Cholesteatoma co-existing with schwannoma

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SUMMARY

Schwannoma is a benign, encapsulated tumor of Schwann cell origin. Approximately 25-45% of all Schwannomas occur in the head and neck. Still, schwannomas of the external auditory canal are a rare finding, and less than 10 cases are reported in literature worldwide. This report presents a rare case of a 55-year-old female with concurrent cholesteatoma and schwannoma in the external auditory canal, a combination seldom reported. Initially diagnosed as cholesteatoma based on clinical findings and imaging, the patient underwent modified radical mastoidectomy. Histopathology revealed the unexpected coexistence of schwannoma and cholesteatoma. Complete surgical excision ensured symptom resolution, with no recurrence over two years of follow-up. This case highlights the importance of histopathological evaluation in ear pathologies to uncover rare coexisting conditions, emphasizing accurate diagnosis and tailored management to optimize patient outcomes in clinical otology. This case report further emphasizes the distinction between cholesteatoma and schwannoma, as demonstrated through imaging characteristics and histopathological findings.

INTRODUCTION

A solitary schwannoma also referred to as a neurinoma or neurilemmoma, is characterized as a benign tumor encapsulated well, exhibiting slow growth, and often associated with pain.[1] These tumors originate from Schwann cells of nerves, explaining their diverse occurrence across the body, as Schwann cells are ubiquitous in peripheral nerves.[1,5] Notably, certain nerves, such as the optic and olfactory nerves, lack the Schwann cell coating, making them exempt from schwannoma development.[1,2] Head and neck regions harbor approximately 25-45% of all schwannomas,[1,4,7] with the cranial nerve VIII representing the most frequent intracranial site and the lateral aspects of the neck being the most common extracranial location.[1,5] According to Morais et al, the cranial nerve VIII in the head is the most afflicted, followed by the sensory nerves in order of frequency, as the motor nerves are only very seldom impacted.[1] This paper presents the incidental finding of schwannoma of the external auditory canal, evidenced by histopathological examination post-surgery for a patient who was clinically being treated as cholesteatoma and outcome based on our therapeutic management. It also emphasizes the distinction between cholesteatoma and schwannoma, as demonstrated through imaging characteristics and histopathological findings.

CASE PRESENTATION

A 55-year-old female, with 3 weeks history of chronic right otorrhea, reduced hearing, and otalgia, been treated by a General Practitioner multiple times with antibiotic ear drops however the pain persists and hence was referred to our centre for further management. Clinically patient appears well, not septic looking, and hemodynamically stable with intact facial nerve function. Further, otoendoscopy revealed a friable polyp totally occluding the right external auditory canal (EAC) (Figure 1). On palpation, the base of the polyp seems to arise from the floor of the EAC. A punch biopsy was done which revealed inflamed polypoid granulation tissue with acute on chronic inflammation. Computed tomography (CT) mastoid performed, showed large soft tissue density filling the right EAC and middle ear with erosion of scutum (Figure 2) erosion of the posterior EAC wall with extension into the mastoid air cells (Figure 3). The absence of abnormalities in the facial nerve bony canal reduces the likelihood of a facial nerve schwannoma. Pure tone audiometry done revealed moderate to severe hearing loss over right ear whereas left ear mild to moderate hearing loss.

Pre-operative diagnosis of cholesteatoma made based on clinical findings and Imaging done. The patient was counselled for surgery and underwent a right-modified radical mastoidectomy. Intra-operatively, the polyp was pedunculated at the anterior EAC wall and was excised fully with its base and sent for Histopathological examination. Further exploration revealed a large cholesteatoma sac medial to the excised polyp. The sac extended through an erosion of the posterior EAC wall extending into the mastoid and causing bony dehiscence over the sigmoid sinus. The cholesteatoma also extended medially onto the middle ear with erosion of the malleus. The stapes supra structure was preserved and the temporalis fascia was laid over this. Post-operative recovery was uneventful, with no cranial nerve palsy and hearing been improved.

