# Surgical Management of Tympanojugulare Paragangliomas: A Rare and Challenging Case of Vascular Neoplasma

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**ABSTRACT**
Tympanojugulare paragangliomas (TJPs) are benign tumors arising from neural crest cells located along the jugular bulbus and the tympanic plexus. TJPs have always presented a challenge to surgeons due to the fact that these tumors are vascularize and locally aggressive and involve important neurovascular structures. The treatment is guided by the classification of Fisch. We present a case of 54 years woman with a mass in the right ear canal associated with pulsatile tinnitus, hearing impairment and multiple lower cranial nerves palsy (CN VII, Xl & Xll). The clinical manifestations and paraclinical investigations established the diagnosis of tympanojugulare paraganglioma Fisch B2. According to the preoperative plan of the case, we decided to resected the tumour subtotally with canal wall down mastoidectomy procedure. Postoperative evaluation shows good healing and improvement of all the symptoms. The evidence-based literature search regarding the management of TJP using 'tympanojugulare paraganglioma as keywords was conducted on PubMed. Further selection is based on inclusion and exclusion criterion. four journals obtained are case reports and related to our case based on clinical manifestation and management of TJPs. The jugulotympanic paraganglioma have the tendency to involve multiple vital structures of the skull base. In most of these tumors, surgery remains the mainstay treatment. A detailed preoperative plan of treatment is required to maximize tumor resection, minimizing morbidity of the great vessels and lower cranial nerves and optimize the result.

***Keywords:*** *Paragangliomas, Tympanojugular*

## 1. INTRODUCTION

Tympanojugular Paraganglioma (TJP) is generally considered to be benign. TJP is a highly vascular tumor that is slow-growing, but often locally aggressive and infiltrates the skull base [3]. Temporal bone paraganglioma arises from various places in the temporal bone, and is classified into 2 types. The first is a tympanomastoid paraganglioma, commonly known as "glomus tympanicums," which is originates in the glomus body located along the Jacobson's nerve (tympanic branch IX), and Arnold's nerve (auricular branch X). The second group of tumors is the TJP or "glomus jugulare," which originates from the paraganglia located in the adventitia dome jugular or from the hypotympanum with the second invasion of the jugular bulb [4].

Fisch and Mattox and Moe et al classified temporal bone paragangliomas based on high-resolution CT examination, and modified by Shin et al (Table 1).

 

**Table 1.** Modified Fisch classification of temporal bone paraganglioma

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| Tympanomastoid paragangliomas | Tympanomastoid paragangliomas | Tumors confined to the middle ear<br>A1 Tumor margins clearly visible on otoscopic examination.<br>A2 Tumor margins not visible on otoscopy. Tumors may extend anteriorly to the eustachian tube and/or to the posterior mesotympanum.<br>Tumors confined to the tympanomastoid cavity without destruction of bone in the infralabyrinthine compartment of the temporal bone.<br>B1 Tumors involving the middle ear with extension to the hypotympanum.<br>B2 Tumors involving the middle ear with extension to the hypotympanum and the mastoid.<br>B3 Tumors confined to the tympanomastoid compartment with erosion of the carotid canal. |
| Tympanojugular paragangliomas | Tympanojugular paragangliomas | Tumors extending beyond the tympanomastoid cavity, destroying bone of the infralabyrinthine and apical compartment of the temporal bone and involving the carotid canal.<br>C1 Tumors with limited involvement of the vertical portion of the carotid canal.<br>C2 Tumors invading the vertical portion of the carotid canal.<br>C3 Tumors with invasion of the horizontal portion of the carotid canal.<br>C4 Tumors reaching the anterior foramen lacerum.<br>Tumors with intracranial extension<br>De1 Tumors up to 2 cm dural displacement.<br>De2 Tumors with more than 2 cm dural displacement.<br>Di1 Tumors up to 2 cm intradural extension.<br>Di2 Tumors with more than 2 cm intradural extension.<br>Di3 Tumors with inoperable intradural extension.<br>Tumors involving the vertebral artery.<br>Ve Tumors involving the extradural vertebral artery.<br>Vi Tumors involving the intradural vertebral artery. |

. The peak age for occurrence of TJP is between the 5th and 6th decades of life with a female-to-male ratio of 4-6:1 [1]. The most common symptoms of TJP are throbbing tinnitus and hearing loss. Jugulotympanic paraganglioma may present with symptoms secondary to mass effect on surrounding structures, including blood vessels or lower cranial nerves (VII, IX, X, IX) such as facial paralysis, dysphagia, dysphonia, or vertigo [5].

