Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pathology

Type 4 Tympanic Paraganglioma Mimicking a Cholesteatoma: A Case Report

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DOI: https://doi.org/10.36347/sjmcr.2026.v14i07.016 | **Received:** 06.06.2026 | **Accepted:** 17.07.2026 | **Published:** 18.07.2026

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Abstract | **Case Report**

Introduction: Tympanic paraganglioma is a rare tumor that predominantly affects women. Type 4 is uncommon. It can clinically and radiologically mimic a cholesteatoma. ***Clinical Case****:* We present a case of a type 4 tympanic paraganglioma in a 63-year-old male with no significant past medical history who presented with right-sided hearing loss and tinnitus that had been progressing for 15 months, complicated by ear pain. Otoscopic examination revealed a non-pulsatile mass in the external auditory canal. The Weber and Rinne tests, as well as the audiogram, demonstrated conductive hearing loss. Computed tomography (CT) scanning revealed a mass occupying the middle ear with mild bone erosion and extension into the external auditory canal, suggesting a cholesteatoma. Laboratory tests were normal. Surgical resection revealed a firm, brownish, polypoid lesion measuring 2 x 1.5 cm. Histologically, a tumor proliferation arranged in nests was observed within a hypervascularized stroma. The tumor cells had regular nuclei with "salt-and-pepper" chromatin. Immunohistochemical analysis revealed that the tumor cells expressed synaptophysin and chromogranin A. Pancytokeratin was negative. S100 highlighted the sustentacular cells, and Ki-67 was low. The postoperative course was uneventful. The prognosis was favorable after 10 months of follow-up. ***Conclusion****:* The symptoms and erosive appearance on imaging may lead to a misdiagnosis of cholesteatoma. Pathological examination then corrects the diagnosis. Testing for germline mutations is necessary to identify familial forms.

Keywords: Tympanic paraganglioma, cholesteatoma, histopathology, immunohistochemistry.

INTRODUCTION

Paragangliomas (PGLs) are non-epithelial neuroendocrine tumors that originate in the paraganglia or glomus cells of the autonomic nervous system, which are derived from the neural crest [1]. Paragangliomas of the head and neck account for less than 0.5% of head and neck tumors, with an annual incidence of 0.001%. The carotid location is the most common, whereas tympanic paragangliomas are rare. They frequently occur in women in their 50s, either sporadically or, in some cases, as a hereditary condition [2]. Type 4 is rare. Clinically and radiologically, it may resemble a cholesteatoma. The definitive diagnosis is made by histopathological examination [3].

We present a case of a type 4 right tympanic paraganglioma in a 63-year-old man.

CASE REPORT

This is a 63-year-old male patient with no personal history of cancer, ear surgery, ear trauma, or chronic otitis media. He was seen in the otolaryngology department at Bogodogo University Hospital for hearing loss in his right ear and tinnitus that had been present for 15 months and had been complicated by ear pain. He reported neither ear discharge nor vertigo. Otoscopic examination revealed a reddish, non-pulsatile mass occupying the external auditory canal and obscuring the tympanic membrane. The Weber test was lateralized to the affected ear, and the Rinne test was negative. The remainder of the physical examination revealed no

cranial nerve involvement and was otherwise unremarkable.

The audiogram revealed a conductive hearing loss of 30 dB. A CT scan of the temporal bones showed a heterogeneous mass occupying the middle ear with mild erosion of the ossicles and extension into the external auditory canal, without intracranial extension, suggesting a cholesteatoma. Laboratory tests, including a complete blood count and basic metabolic panel, were normal.

The lesion was excised via tympanotomy. Macroscopic examination revealed a firm, brownish, polypoid lesion measuring 2 x 1.5 cm (Figure 1).

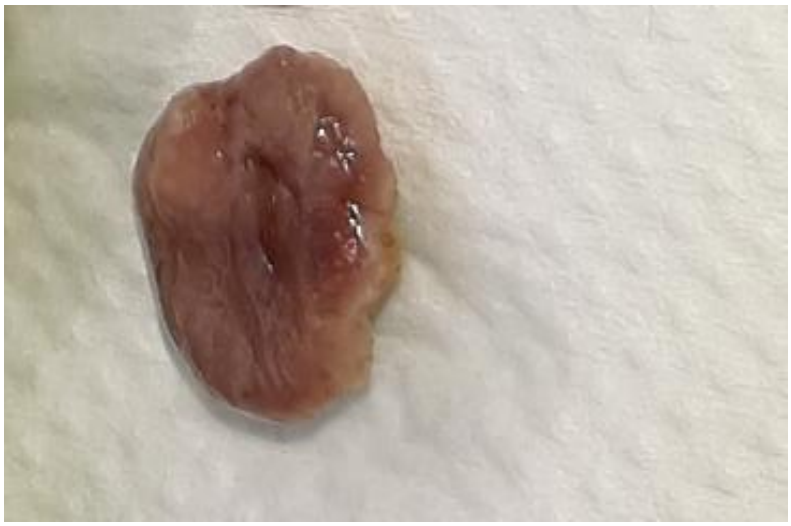

Figure 1: Macroscopic image of a polypoid paraganglioma measuring 2 x 1.5 cm, firm and brownish

