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REVIEW ARTICLE

Extension patterns of vestibular schwannomas towards the middle ear: three new cases and review of the literature

Aina Venkatasamy[1,3] · Caroline Nicolas-Ong[1] · Hella Vuong[2] · Anne Charpiot[2] · Francis Veillon[1]

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Abstract

Objective Middle ear extension of vestibular schwannomas is not a common occurrence, and only a few cases have been described so far in past publications. We report three new cases of vestibular schwannomas extending to the middle ear and reviewed the literature to specify the patterns of such an extension.

Materials and methods We analysed databases of previously published articles to search for additional cases of middle ear extension of vestibular schwannomas and compared them to the cases we have documented. Extension patterns of the tumours were analysed, especially focusing on the extension through the round and oval windows.

Results and conclusion Middle ear vestibular schwannomas are uncommon tumours and only 13 cases have been published so far. The vestibular schwannoma (internal auditory canal or intralabyrinthine) has to invade the labyrinth first (complete invasion in 88% of the cases, $n = 14$), before reaching the middle ear. In the majority of cases (69%, $n = 11/16$), internal auditory canal vestibular schwannomas or intralabyrinthine schwannomas extended in the middle ear though the round window.

Keywords Vestibular schwannomas · Middle ear · Round window · CT · MRI

Introduction

Vestibular schwannomas are benign tumours, representing around 80% of cerebellopontine angle tumours. Their most common symptoms are asymmetrical progressive sensorineural hearing loss, vertigo and tinnitus [1, 2]. Vestibular schwannomas commonly arise from Schwann cells of the vestibular nerve sheath in the internal auditory canal and have a natural tendency to extend towards the cerebellopontine angle due to the absence of a bony obstacle. Intralabyrinthine schwannomas are far less common (1% of all vestibular schwannomas), with a prevalence of 0.1–0.4%, and arise from the peripheral intralabyrinthine branches of the vestibulocochlear nerve [1]. Most of the inner ear schwannomas (80%) occur in the cochlea, according to Tieleman et al., especially in the basal and second turn of the cochlea and more frequently in the scala tympani [2]. Vestibular schwannoma is usually not the first diagnosis routinely considered when facing a middle ear mass, as inflammatory pathologies (chronic otitis media, cholesteatomas, cholesterol granulomas…) or other tumours types are more common [1, 3]. The middle ear extension of an intralabyrinthine or internal auditory canal schwannoma is uncommon and the patterns of extension have yet to be specified. We report three new cases of vestibular schwannomas extending to the middle ear. We also analysed all previously published cases of internal auditory canal vestibular schwannomas or intralabyrinthine schwannomas extending in the middle ear and presenting as middle ear masses. The aim is to better understand the potential pathways of extension of such tumours, from the labyrinth towards the middle ear cavity.

✉ Aina Venkatasamy
aina.venkatasamy@chru-strasbourg.fr

1 Service de Radiologie 1, Hôpital de Hautepierre, Hôpitaux Universitaires de Strasbourg, 1 avenue Molière, 67098 Strasbourg CEDEX, France

2 Service de chirurgie ORL, Hôpitaux Universitaires de Strasbourg, 1 avenue Molière, Strasbourg 67098, France

3 Université de Strasbourg, Inserm IRFAC UMR_S1113, Laboratory Stress Response and Innovative Therapies "Streinth", Strasbourg, France

Materials and methods

Case reports

We report three cases of vestibular schwannoma presenting as middle ear masses, imaged by CT and MRI and examined by ear–nose–throat surgeons at our institution.

Data sources and study selection

Pubmed and Google Scholar databases were used to select literature for review and meta-analysis. The languages covered were English, French, Portuguese, Spanish and German. All articles (including original articles, case reports and imaging cases) were reviewed from 1950 to 2018. Thirteen cases have been reported in past publications from 1974 to 2015 [4–15].

Data extraction

Every selected article was read and analysed.

Extracted data (from the three cases we report and selected articles) were:

- The origin of the schwannoma: internal auditory canal and/or inner ear (labyrinth).
- In case of labyrinthine extension, the topography of the tumour: cochlea, vestibule, semi-circular canals.
- The dimension of the middle ear extension of the schwannoma.
- The management of the tumour.
- The patterns of extension of the inner ear tumour towards the middle ear were analysed: round window/oval window/though a bone defect.