Otoscopic examination of a middle ear paraganglioma may reveal a reddish-blue, pulsating mass medial to the tympanic membrane. Catecholamine secretion occurs in only 2% of cases. Investigations of TJP should include a CT scan of the head to identify the extent of bone damage and an MRI to determine the extent of the tumor; T2-weighted images show a “salt and pepper” appearance of the paraganglioma. Diagnostic angiography can be confirmed and shows the specific vascular supply of the paraganglioma [5].

Three main recommended treatment options for TJP are: surgery, wait-and-scan, and radiotherapy (RT). Although surgical management has been the mainstay of TJP, the success in terms of complete disease eradication and minimal complications achieved by some centers of excellence has not been uniformly replicated elsewhere.

The main reason for this is that the TJP is trans-cranio-temporo-cervical, therefore, requiring the surgeon to have a thorough understanding of the skull base anatomy and surgical techniques to treat it effectively. RT as effective as surgical techniques are being developed to treat this subset of tumors. On the other hand, the realization that many of these tumors exhibit a sluggish growth pattern is, quite rightly, leading to a wait-and-scan policy [4].

TJP is often locally aggressive and infiltrates the skull base of the bone and regional cranial nerves (CNS). Surgical management of these lesions remains very challenging as a result of the complex anatomic location and the potential for intraoperative and postoperative complications. The main problems include: (1) inadequate exposure as this tumor can spread in 3 different compartments (intrapetrous, extracranial, and intradural); (2) the vertical portion of the facial nerve is centered and closely related to the jugular bulb; and (3) close connections with important neurovascular structures, such as the lower cranial nerves, inferior petrosal sinus, and internal carotid artery (ICA) [6]. The purpose of this case report is to share comprehensive information about the diagnosis and the management of TJPs.

## 2. Case Report

A 54-year-old woman was referred to the ENT clinic of Wahidin Sudirohusodo Hospital on February 5, 2021 with a diagnosis of right ear canal tumor. The patient's complaint of pulsatile tinnitus 3 years ago, headache, bloody otorrhoea in the last 1 year, otalgia, hearing loss, and dizziness. The patient also complained about how her face was asymmetrical and this was from 3 months ago, also weakness of the right shoulder. The patient had a history of previous right ear surgery 1 month before admission.

From inspection we found right facial nerve palsy House-Brackmann III, atrophy on right side of tongue, difficulty to up right shoulder, right retroauricular show surgery incision scar. Otoendoscopy on right ear show

bloody secretion surround a pulsatile mass on ear canal and blocking visualization to the tympanic membrane

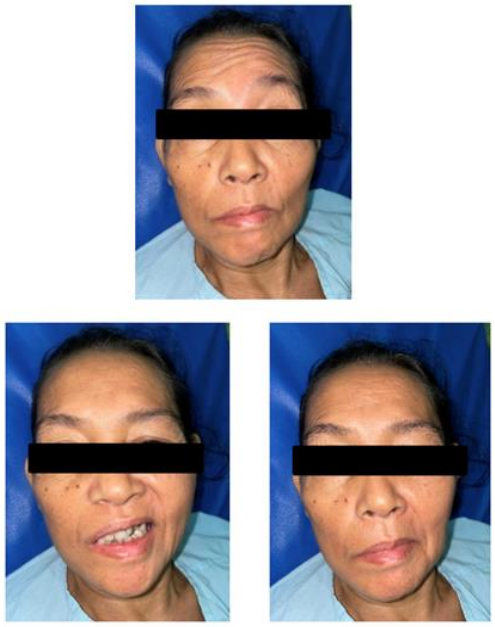

**Figure 1** Pre-Operative Image

Anterior rhinoscopy and pharyngoscopy did not reveal any abnormalities. Neck assessment: no enlargement of the lymph nodes. Pure tone audiometry test revealed right Profound Mixed Hearing Loss and normal hearing on left ear.

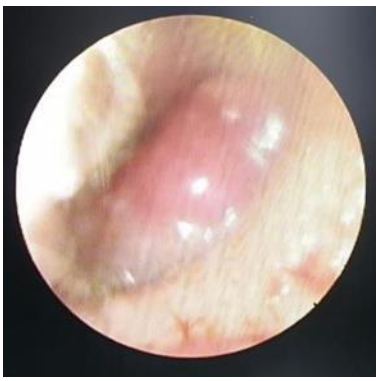

**Figure 2** Otoscopy A Red Mass the External Ear Canal

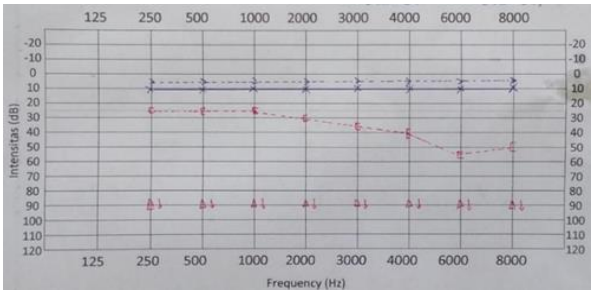


**Figure 3** Audiogram Laboratory Results Normal Performed

CT scan of the temporal bone without contrast found soft tissue mass filled in the right external acoustic canal that extends to the tympanic cavity and mastoid antrum. Enlargement of internal jugular vein and erosion of the jugular bulb plate.