The patient is currently about 2 years post-surgery, and there has been no recurrence of the schwannoma nor cholesteatoma based on clinical and otoendoscopy findings. From a histological perspective, the tumor is comprised of long, spindle-shaped cells, commonly arranged in a palisade pattern around their elongated nuclei. Antoni type A (Verocay Body) refers to areas with dense cell concentrations, while Antoni type B refers to areas with loose, asymmetrically placed cells. A positive S-100 protein indicates the origin of Schwann cells.[6,7]

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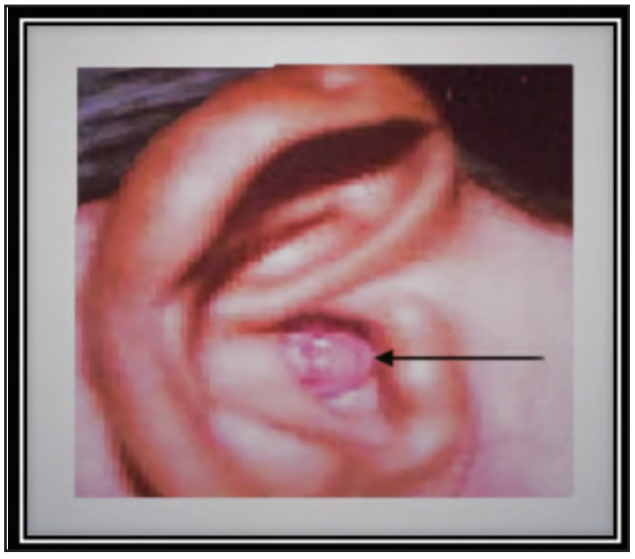

Fig. 1: Direct visualization of aural polyp over the EAC

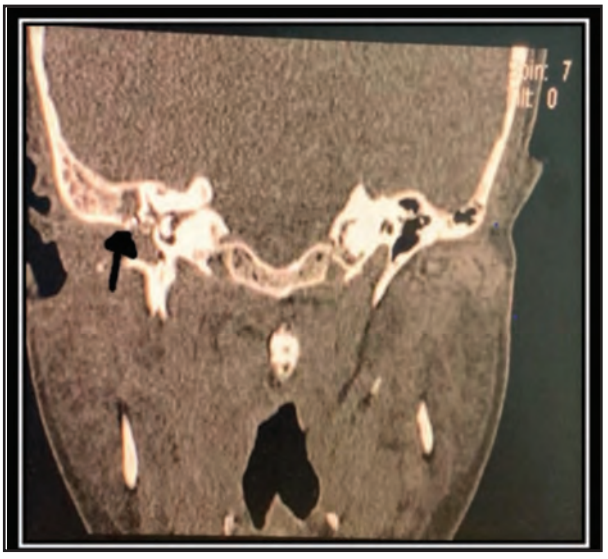

Fig. 2: CT scan without contrast Mastoid Coronal view showing scutum erosion (black arrow)

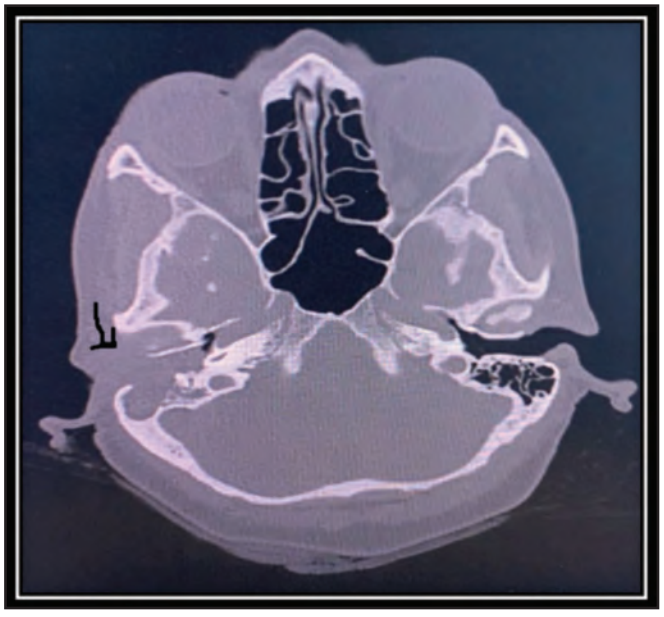

Fig. 3: CT scan without contrast Mastoid Axial view showing middle ear opacity over the right side with erosion of posterior EAC wall with extension into mastoid air cells (black arrow)

The recommended treatment for schwannoma is complete surgical excision. Although the diagnosis for the patient was confirmed post-operatively, she did not require further surgery because the lesion was prudently completely excised.

