CASE REPORT

External Auditory Canal Schwannoma in Type II Neurofibromatosis: A Case Report

Giulia Elvira Malzanni[1], Michele Tulli[2], Omar Gatti[3], Mario Bussi[4], Lucia Oriella Piccioni[5]

Received on: 09 June 2020; *Accepted on:* 07 June 2021; *Published on:* 31 March 2025

Abstract

Introduction: In the English literature, very few cases of schwannomas arising from the external auditory canal (EAC) were reported.

Case description: We describe a 37-year-old patient affected by type II neurofibromatosis who presented with a soft mass occluding the external auditory meatus. He was treated with an en bloc excision with an endaural approach under general anesthesia. The histopathologic examination of the specimen revealed a schwannoma.

Discussion: We indicate the diagnostic and therapeutic workup for ear canal schwannomas reviewing the cases reported in the literature.

Keywords: Case report, External ear canal, Extracranial schwannoma, Nerve sheath tumor.

Otorhinolaryngology Clinics: An International Journal (2024): 10.5005/jp-journals-10003-1536

Introduction

Schwannomas (neurinoma) are slow-growing benign neural crest-derived tumors. The tumor arises from Schwann cells that surround the peripheral, cranial or autonomic nerves in different anatomic locations. Although they can develop in almost all regions of the body, 5–45% of schwannomas are found in the head and neck region. Schwannoma of the vestibulocochlear nerve is the most common and represents 8% of all intracranial tumors.[1] Their localization in the external auditory canal (EAC) is rare. We analyze a clinical case of schwannoma arising from the EAC and review the literature to define a diagnostic and therapeutic workup.

Case Description

A 37-year-old man affected with type II neurofibromatosis (with ring chromosome 22) presented with an EAC slowly growing mass. He was known for previous surgeries. The excision of a left vestibulocochlear schwannoma with consequent deafness, a cervical schwannoma and a thoracic schwannoma. He didn't refer to hearing loss, because of his cognitively poor condition, but his parents reported a suspicion of right hypoacusis. The objective examination showed a soft mass with a smooth surface, covered with normal skin that was neither bleeding nor ulcerated, which occluded the right external acoustic meatus and hindered the visualization of the tympanic membrane (Fig. 1). A computed tomography (CT) scan showed a well-circumscribed round mass obliterating the external auditory meatus arising from the posterosuperior wall of the ear canal (Fig. 2). The middle ear, mastoid and internal auditory canal were normal; no bone erosion was observed. A surgical excision with an endaural approach under general anesthesia was performed; a helicotragal incision was executed, and the mass was totally removed, preserving the integrity of the overlying skin and the surrounding osteocartilaginous structure (Fig. 3). At the end of the procedure, the tympanic membrane appeared intact and disease-free, and the canal was packed with a non-absorbable swab.

On postoperative day one, the patient was discharged in good conditions. Histopathologic examination of the specimen revealed a schwannoma. The mass was strongly positive for S-100 protein on an immunohistochemical staining. The patient is currently under

[1–5]Department of Otorhinolaryngology, San Raffaele Scientific Institute, Milano, Italy

Corresponding Author: Giulia Elvira Malzanni, Department of Otorhinolaryngology, San Raffaele Scientific Institute, Milano, Italy, Phone: +39 3345048330, e-mail: giuliaemalzanni@gmail.com

How to cite this article: Malzanni GE, Tulli M, Gatti O, *et al.* External Auditory Canal Schwannoma in Type II Neurofibromatosis: A Case Report. Int J Otorhinolaryngol Clin 2024;16(2):105–107.

Source of support: Nil

Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

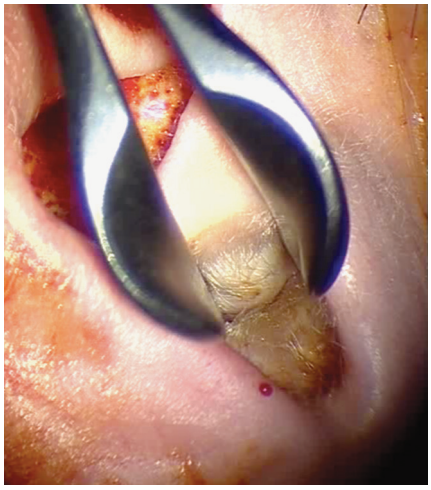

Fig. 1: A mass occluding external ear canal was observed on otoscopic examination