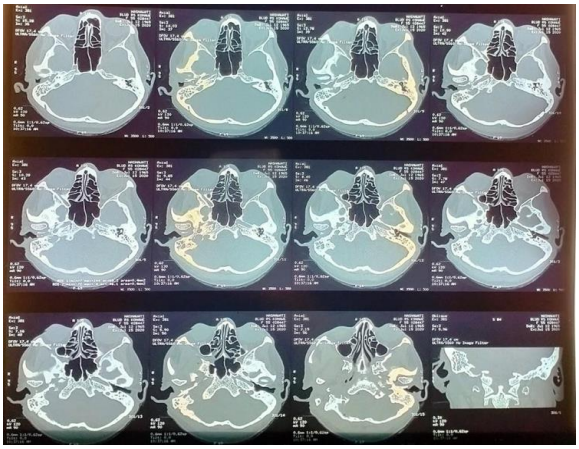

**Figure 4** CT Scan temporal bone

MRI of the brain impression of right glomus tympanojugulare and persistent cavum septum pellucidum et cavum vergae.

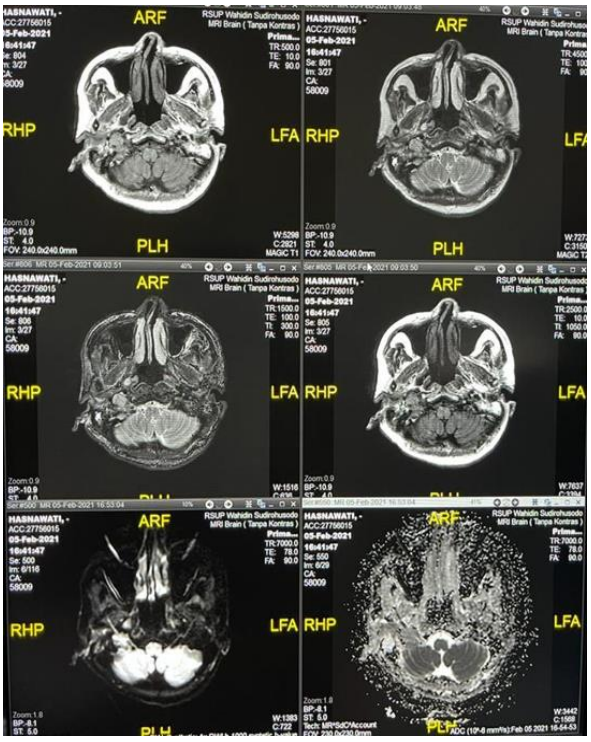


**Figure 5.** MRI

MSCT Scan of cerebral and carotid angiography obtained the impression of a suspected thrombus proximal to the right jugular vein. Four days before surgery embolization proceeded. Subtotal tumor resection with canal wall down mastoidectomy procedure performed on 22 march 2021.

Intraoperative finding: mass on the ear canal (polyp), tympanic membrane very thick, vascularized tumor fulfill the tympanic cavity extended to mastoid antrum and necrotic tissue surrounding, erosion on lateral ear canal wall, scutum, and ossicle (malleus, incus), facial nerve intact, lateral semicircular canal intact.

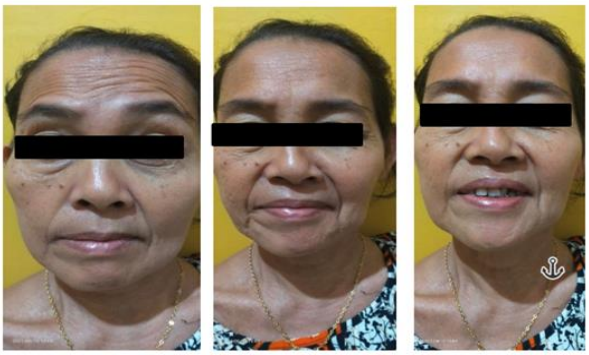

**Figure 6** Post operative image

## 3. METHOD

Structured literature search through PubMed using the keywords tympanojugulare paraganglioma and treatment. The search was conducted on June 15, 2021. Inclusion criteria were 1) literature on TJPs, 2) published in the last 10 years, 3) Free Full Text, 4) text in English. While the exclusion criteria are: article review, lack of relevance.