DISCUSSION

Solitary schwannomas are benign tumors that were first described in 1908 by Verocay, who named them neurinomas. In 1974, Batsakis introduced the name schwannoma. These tumors have been referred to by various terms, including neurinoma, neurilemmoma, mioschwannoma, and schwannoglioma.[1] The occurrence of schwannoma and cholesteatoma presenting concurrently is exceedingly rare. Schwannoma may arise alone or in conjunction with type 2 neurofibromatosis (NF2). It is also possible to see many schwannomas, particularly when NF2 or schwannomatosis is present.[3] Most extracranial schwannomas in the head and neck originate from branches that innervate the muscles or skin of the brachial or cervical plexus.[3,4,6]

The auriculotemporal nerve (V), the nervus intermedius of Wrisberg (VII b), and the auricular branch of the vagus nerve (X) make up the complex sensory innervation of the external auditory canal.[3,5,6,8,9] As a result of this, it can be tricky to determine the nerve that gives rise to an external auditory canal schwannoma, as in this case.[3,8]

The primary symptom of schwannoma is typically a slow-growing, painless mass. It is uncommon for schwannomas to cause neurogenic symptoms like pain or paresthesia, or motor issues, particularly when the tumor affects a motor nerve.[3] Managing this combination of pathologies necessitates careful consideration of the risks and benefits associated with various treatment options. It is uncertain whether the reduced hearing and otalgia experienced by our patient were linked to the schwannoma or chronic inflammation in the middle ear, as both conditions can manifest with these same symptoms. Presently, the preferred treatment for chronic middle ear inflammation with cholesteatoma involves a canal wall-down procedure with the complete removal of middle ear pathologies. It is crucial to ensure thorough removal of the cholesteatoma matrix.[1]

The concurrent presentation of cholesteatoma and schwannoma posed a diagnostic and therapeutic challenge. Our patient was initially diagnosed with cholesteatoma along with hearing impairment. The symptoms of the schwannoma were obscured by the diffuse symptoms of the cholesteatoma. The hearing impairment was initially attributed to the prolonged symptoms of the cholesteatoma. A routine preoperative CT scan confirmed a large soft tissue

Table I: Difference of external ear schwannoma and cholesteatoma

Differentiation Between Schwannoma of the External Ear and Cholesteatoma

Characteristic	Schwannoma (External Ear)	Cholesteatoma
Definition	A benign tumor arising from Schwann cells of the nerve sheath.	A collection of keratinizing squamous epithelium in the middle ear or external ear.
Etiology	Arises from nerves (e.g., auriculotemporal or great auricular nerve).	Chronic ear infections or eustachian tube dysfunction leading to retraction pockets.
Location	Typically occurs in the external auditory canal or adjacent areas.	Commonly found in the middle ear or mastoid; rarely extends to the external auditory canal.
Growth Pattern	Slow-growing, well-encapsulated mass.	Expansive and erosive growth that destroys adjacent bone.
Symptoms	- Painless mass in the external auditory canal.	- Otorrhea (foul-smelling discharge), hearing loss, ear pain.
	- Hearing loss or tinnitus (rare).	- Recurrent infections or fullness in the ear.
Appearance on Examination	Smooth, firm, and well-circumscribed lesion in the canal.	Whitish, irregular debris with granulation or bony erosion.
Imaging Findings	- Well-defined mass; no bony erosion unless large.	- Bony destruction on CT; hyperintense lesion on T2-weighted MRI with peripheral enhancement.
	- No keratin or soft tissue opacity.	- Soft tissue opacity and signs of infection or granulation tissue.
Histopathology	- Antoni A (cellular) and Antoni B (myxoid) regions.	- Keratinizing squamous epithelium with cholesterol crystals and inflammation.
	- Positive for S-100 protein.	- No S-100 positivity.
Management	Surgical excision with preservation of nerve function.	Surgical removal (e.g., tympanomastoidectomy) to prevent recurrence and complications.
Prognosis	Excellent; recurrence is rare.	Variable; recurrence or complications (e.g., intracranial spread) possible if not treated.