Results

Case 1

A 58-year-old woman with right progressive sensorineural hearing loss presented with an acute onset of vertigo. Temporal bone CT showed a hypodense mass filling most of the right tympanic cavity (Fig. 1a). A thin hypodensity was also observed in the postero-superior portion of the walls of the lateral semicircular canal. MRI showed a tumour occupying the entire internal auditory canal, medially extending in the cerebellopontine angle (Fig. 1b). It presented with an intermediate signal on T2-weighted images gradient-echo sequence and a strong gadolinium uptake on T1-weighted images contrast-enhanced sequences. The mass arose from the vestibular nerve and anteriorly displaced the facial nerve. It completely filled the inner ear and reached the middle ear through a small defect in the wall of the lateral semicircular canal and the round window. A translabyrinthine approach was performed and surgery confirmed the lateral semicircular canal wall defect and the extension pathways of the tumour. Histology confirmed the diagnosis.

Case 2

A 57-year-old man presented with progressive right sensorineural hearing loss and chronic instability. Temporal bone CT showed a hypodense nodule at the level of the round window, without any signs of chronic otitis media (Fig. 2a). As the clinical context was misleading, an MRI was performed. MR showed a right vestibular schwannoma occupying the entire internal auditory canal and extending towards the inner (entire labyrinth) and middle ear. The mass reached the middle ear through the round window

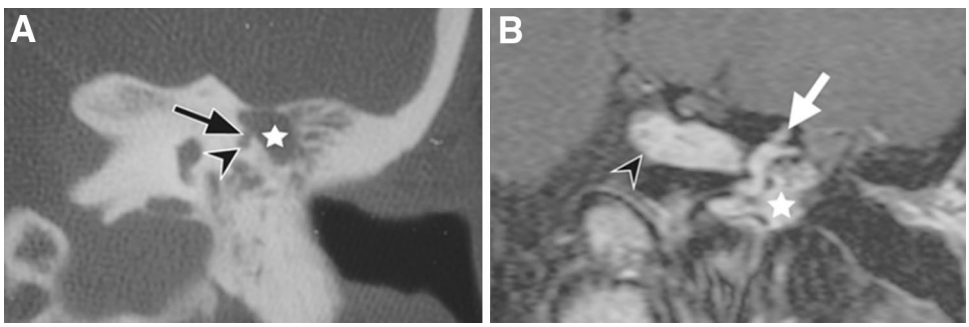

Fig. 1 Case 1, pathologically-proven vestibular schwannoma extending towards the middle ear through the inner ear. **a** Coronal non-enhanced temporal bone multidetector CT showing a hypodense filling of the middle ear cavity (star). A thin hypodensity was also observed (arrow) in the postero-superior portion of the walls of the lateral semicircular canal (arrow head). **b** Coronal T1-weighted contrast-enhanced spin-echo sequence at 1.5 T, acquired 10 min after intravenous gadolinium injection, showing a vestibular schwannoma in the internal auditory canal (arrow head) and extending towards the cerebellopontine angle. The tumour enhanced homogeneously. The tumour extended laterally towards the labyrinth and completely invaded the inner ear (arrow) and extended towards the middle ear where enhancing tumour was observed (star)