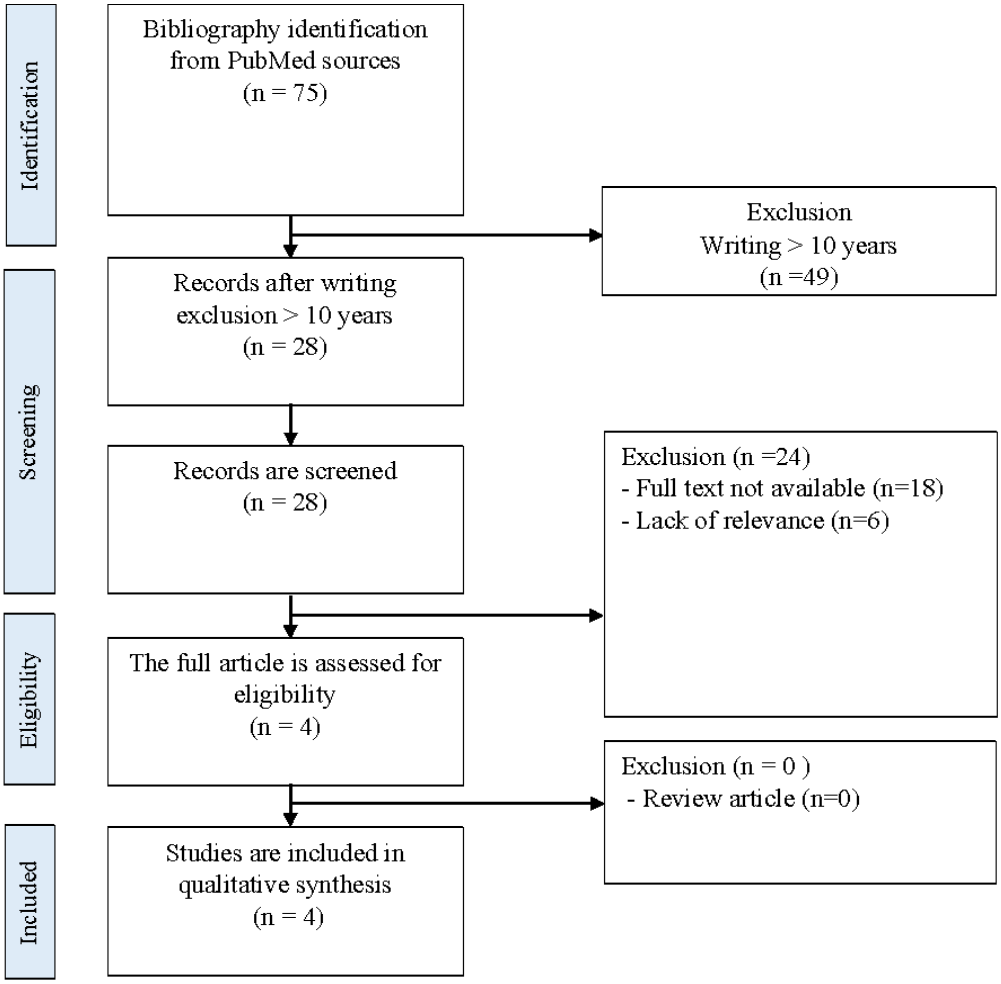


**Figure 7** Literature search strategy and results

## 4. RESULT

The search results in Pubmed journals obtained 75 journals that were adjusted to the keywords entered in each database. (Chart 1). The four articles obtained are of relevance for critical review related to clinical questions. The critical assessment is described in Table 2.