Table II: MRI findings to differentiate external ear schwannoma and cholesteatoma

Characteristic	Cholesteatoma	Schwannoma (External Ear)
Signal Intensity on T1	- Typically hypointense (low signal) due to keratin and debris.	- Isointense or slightly hypointense relative to muscle.
Signal Intensity on T2	- Hyperintense (bright) due to fluid content in keratin debris.	- Hyperintense (bright), especially in Antoni B regions, due to the myxoid stroma.
Contrast Enhancement	- Minimal to no enhancement after gadolinium contrast, as cholesteatoma lacks vascularity.	- Strong, uniform, or heterogeneous enhancement post-gadolinium due to vascularized Schwann cells.
Diffusion-Weighted Imaging (DWI)	- Hyperintense on non-echo-planar DWI due to restricted diffusion of keratin debris.	- Typically no restricted diffusion or only mild hyperintensity due to cellularity in Antoni A regions.
Location and Pattern	- Irregular, non-encapsulated lesion, often associated with bone erosion in middle ear or canal.	- Well-circumscribed, encapsulated lesion without bone destruction unless very large.
Bone Involvement	- Frequently involves bone erosion or destruction visible on adjacent CT imaging.	- Rarely causes bone erosion unless it reaches a significant size.

density lesion in the right external and middle ear, along with suspicious erosion of the scutum due to extensive cholesteatoma. MRI examination is not typically part of the routine evaluation for cholesteatoma cases. The definitive diagnosis was established postoperatively based on histopathological findings revealing a benign spindle cell neoplasm suggestive of schwannoma.[1]

Schwannomas are firm, smooth-surfaced tumors covered by normal skin. Originating from nerve sheaths, they are encased in a perineurium capsule and grow by expanding, pushing nerve fibers toward the periphery. Due to their similarities with other soft tissue tumors, the differential diagnosis includes conditions such as sebaceous adenoma, eosinophilic granuloma, fibroma, chondroma, and leiomyoma.[3] From a histological perspective, the tumor is comprised of long, spindle-shaped cells, commonly arranged in a palisade pattern around their elongated nuclei. Antoni type A (Verocay Body) refers to areas with dense cell concentrations, while Antoni type B refers to areas with loose, asymmetrically placed cells. A positive S-100 protein indicates the origin of Schwann cells.[6,7] The summarized tables detailing the presentations and MRI findings of external ear schwannomas and cholesteatomas as below (Table I and Table II).[10]

After thoroughly analyzing the treatment options in the case presented, we discussed the available choices with the patient and their family members upon receiving the histopathological specimen suggestive of schwannoma post-surgery. The choices of treatment that were explained to the patient and family members after receiving the histopathology were conservative treatment (serial monitoring by otoscopic and symptoms as intraoperatively the lesion has been completely excised), or performing MRI to assess further and keep in view for surgical excision if indicated, however patient and family members keen for conservative management by serial monitoring. From a socioeconomic perspective, since the family provided financial support for the patient's treatment, and clinical outcomes showed improved hearing, the family preferred the conservative treatment approach.

CONCLUSION

This case underscores the rare coexistence of schwannoma and cholesteatoma in the external auditory canal, initially misdiagnosed as isolated cholesteatoma. Histopathological examination was crucial for accurate diagnosis, preventing misidentification as granulation tissue. Given its rarity, our findings provide valuable insight, emphasizing the importance of thorough surgical assessment and routine histopathological evaluation and distinguish characteristics of cholesteatoma and schwannoma accordingly.

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