Histologically, there was a fairly well-defined tumor proliferation, arranged in nests within a hypervascularized stroma, without necrosis. The tumor cells were eosinophilic, epithelioid, with regular nuclei, "salt-and-pepper" chromatin, and few mitoses (Figure 2). This histological appearance ruled out the possibility of a cholesteatoma. It was consistent with a paraganglioma, a hemangioma, or a neuroendocrine tumor.

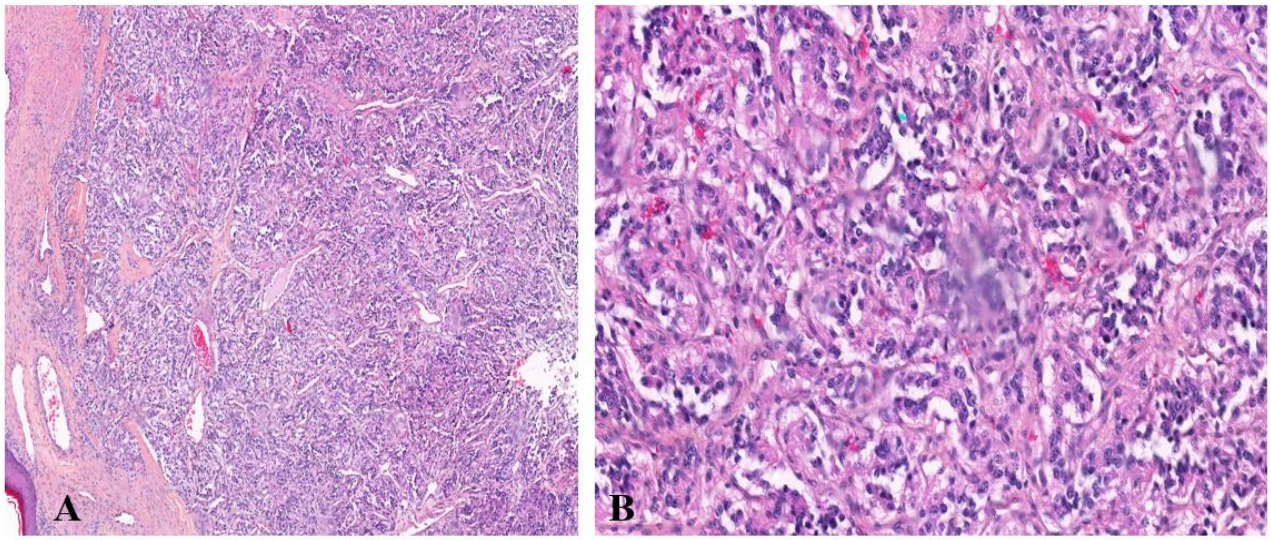

Figure 2 : Histological images of a paraganglioma. (A) – Hematoxylin-eosin (HE) stain, magnification (M) x 100: hypervascularized tumor proliferation. (B) – HE stain, M x 400: Clusters of eosinophilic, epithelioid tumor cells with regular nuclei and "salt-and-pepper" chromatin.

On immunohistochemical examination, the tumor cells expressed neuroendocrine markers (synaptophysin and chromogranin A). Pancytokeratin was negative. S100 highlighted the sustentacular cells, and CD34 revealed hypervascularization (Figure 3). The Ki-67 proliferation index was 1–2%. We therefore concluded that this was a type 4 tympanic paraganglioma, with no signs of malignancy.