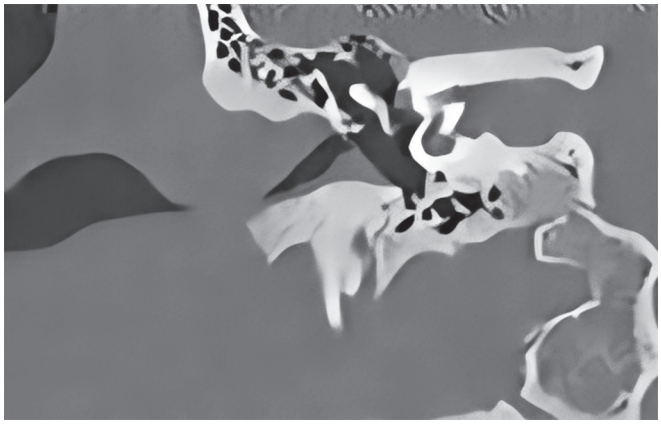

Fig. 2: In the coronal view of temporal bone CT scan, soft tissue shadows were observed in the posterosuperior wall of the ear canal

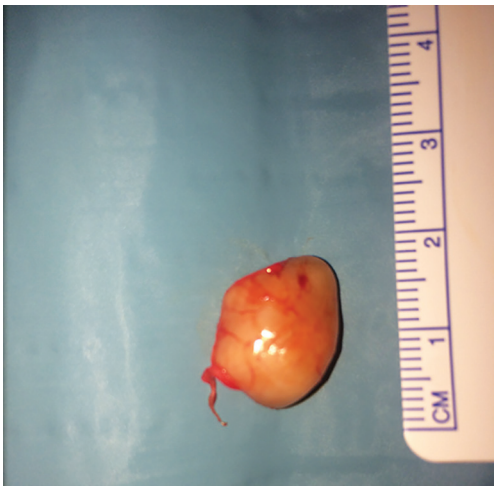

Fig. 3: Surgical specimen

follow-up (16 months from surgery), with no evidence of recurrence or narrowing of EAC.

Discussion

Schwannomas in the head and neck are most commonly associated with large nerve trunks. Schwannomas arising from the EAC are very rare. Usually it is impossible to define the origin of the neurinoma because innervation of the EAC involves complex overlapping of cranial Nerves (CNs) V, VII, IX, X and nerves from cervical plexus.

Only few cases of EAC schwannomas are reported in literature and their characteristics are summarized in Table 1.[2–12]

The clinical presentation of schwannoma is usually a slowly growing and asymptomatic mass that closes the EAC. The differential diagnosis should be made with other soft tissue neoplasms, such as fibroma, chondroma, and leiomyoma.

Ten of the eleven cases reported presented hearing loss, but only three of them had recurrent external otitis.[4,6,7]

In most of the cases (10/11), a CT scan was performed before surgery, and this was useful to define the extension of the mass and the possible bone or cartilage erosion.

Treatment is en bloc excision of the tumor via either a transmeatal or retroauricular approach. Some authors have chosen a transmeatal approach while others prefer a retroauricular approach.[2,3,5–10] A transmastoid approach was performed in one case, since the mass eroded the mastoid bone. We believe that a preoperative CT is mandatory to define the extension of the mass and the condition of the temporal bone and middle ear cavity. The choice of approach will depend on tumor size, location, and relation to surrounding structures.

The definitive diagnosis of schwannoma is made by histopathologic and immunochemical examination (strongly positive for S-100). In only one case, a chondroid metaplasia was seen.[10] The mean follow-up is 16 months (range 3–36) and none of the cases developed recurrence.

About postoperative imaging, Kim et al. performed a CT scan 8 months after surgery to confirm the success of EAC reconstruction,

Table 1: Characteristics of EAC schwannoma reported in literature

Authors	*Symptoms of presentation*	*Preoperative imaging*	*Bone/middle ear/ cartilage involvement*	*Surgical approach*	*Histology*	*Postoperative imaging*	*Postoperative complications*	*Follow-up*
Wu C et al.	Otalgia, bloody discharge	CT, MRI	No	Retroauricular	Typical schwannoma	–	None	12 months (no recurrence)
Harcourt JP et al.	Hearing loss, bloody discharge	–	No	Retroauricular	Typical schwannoma	Temporal bone CT	Not reported	18 months (no recurrence)
Lewis JB et al.	Hearing loss, recurrent external otitis	CT, MRI	No	Not specified	Typical schwannoma	–	Not reported	12 months (no recurrence)
Galli J et al.	Hearing loss	CT	No	Transmeatal	Typical schwannoma	Total body MRI	None	3 months (no recurrence)
Gross M et al.	Hearing loss and recurrent otitis	CT	No	Transmeatal	Typical schwannoma	–	Not reported	24 months (no recurrence)
Topal O et al.	Hearing loss, recurrent external otitis	CT	No	Transmeatal	Typical schwannoma	–	Not reported	6 months (no recurrence)

(Contd...)