**Table 2.** The Critical Assessment

| No | Paper Identities | Result |
|---|---|---|
| 1 | **Author Journal:**<br>Harati, et al, 2014. (TJ 1) [7]<br>**Type Publication:** Research<br>**Population/Patient/ProblemIntervention/Index/Indicator:**<br>14 patients<br>with large tympanojugular glomus (Fisch grades C and D)<br>**Comparator:**<br>(Group 1)<br>Radical tumor removal (interdisciplinary microsurgical resection)<br>1 patient with tympanojugulare paraganglioma FISCH type B2<br>(Group 2) | **Outcome:**<br>(Group 1)<br>Pre-surgery:<br>- 11 patients with hearing loss<br>- 2 patients with progressive facial nerve palsy<br>- 2 patients had dysphagia and dysarthrophonia<br>- 2 patients with tumor regrowth (Both had facial nerve palsy [HB grade V] and lower cranial nerve palsy.<br>-All patients received cranial digital subtraction angiography (DSA)  Multistage preoperative tumor embolization was performed in all but one patient.<br><br>Pre-surgical case:<br>Complaints of tinnitus, otorrhoea, hearing loss, and facial nerve palsy HB III<br>The patient received DSA<br><br>Post-surgery:<br>- Radical tumor removal was performed in 10 patients, Near-total resection in 4 patients<br>- Hearing was maintained in 4 patients with normal preoperative audiograms.<br>- The facial nerve was preserved in all patients.<br>- Temporary facial nerve paralysis occurred in 2 patients and improvement in long-term follow-up<br>- 3 previous patients with facial nerve palsy, nothing changed.<br>-Persistent vocal fold paralysis occurred in 3 patients and laryngoplasty was performed.<br>-Overall recovery based on the Karnofsky performance scale was 100% in 10 patients and 90% in 4 patients.<br>-No major complications such as major vessel injury, intracranial hemorrhage, or ischemia. There were no cases of postoperative ataxia or transient limb paralysis.<br><br>(Group 2)<br>Pre-surgery:<br>-The main complaint of the patient is pulsatile tinnitus, hearing loss accompanied by bloody otorrhoea<br>House Brackman (HB) III facial nerve paresis<br>-Patients received tumor embolization before surgery<br><br>Post-surgery:<br>-carried out subtotal resection of the tumor<br>-HB III  HB II<br>- In general, recovery based on Karnofsky performance scale is 100%<br>- no postoperative complications |
| 2 | **Author Journal**:<br>Duzlu et al, 2018 (TJ 5) [8]<br>**Type Publication:**<br>Research<br>**Population/Patient/ProblemIntervention/Index/Indicator:**<br>18 TMP patients with simple or modified radical mastoidectomy (MRM) and tympanoplasty after retroauricular incision | **Outcome:**<br>Demographic data<br>mean age 50.3 ± 11.7 (SD 25-71 years).<br>-The overall female: male ratio of cases is 1.8:1 and specifically:<br>TMP = 2.6:1<br>TJP = 1.2:1<br>-The ratio of events on the right: left =16:18<br>-The most common symptoms in both: tinnitus and hearing loss |

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| | 16 TJP patients with type A infratemporal fossa procedure (ITFA-A)<br>**Comparator:**<br>- Demographic characteristics,<br>- Symptoms and signs that appear<br>- Radiological findings,<br>- Surgical technique,<br>- Complications. | -Symptoms only in TJP: Facial paralysis, dysphagia, and dysphony<br>-Increased catecholamine levels in plasma and urine in 3 TJP patients and 1 TMP patient.<br><br>Tumor characteristics:<br>-TMP : Fisch type A 9 cases, Fisch type B 9 cases<br>-TJP C1: 11 cases, C2: 4 cases, and Di1: 1 case<br>- Average maximum tumor size in the axial plane:<br>TMP 11.2 ± 5.2 mm (range 5---23) and TJP 30.2 ± 10.0 mm (range 15---70), respectively (p = 0.001)<br>-DSA:<br>TMP: posterior auricular artery<br>TJP: occipital artery<br><br>Surgery:<br>-TMP □<br>17 cases: total tumor excision<br>4 cases CWD<br>2 cases via transanal<br>6 cases of CWU<br>-TJP □<br>8 cases: total tumor excision<br>(5 with preoperative embolization, 3 without embolization)<br>5 cases with subtotal resection received stereotactic radiotherapy<br><br>Follow up :<br>Average 25.8 months (4-108 months)<br>TJP : 40.1 months (10-108)<br>TMP: 10.2 months (1-26)<br><br>Complications:<br>TJP:<br>- Facial nerve paralysis:<br>3 patients with HB grade IV<br>2 patients with non-maintained facial nerve<br>-External carotid artery not preserved (1 patient)<br>- Otorrhea and tympanic membrane perforation (3 patients)<br>- Iatrogenic cholesteatoma (1 patient)<br>- SNHL profound (3 cases) |
| 3 | **Author Journal:**<br>Merzouqi B, et al. 2021 (JT 6) [2]<br>**Type Publication:**<br>Research<br>**Population/Patient/ProblemIntervention/Index/Indicator:**<br>15 cervical paraganglioma patients (group 1)<br>11 patients with TJP<br>(Group 2)<br>**Comparator:**<br>- clinical features<br>- Support<br>- The clinical findings<br>- Treatment results | **Outcome:**<br>Grup 1:<br>-Epidemiology:<br>Mean age 56.67 years (33-77)<br>Female dominant (0.25 ratio)<br>-Clinical characteristics:<br>Clinical features:<br>assertive and pulsating mass (86.6%)<br>No pain (73.3%)<br>Movable (26%)<br>Fixed (40%)<br><br>Affected side:<br>7 right<br>6 left<br>2 Bilateral, firm, pulsatile and asymmetric<br><br>100% normal otoscopy<br>Vagus nerve lesion 4 patients (2 patients with unilateral vocal cord paralysis)<br>Hypoglossal nerve lesion in 1 patient<br><br>Radiological characteristics<br>Ultrasound of the neck: 46.7% with a hypervascular mass, 57.14% near the great vessels of the neck |