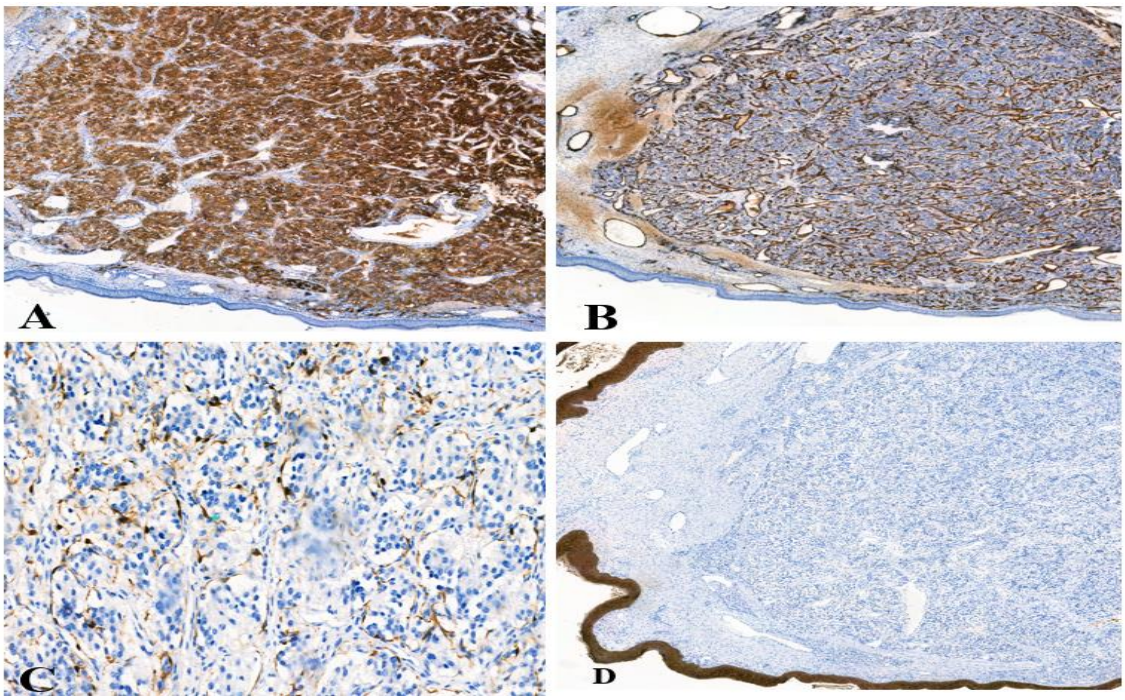

Figure 3 : Immunohistochemical images of a paraganglioma. (A) Chromogranin: cytoplasmic expression in tumor cells. (B) CD34: staining of blood vessels. (C) S100: staining of sustentacular cells. (D) Pancytokeratin: negative.

The postoperative course was uneventful. Symptoms returned to normal, with no evidence of recurrence after 10 months of follow-up.

DISCUSSION

A paraganglioma (PGL) is a tumor—most often benign—of the paraganglia, which originate from the neural crest. Tumors in the head and neck are fairly common, particularly in the carotid, jugular, tympanic, jugulo-tympanic, and vagal glomus. Paragangliomas of the tympanic glomus or middle ear are rare, accounting for approximately 30 to 40% of PGLs in the head and neck. There are four types, classified according to the tumor's location and extent. Type 4 is a tumor of the middle ear that extends into the external auditory canal [4,5]. Its incidence is estimated at about one case per 1.4 million people per year [6]. The majority are women (90.4%), with an average age of 55.2 years [6,7]. Hereditary forms often manifest in younger individuals and are sometimes bilateral or multifocal in about one-quarter of cases [8].

Paraganglioma is often sporadic. However, hereditary forms account for 40% of cases and are often associated with von Hippel-Lindau syndrome, neurofibromatosis type 1, multiple endocrine neoplasia (MEN) type 2, and paraganglioma syndrome [4,5]. In this case, it is likely a sporadic form, given our patient's age and the fact that the tumor is solitary.

Chronic hypoxia is a risk factor for PGL. The succinate dehydrogenase (SDHx) enzyme complex, composed of the proteins SDHA, SDHB, SDHC, SDHD, and SDHAF2, is often involved. It is an essential component of the link between the Krebs cycle and the mitochondrial electron transport chain. Thus, a deficiency or dysfunction leads to the production of adenosine triphosphate (ATP) via glycolysis and enables tumor cell proliferation even in a hypoxic environment [8].

The symptoms are dominated by pulsatile tinnitus (90%), hearing loss (80%), vertigo, and sometimes involvement of the lower cranial nerves; otorrhea or otalgia are rare. Functional forms are rare and may lead to Ménard's triad (palpitations, headaches, and sweating).[4,6,9]. On otoscopic examination, it appears as a painless, pulsating, bluish-red retro-tympanic mass, giving the appearance of a "rising sun" [2,10]. Audiometry reveals conductive hearing loss, as in our case. Sensorineural hearing loss may be associated with large tumors that have spread into the cranial cavity [2]. In our case, the presence of hearing loss, tinnitus, conductive hearing loss, and a painful, non-pulsating mass may indeed initially suggest a cholesteatoma, despite the absence of risk factors such as a personal history of ear surgery or trauma, or chronic otitis media.