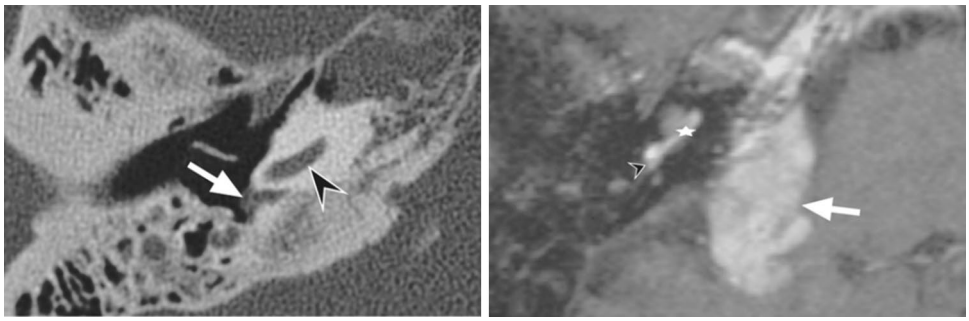

Fig. 2 Case 2, pathologically-proven vestibular schwannoma extending towards the middle ear through the inner ear. **a** Axial non-enhanced temporal bone CT showing a hypodense nodule at the level of the round window (arrow). On CT the cochlea (arrow head) and the rest of the tympanic cavity appeared normal. **b** Axial T1-weighted contrast-enhanced spin-echo sequence acquired at 3 T, 10 min after intravenous gadolinium injection, showing a large vestibular schwannoma in the internal auditory canal, extending towards the cerebellopontine angle (arrow). The tumour also extended in the labyrinth and completely filled the inner ear. The cochlea was completely occupied by the tumour (star) and the tumour extended towards the middle ear through the round window (arrow head)

(Fig. 2b). Medially, the schwannoma extended towards the cerebellum and brainstem. Videonystagmography revealed a caloric areflexia of the lateral semicircular canal and saccadic pursuit, attesting of central damage by compression of the brainstem. There were signs of brainstem compression. As a result, surgery with a translabyrinthine approach was quickly performed and confirmed the diagnosis and the extension through the round window.

Case 3

A 75-year-old man with a history of type 2 neurofibromatosis, presented with left sensorineural hearing loss, which progressively evolved towards complete loss of hearing, associated with left tinnitus and instability. There was no nystagmus, but a Halmagyi test was positive and there was a caloric areflexia of the right lateral canal. The temporal bone CT showed a hypodense middle ear mass filling the round window niche and surrounding the ossicles. The mass also extended towards the Eustachian tube. An MRI showed a left vestibular schwannoma in the internal auditory canal, extending in the cerebellopontine angle (Fig. 3). The tumour completely filled the inner ear and reached the middle ear through the round window. Serous otitis media was secondary to the extension in the Eustachian tube. This patient also presented with multiple schwannomas of other cranial nerves (related to type 2 neurofibromatosis). The patient has been monitored by MRI for a few years and no significant evolution of the tumour was observed afterwards.

Review of the literature

The main characteristics of the three cases we report and the 13 cases of the literature are featured in Table 1.

The three cases we report all arose in the internal auditory canal, which is in agreement with previously published

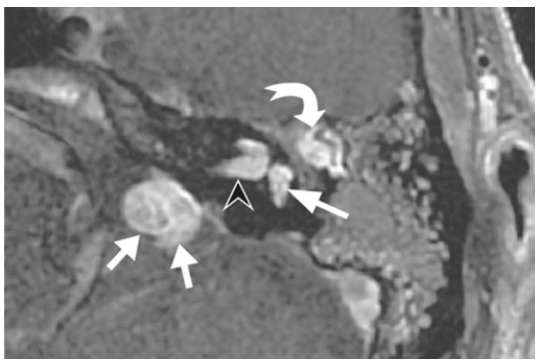

Fig. 3 Case 3, vestibular schwannoma extending towards the middle ear through the inner ear in a patient with type 2 neurofibromatosis. Contrast-enhanced axial T1-weighted sequence showing a vestibular schwannoma in the internal auditory canal (arrow head) and the cerebellopontine angle (double white arrows) extending towards the inner ear, completely filling the cochlea and the vestibule (arrow) and developing in the middle ear (curved arrow), where it surrounds the ossicles

data. In the literature, the majority of vestibular schwannomas extending in the middle ear (77%, $n = 10/13$) arose in the internal auditory canal [4–11, 13], while 23% of them ($n = 3/13$) arose from the labyrinth [12, 14], as illustrated in Fig. 1.

In our cases, all schwannomas filled the entire labyrinth. In previous publications, the authors specified the labyrinthine extension of their tumours in 12 cases (92%): the entire labyrinth was invaded by the tumour in 11 cases (85%) [4–10, 12–14], while Sone et al. only refer to a cochlear extension of their tumour (and no vestibular involvement) [6]. What emerges from these observations, is that the labyrinth had to be filled by the tumour, at least partially (100%, $n = 16$), before the tumour could reach the middle ear cavity. A complete filling of the labyrinth by the tumour was observed in 88% of the cases ($n = 14$).