Table 1: *(Contd...)*

Authors	*Symptoms of presentation*	*Preoperative imaging*	*Bone/middle ear/ cartilage involvement*	*Surgical approach*	*Histology*	*Postoperative imaging*	*Postoperative complications*	*Follow-up*
Magliulo G et al.	Hearing loss	CT, MRI	Cartilage erosion	Retroauricular + skin graft from postauricular region	Typical schwannoma	–	Not reported	36 months (no recurrence)
Bakshi SS et al.	Hearing loss	CT	Temporoman-dibular joint erosion	Retroauricular	Typical schwannoma	–	None	8 months (no recurrence)
Bennani A et al.	Hearing loss	CT	No	Transmeatal	Schwannoma with chondroid metaplasia	–	None	8 months (no recurrence)
Lee DH et al.	Hearing loss	CT	No	Retroauricular	Typical schwannoma	–	None	30 months (no recurrence)
Kim YH et al.	Hearing loss	CT	Mastoid erosion	Transmastoid + reconstruction through tragal cartilage and skin graft from postauricular region	Typical schwannoma	Temporal bone CT	Not reported	24 months (no recurrence)

as they had to reconstruct the lost lateral bony region of the EAC and the entire posterior surface of the cartilaginous portion with tragal cartilage. We agree about the necessity of a temporal bone CT scan in this case since the reconstruction has been wide, but normally a clinical examination is enough to rule out a narrowing of the EAC or a bad surgical outcome. Otherwise, we agree with Galli et al.[5] suggesting a total body MRI during the postoperative follow-up since schwannoma is associated with type II neurofibromatosis. The case we presented is the first report of an EAC schwannoma in which an association with NF2 is present, but due to the usualness of these schwannomas we believe that a total body MRI is mandatory after the histopathologic diagnosis of an EAC schwannoma.

Conclusion

Schwannoma should be considered in the differential diagnosis of benign or malignant tissue masses involving the external ear canal. The treatment is surgical (with a retroauricular or transmeatal approach). When the histopathological diagnosis gives evidence of an EAC schwannoma, type II neurofibromatosis should be excluded with MRI or genetic investigations.

Orcid

Giulia Elvira Malzanni https://orcid.org/0000-0003-4016-0642

References

1. Nager GT. Acoustic neuromas. Acta Otorhinolaryngol 1985;99(3–4): 245–261. DOI: 10.3109/00016488509108905.
2. Wu CM, Hwang CF, Lin CH, et al. External ear canal schwannoma: An unusual case report. J Laryngol Otolaryngol 1993;107(9):829–830. DOI: 10.1017/s0022215100124545.
3. Harcourt JP, Tungekar MF. Schwannoma of the external auditory canal. J Laryngol Otolaryngol 1995;109(10):1016–1018. DOI: 10.1017/s0022215100131913.
4. Lewis WB, Mattucci KF, Smilari T. Schwannoma of external auditory canal: An unusual finding. Int Surg 1995;80(3):287–282. PMID: 8775624.
5. Galli J, d'Ecclesia A, La-Rocca LM, et al. Giant schwannoma of external auditory canal: A case report. Otolaryngol Head Neck Surg 2001;124(4):473–474. DOI: 10.1067/mhn.2001.114797.
6. Gross M, Maly A, Eliashar R, et al. Schwannoma of the external auditory canal. Auris Nasus Larynx 2005;32(1):77–79. DOI: 10.1016/j.anl.2004.09.001.
7. Topal O, Erbek SS, Erbek S. Schwannoma of the external auditory canal: A case report. Head Face Med 2007;3:6. DOI: 10.1186/1746-160X-3-6.
8. Magliulo G, Parrotto D, Sardella B, et al. Cavernous hemangioma of the tympanic membrane and external ear canal. Am J Otolaryngol 2007;28(3):180–183. DOI: 10.1016/j.amjoto.2006.03.012.
9. Bakshi SS, Shankar K, Parida PK. A large schwannoma of external auditory canal: An unusual case. Kulak Burun Boğaz Ihtis Derg 2015;25(4):229–231. DOI: 10.5606/kbbihtisas.2015.43402.
10. Bennani A, Karich N, Kamaoui I, et al. Schwannoma with chondroid metaplasia of the external auditory canal–A rare finding in a rare location: A case report J Med Case Rep 2018;12(1):66. DOI: 10.1186/s13256-018-1584-4.
11. Lee DH. Schwannoma of external auditory canal. Auris Nasus Larynx 2018;45(3):630–632. DOI: 10.1016/j.anl.2017.07.022.
12. Kim YH. Massive schwannoma originating from the external auditory canal invading the mastoid. Ear Nose Throat J 2019;98(1):25–27. DOI: 10.1177/0145561318824808.