid anomaly or intracranial extension was identified. These imaging findings were suggestive of an intratympanic schwannoma or a granulomatous/cholesteatomatous lesion, with histological correlation recommended for definitive diagnosis (**Figure 3**).



**Figure 3:** MRI of the temporal bones. The intratympanic mass demonstrates weak enhancement without the classic ‘salt-and-pepper’ pattern, initially orienting the diagnosis toward a lesion of tumoral origin

Angio-CT of the temporal bones and cervical vessels, performed using volumetric helical acquisition with 0.6-mm collimation and MIP and 3D reconstructions, demonstrated a 15 × 6.5 mm mass with intense arterial-phase enhancement. Key findings included erosion of the anterior wall of the jugular

foramen associated with a slightly prominent jugular bulb, ossicular chain erosion, involvement of the second portion of the facial canal, and mastoid opacification. Notably, the absence of a bony septum between the tumor and the jugular bulb, combined with anterior jugular foramen erosion, raised radiological concern for jugular involvement. These findings were consistent with a strongly enhancing mass, favouring a schwannoma or glomus tumor with possible tympanojugal extension (**Figures 4A-4B**).



**Figure 4A:** High-resolution CT bone window. The mass occupies the meso-hypotympanum. Note the absence of a bony plate between the tumor and the jugular bulb



**Figure 4B:** Angio-CT coronal reformat. The strongly enhancing mass (1.46 × 0.65 cm) is directly contiguous with the jugular bulb, erroneously suggesting jugular invasion

Intraoperatively, a dark red, highly vascularized, and pulsatile mass was identified in the meso- and hypotympanum, adherent to the cochlear promontory at Jacobson's nerve. Systematic exploration confirmed hypotympanic extension without transgression of the tympanic floor or jugular infiltration. Careful exploration of the hypotympanic floor revealed a complete jugular bulb dehiscence: the bony plate separating the tympanic cavity from the superior wall of the jugular bulb was entirely absent, leaving the jugular bulb directly exposed and adjacent to the tumor, yet clearly uninvaded. This finding led to an intraoperative reclassification as Fisch type B, with the dehiscence identified as the sole explanation for the jugular foramen erosion erroneously suggested on preoperative imaging. The jugular bulb wall was preserved throughout the procedure. The tumor was dissected en bloc, achieving complete macroscopic resection without vascular complication. Pathological analysis of the surgical specimen confirmed a paraganglioma (glomus tympanicum) displaying the classic zellballen architecture. No malignant features were identified. The final diagnosis was a benign tympanic paraganglioma, classified as Fisch type B. The postoperative course was uneventful and free of complications. The patient was discharged on postoperative day two. At the one-month follow-up, examination revealed a well-healed tympanic membrane with no residual mass, along with a marked improvement in tinnitus and audiometric recovery

of the conductive component. Facial nerve function remained intact throughout. No lower cranial nerve deficit or vascular complication was observed.

## Discussion

Glomus tympanicum tumors predominantly affect females (4:1 ratio), with a peak incidence at 50-60 years<sup>8</sup>. The most common complaints are pulsatile tinnitus, conductive hearing loss, and aural fullness<sup>9</sup>. A positive Brown's sign confirms direct contact with the tympanic membrane<sup>8</sup>.

The key originality of this case is that both clinical presentation and multimodal imaging independently and jointly oriented toward a Fisch type C tympanojugal paraganglioma. The severity of the conductive loss, the large pulsatile mass, and the three-year duration were clinically consistent with a jugular fossa-involving lesion. The angio-CT showed strong vascular enhancement and anterior jugular foramen erosion with direct tumor-jugular bulb contiguity. This convergence of clinical and imaging arguments made the case diagnostically challenging and the intraoperative reclassification to Fisch type B all the more significant.

A secondary pitfall was the discordance between MRI and CT enhancement. The MRI demonstrated weak gadolinium uptake, atypical for a paraganglioma, initially suggesting a schwannoma. Small or predominantly solid tympanic paragangliomas may lack the classic 'salt-and-pepper' pattern<sup>10</sup>. The subsequent angio-CT correctly identified the vascular blush but overestimated extent due to the jugular bulb dehiscence.

The central finding is the coexistence of a Fisch type B paraganglioma with complete jugular bulb dehiscence. This variant - absence of a bony plate covering the jugular bulb superior wall - has a prevalence of 6-19% in temporal bone dissection studies<sup>11,12</sup>. When adjacent to a middle ear mass, it generates a CT appearance indistinguishable from jugular invasion<sup>13</sup>. Shinohara, et al. found this variant accounts for radiological overclassification in ~15% of cases initially staged as tympanojugal paragangliomas<sup>14</sup>.

The intraoperative risk of an unrecognized dehiscence is significant: jugular bulb laceration can cause severe hemorrhage<sup>15</sup>. Semaan and Megerian identified this variant in 23% of operated tympanic paragangliomas, finding it an independent predictor of intraoperative blood loss<sup>16</sup>. Early recognition in our case was decisive for safe complete resection.

The distinction between Fisch type B and type C is of paramount surgical importance. Type C lesions require infratemporal fossa approaches with risks of up to 30% facial nerve palsy, permanent lower cranial nerve deficits, CSF leak, and major venous hemorrhage<sup>6,15</sup>. The intraoperative reclassification in our case directly prevented this unnecessary escalation.

## Conclusion

We report a Fisch type B tympanic paraganglioma in which a coexisting jugular bulb dehiscence led to convergent clinical and radiological overclassification as a Fisch type C lesion. Intraoperative exploration established the correct diagnosis and allowed complete transmeatal resection with excellent outcome. This case underscores that jugular bulb dehiscence must be systematically sought in temporal bone imaging, explicitly described in radiological reports, and anticipated intraoperatively.

A stepwise exploratory approach should always be considered before committing to an irreversible high-morbidity intervention when staging remains uncertain.

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