High-resolution computed tomography with contrast or magnetic resonance imaging (MRI) of the cervicofacial or temporal regions is the imaging modality of choice for diagnosis, showing a well-defined intratympanic mass with "salt-and-pepper" enhancement (hypo- and hypersignal). The absence of bone destruction is the main distinguishing feature from cholesteatoma. Angiography shows a hypervascularized tumor [2,4,5,9]. Only a CT scan was performed on our patient. The erosive appearance of the ossicles was

consistent with a cholesteatoma, though it did not definitively rule out a paraganglioma.

Elevated levels of free plasma metanephrine and urinary metanephrine fractions above 1.5 times the reference value are found in secretory forms, which are rare (4.6%) [4,5]. This test was not performed in our case, as there was no suspicion of a paraganglioma following clinical and radiological examinations.

Macroscopic examination reveals an ovoid lesion with a firm to rubbery consistency and a heterogeneous appearance, particularly after embolization [8]. It often has an average size of 0.7 (±0.6) cm [5]. A biopsy is not recommended given the risk of bleeding [4,9]. Our lesion measured 2 cm along its longest axis; it was polypoid, ovoid, homogeneous, and was completely resected.

Histopathology typically reveals an alveolar or nest-like pattern known as "Zellballen," surrounded by sustentacular cells within a hypervascularized stroma. The tumor cells are amphophilic to eosinophilic, predominantly epithelioid and sometimes spindle-shaped. Their nuclei are round, with "salt-and-pepper" chromatin. Mitoses are rare. Sclerosing, pigmented, and clear areas may be observed. Cell pleomorphism, necrosis, and an infiltrating pattern involving soft tissues or bone may occasionally be noted (2%), but these findings are not predictive of aggressive behavior with the potential for metastasis. However, the distinction between multifocality and true metastasis has not been clearly established [1,4,8]. Our tumor had a classic histological appearance, with no signs of malignancy, but it was highly vascularized, which could also suggest a hemangioma.

On immunohistochemical analysis, the tumor cells express neuroendocrine markers such as synaptophysin, chromogranin A, and CD56. The GATA-3 transcription factor is positive (89%). They are negative for cytokeratin. S100 highlights the sustentacular cells. Ki-67 is often low [4,8]. This profile helps distinguish it from meningiomas, hemangiomas, adenomas, and neuroendocrine tumors of the middle ear [1,8]**.** The immunohistochemical findings were consistent with paraganglioma in our case.

Another important factor is the detection of germline mutations in the *SDHx* gene, characterized by a loss of expression in tumor cells as determined by immunohistochemistry. These mutations involve the SDHD (66%), SDHB (15%), and SDHC (6%) subunits. The SDHB mutation is of greatest concern due to the higher risk of metastasis. In rare cases of Carney's triad, this mutation may be sporadic and due to methylation of SDHC**.** Molecular biology testing also screens for other mutations, such as those in the *VHL*, *NF1*, and *RET* genes[1,4,5]. Our patient did not undergo this in-depth evaluation because these tests were not available.

Treatment involves surgery, fractionated radiation therapy, or active surveillance. Complete surgical resection is often the preferred option. However, subtotal resection or fractionated radiation therapy may be indicated for lesions that are infiltrative or adherent to vascular and nerve structures in order to avoid morbidity. [6,7,11]. Preoperative embolization may be performed to induce regression of the tumor [10]. Tumor résection was successfully performed.

The natural course of the condition is favorable, with slow growth (about 0.8 mm/year), no complications, and occasional local invasion [2,5,6]. Complications are often postoperative and include facial paralysis, perforation of the eardrum, surgical site infection, and cerebrospinal fluid leakage. The mortality rate is very low, as is the recurrence rate [2,10]. The outcome was favorable in this case, with no postoperative complications.

CONCLUSION

The symptoms of a middle ear lesion and its erosive radiographic appearance may lead to a misdiagnosis of cholesteatoma. Histopathological examination, combined with immunohistochemical analysis, is extremely helpful in correcting the diagnosis and identifying signs of malignancy. However, testing for germline mutations is necessary to identify familial forms of the disease.

Conflicts of Interest

The authors declare no conflicts of interest.

Author Contributions

All authors have read and approved the final version of the manuscript.

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