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| | | CT scan is performed in 40% of patients with findings of:<br>-vascular massMa<br>-Described the exact location and measurements<br>-Determines proximity to major blood vessels, permeability, state of the internal jugular vein<br>-determines the presence of extension, and erosion of the structure of the skull base<br><br>MRI is performed in 80% of cases, depicting:<br>- Vascular mass, with characteristic "salt and pepper"<br>-T1 punctate hyperintense picture<br>-With contrast indicates fast washing<br><br>“The Shambin Classification”<br>Type 1: 1 patient<br>Type II : 7 patients<br>Type III: 2 patients<br><br>Examination<br>Urinary catecholamines and serum catecholamines are normal,<br>except 1 patient: increased >1.5 times normal<br><br>Operation procedure:<br>93.33% total resection<br>The rest is due to tumor expansion, a difficult vascular axis.<br>Group 2 (TJP):<br>-epidemiology:<br>Mean age 54.72 years (34-80)<br>Female dominance (ratio 0.37)<br><br>-Clinical features:<br>90.9% associated unilateral pulsating tinnitus and progressive hearing loss,<br>45.45% otorrhoea<br>18.18% authority<br>18.18% headache<br>1 patient with cervical mass<br><br>Otoscopy:<br>63.63% of the mass pulsates behind the tympanic membrane<br>18.18% mass raised on the inferior wall of the CAE<br>18.18% reddish mass in CAE<br><br>Affected side<br>-right 72.72%, left 27.27%<br><br>- Facial nerve lesions in 4 patients (36.36%):<br>HB II in 1 patient,<br>HB III in 1 patient<br>HBV in 2 patients.<br>-2 patients with hypoglossal nerve lesions and facial nerve lesions<br>-1 patient presented with Glossopharyngeal nerve lesions (velar paralysis leading to impaired swallowing), vagal nerve palsy with dysphonia and dysphagia and spinal cord lesions complementing Collet-Sicard syndrome.<br><br>-Audiometry:<br>normal hearing:1<br>mild hearing loss: 1<br>moderate hearing loss: 2<br>severe hearing loss: 3<br>very severe hearing loss: 4<br><br>Radiological feature: |

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| | | -CT scan of the temporal bone was performed in 10 patients (90.90% of cases)<br>-MRI was performed in 8 patients (72.72% cases). 4 patients had no intracranial invasion, 1 patient had pontocerebellar angle invasion, temporal lobe invasion was seen in 2 patients, and infratemporal fossa in 1 patient<br>-Classification of this tumor according to FISCH is as follows: type A (glomus tympanicum) 1 case; type B (glomus hypotympanicum) 4 cases; type C1 2 cases; type C2 De1 2 cases; type C3 Di3 2 cases.<br><br>-Lab checks:<br>Urine catecholamine test (every 24 hours) and/or serum catecholamines<br>the screening was normal in all 7 patients tested.<br><br>- Embolization:<br>Performed in 3 patients out of 8 operated on and 27.27% of all cases, all paraganglioma type C, 2 of which were classified as C1 and one as De1C2.<br><br>Surgical procedure:<br>-retro-auricular approach: 4 patients,<br>-infratemporal approach type A p:3 patients<br>-transcanal approach: 1 patient with type A TP but mastoidectomy was also performed.<br>-3 patients are inoperable.<br>-3 patients ossicular chain preserved<br>ossicular chain reconstruction with anossiculoplasty in 1 patient,<br>-3 patients who cannot be operated on are considered for radiotherapy<br><br>Post-surgery:<br>- 2-7 days treatment on average<br>- vertigo(+), tinnitus(+)<br>- audiometry: preservation of hearing level in 4 patients whose middle ear can be preserved<br><br>Histopathology: The diagnosis of paraganglioma was confirmed in 100% of patients<br>after histopathological examination. |
| 4 | **Author Journal:**<br>Bacciu A et al. 2014 (TJ 4) [3]<br>**Type Publication:**<br>Research<br>**Population/Patient/ProblemIntervention/Index/Indicator:**<br>122 Fisch class C TJP patients who have undergone surgical procedures and facial nerve management (minimum 1 year postoperative)<br>**Comparator:**<br>Demographic data, surgical procedures, intraoperative FN management, preoperative and postoperative FN function.<br>Clinical improvement of facial nerve paresis (House-Brackmann) | **Outcome:**<br>Demographic Data:<br>- Male: 37 (30.3%), female 85 (69.7%).<br>-Mean age of the patient at the time<br>surgery 45.5 ± 12.3 years (16-67).<br>-67 tumors on the left side (54.9%), and 55 (45.1%) on the right<br>-Distribution of tumors (Fisch classification):<br>Type C1 (17 patients/13.9%), type C2 (73 patients/59.8%), type C3(29 patients/23.8, type C4 (3 patients/2.4%)<br><br>Surgical procedure:<br>- Single-stage tumor removal:111 patients<br>- IFTA-A :109 patients<br>- fallopian bridge<br>Technique 2 patients with Fisch class C1 TJP with posterior dominant disease<br>- The planned stepwise procedure was adopted using IFTA-A as the initial operation in 11 patients who had an intradural tumor extension of more than 2 cm; a petro-occipital-trans-sigmoid approach was used for planned second-stage intradural tumor removal in 5 cases, a transcochlear approach was used in 5 cases, and an extreme lateral approach in the remaining cases. |

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| | | Facial nerve management:<br>- The facial nerve was left in place in 2 patients (1.6%), diverted anteriorly in 97 patients (79.5%), diverted anteriorly by segmental resection of the epineurium in 7 patients (5.7%), and sacrificed and reconstructed in 15 patients (12.3%).<br>-1 patient underwent a cross facial nerve graft.<br>-At the last follow-up, House-Brackmann grades I to II were achieved in 51.5% of patients who underwent anterior rerouting and 28.5% of those who underwent anterior rerouting with epineurium resection. House-Brackmann grade III was achieved in 73.3% of patients who underwent cable nerve graft interposition.<br>-2 patients whose facial nerve was left behind had grade I and grade III, respectively. Patients who underwent facial nerve grafts had grade III. Total resection was achieved in 105 cases (86%)<br><br>Complications:<br>- Perioperative complications of cerebrospinal fluid leakage: two cases (1.6%).<br>-Total tumor removal was performed in 105 cases (86%). Of these, three cases (2.8%) had tumor recurrence. resection of a subtotal of 17 cases (14%). Fifteen of these residual tumors were intracranial involving the cavernous sinus, posterior dura fossa, and vertebral and basilar arteries.<br>-For extracranial residual tumors, partial resection was performed in two cases due to significant cardiac morbidity. |

## 5. DISCUSSION

Paraganglioma is a benign, but locally invasive hypervascular neoplasm. TJP accounts for only 0.012%. of all tumors. TJP are found in the carotid body, the vagus nerve, along the internal jugular vein, or the tympanic cavity. The TJP tumor are generally present in the 5th or 6th decade of life with main symptoms including pulsatile tinnitus and hearing loss.[5] This is consistent with this case report which describes the presentation of a jugulotympanic paraganglioma that occurred in a woman in the 5th decade of life with the chief complaint being pulsatile tinnitus as the chief complaint accompanied by accompanying complaints of hearing loss. And according to research conducted by Sivalingam et al found the incidence of TJP was more common in women where the ratio of female to male: male is 1.8:1, this is also following research conducted by Marzouki B et al where the average age was 54.72 years (range 34-80 years) and was dominated by women (ratio 0.37).

Jugulotympanic paraganglioma may present with symptoms secondary to mass effect on surrounding structures, including blood vessels or lower cranial nerves (VII, IX, X, IX) such as facial paralysis, hearing loss (conductive or sensory), tinnitus, dysphagia, dysphonia, or vertigo. This is following this case report wherein this patient facial nerve palsy was found but no lower cranial nerve abnormalities or colli region masses were found. This is also consistent with a study by Harati et al where from 14 TJP patients, 2 patients had progressive facial nerve palsy, 2 patients had dysphagia and dysarthrophonia, 2 patients had tumor regrowth where both had facial nerve palsy [HB grade V], and nerve palsy. lower cranial. Similarly, the study conducted by Merzouki et al studied 11 TJP patients where 1 patient had a cervical mass and 4 patients had facial nerve lesions, and 2 patients had hypoglossal nerve lesions and facial nerve lesions, and patients presented with Glossopharyngeal nerve lesions (velar paralysis leading to impaired swallowing), vagal nerve palsy with dysphonia and dysphagia and spinal cord lesions complementing Collet-Sicard syndrome [2].

Otoscopy examination of the middle ear paraganglioma revealed a reddish blue pulsating mass to the tympanic membrane [9]. In this case report patient, otoscopic examination found a reddish tumor mass filling the right external acoustic canal, the tympanic membrane was difficult to evaluate.

Catecholamine secretion occurs in only 2% of cases.[5] Preoperative assessment of catecholamine secretion by the tumor should be performed in all patients to avoid fluctuations in blood pressure and extensive perioperative tachycardia. In the case of catecholamine-secreting tumors, initial treatment is required and extensive investigations should be performed to exclude the possibility of synchronous pheochromocytoma or sympathetic paraganglioma.[7] This is different from the study conducted by Dzulu et al which examined 16 TJP patients, and only 3 patients underwent catecholamine examination and found no increase in catecholamine levels in plasma or urine.

Merzouq et al also did not perform examinations on all TJP patients, of 11 patients only 7 patients underwent laboratory examination of urinary catecholamines per 24 hours and/or serum catecholamines and obtained normal results in all patients tested. In our patient, we did not do catecholamine examination due to the unavailability of these examination facilities [2].

Given the anatomical predisposition of paranggliomas of the middle ear, patients most often present with audiological disorders. Hearing loss can be sensorineural (SNHL), mixed (MHL), or conductive (CHL). SNHL is rare, often incurable, and is associated with cochlear invasion and fistulization. CHL, which is significantly more common, is often caused by ossicular mass loading, ossicular discontinuity, and tympanic membrane or round window obstruction.10 Of the 11 TJP patients studied by Merzouqi B et al, 1 patient with normal hearing, 1 patient with mild hearing loss, 2 patients with moderate hearing loss, 3 patients with severe hearing loss, 4 patients with very severe hearing loss. In this case report, audiometric examination revealed very severe mixed hearing loss.

Investigations should include a CT scan of the head to identify the extent of bone damage and an MRI to determine the extent of the tumor; The T2-weighted image shows the appearance of a "salt and pepper" paraganglioma. If the diagnosis is not clear, diagnostic angiography can be confirmed, which shows the specific vascular supply of the Paraganglioma.5 This patient underwent a CT scan and revealed obliteration of the right external acoustic canal that extends to the tympanic cavity, protruded jugular bulb, and destruction of the pale jugular. bulb and perselubungan on the mastoid antrum. Followed by MRI examination and obtained the impression according to the picture of glomus jugulare dextra, persistent cavum septum pellucidum et cavum vergae, mastoiditis dextra from these investigations, it was concluded the patient was classified as type C1 TJP. Then CT angiography was performed and found blood supply from the right inferolateral trunk, right posterior auricular artery, right occipital artery, and right superficial temporal artery and it was decided to carry out the embolization procedure.

Treatment of TJP remains controversial. Fractionated radiotherapy, stereotactic radiosurgery, gamma knife radiosurgery, and cyberknife radiosurgery as primary treatments have revealed the control of high rates of tumor growth and relatively low morbidity in current treatment. 7 Surgery for large TJPs is challenging because of its high vascularity and close association with neural and spinal structures. fine blood vessels. However, favorable long-term outcomes can be achieved by incorporating the surgical expertise of specialized neurosurgeons, ENT surgeons, and interventional neuroradiologists in the planning and execution of surgery. In this patient, it was decided to perform a subtotal tumor resection with canal wall down mastoidectomy. Intraoperatively we found; anterior lateral ear canal mass, hypertrophied tympanic membrane, and skin of the ear canal, very thick tympanic membrane, then behind it there is a tumor and necrotic tissue, tumor of the mastoid cavity, mastoid antrum, attic and middle ear, mastoid air cell. diploic bristle tumor mass, lateral wall of the ear canal erosion, minimal erosion of the scutum, ossicle erosions (malleus, incus), facial nerve intact mastoid, pars tympanic unidentified, intact lateral semicircular canal, active bleeding from tumor mass, tumor mass reddish with tissue necrotic, and performed canal wall down mastoidectomy procedure, free the tumor from the mastoid cavity, antrum, attic, tympanic cavity to the hypotympanum and eustachian tube, clean other pathological tissue, removal of ossicular erosion. After subtotal removal of the tumor, meatoplasty was performed and the incision wound was sutured layer by layer. Postoperative evaluation found good healing after surgery, complaints of bloody otorrhoea no longer exist and facial nerve paralysis improved (after surgery to HB grade II). No complications were found in this patient after surgery.

## 6. CONCLUSION

TJP is a benign tumor but need appropriate assessment due to the invasive character to vital structures. Surgery remains the mainstay management of TJPs but long term follow up still needed to anticipate regrowth other complications of the tumor. A detailed preoperative plan of treatment is required to maximize tumor resection, minimizing morbidity of the great vessels and lower cranial nerves and optimize the result.

## ETHICAL DECLARATION

Authors declared that the patient was consented about publishing his case on the scientific journal.

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