Table 1 Clinical and imaging characteristics of the three cases we report compared to the 13 other cases reported in the literature

Case number	Author (reference)	Year	Patient (sex, age in years)	Clinical symptoms	Relevant clinical background	Origin of tumour	Extension	Entire labyrinth involvement	Extension to the middle ear through	Dimensions of middle ear extension	Management of middle ear mass
1	Venkatasamy	2019	W, 58	Profound deafness, vertigo	No	IAC	CPA, IAC, C, V, all SCC, ME	Yes	Round window and lateral SCC	12 × 8 × 18 mm (axial × sagittal × craniocaudal)	S + H translabyrinthine approach
2	Venkatasamy	2019	M, 57	Profound deafness, vestibular areflexia	No	IAC	CPA, IAC, C, V, all SCC, ME	Yes	Round window	3 × 4 × 3 mm	S + H translabyrinthine approach
3	Venkatasamy	2019	M, 57	Profound deafness, chronic instability	NF2 (neurofibromatosis)	IAC	IAC, C, V, all SCC, ME	Yes	Round window	16 × 19 × 17 mm	FU
4	Barett [4]	2015	M, 70	SNHL	No	IAC	CPA, IAC, C, V, ME	Yes	Round window	6.6 mm in ME mesotympanum	FU
5	Ebmeyer [5]	2011	W, 48	Unilateral deafness, dizziness, tinnitus	No	IAC	CPA, IAC, C, V, ME	Yes	Round window	Missing data	S + H translabyrinthine approach
6	Sone [6]	2010	W, 53	Profound deafness (related to previous surgeries) no other symptoms	NF2 (neurofibromatosis) History of surgery with translabyrinthine approach for IAC schwannomas	IAC	CPA, IAC, C, ME	No, only the cochlea; no mention of a vestibular extension	Round window	Missing data	S + H Surgical removal of middle ear schwannoma
7	Carreno [7]	2007	W, 41	Liquid flowing from EAC	History of surgery with occipital approach for IAC schwannoma	IAC	IAC, C, V, all SCC, ME, EAC	Yes	Important lysis of the otic capsule Oval window	Missing data	S + H with a transotic approach and total removal of the tumour
8	Yamada [8]	1998	W, 57	Profound deafness, tinnitus, moderate facial palsy	No	IAC	CPA, IAC, C, V, all SCC, ME, EAC	Yes	Not clear	15 × 15 × 20 in total	S + H Two-stage surgical procedure (translabyrinthine and occipital approaches)

Table 1 (continued)

Case number	Author (reference)	Year	Patient (sex, age in years)	Clinical symptoms	Relevant clinical background	Origin of tumour	Extension	Entire labyrinth involvement	Extension to the middle ear through	Dimensions of middle ear extension	Management of middle ear mass
9	Woolford [9]	1994	W, 54	SNHL, facial palsy, decreased trigeminal nerve function and palsies of the lower cranial nerves	Previous surgery with retrosigmoid approach for vestibular schwannoma of the CPA (2 years before)	IAC	All temporal bone, EAC, infratemporal fossa	Yes	Not clear, extended bony lysis	Missing data	S + H
10	Amoils [10]	1992	W, 48	SNHL, tinnitus, vertigo	No	IAC	CPA, IAC, C, V, all SCC, ME	Yes	Round window	20 × 15 mm in the CPA, missing data for middle ear	S + H
11	Stoney [11]	1991	W, 64	Postural instability, pain in the trigeminal nerve territory, SNHL	SNHL resulting from meningoencephalitis	IAC	CPA, IAC, ME. Precise labyrinthine extension not clear	Not clear	Not clear	Missing data	Missing data
12	Mafee [12]	1990	M, 69	Otalgia, tinnitus, SNHL	No	IL	C, V, ME	Yes	Round window	Missing data "small tissue mass in postero-inferior part of ME cavity"	S + H with labyrinthectomy and complete tumour removal
13	Mafee [12]		W, 12	SNHL	No	IL	C, V, ME	Yes	Round window	Missing data	Tympanotomy with biopsies confirming the schwannoma. Extension not noticed on 1st surgery, only confirmed 3 years later by S + H (translabyrinthine approach)

Table 1 (continued)

Case number	Author (reference)	Year	Patient (sex, age in years)	Clinical symptoms	Relevant clinical background	Origin of tumour	Extension	Entire labyrinth involvement	Extension to the middle ear through	Dimensions of middle ear extension	Management of middle ear mass
14	Huy [13]	1987	W, 56	SNHL, vertigo	No	IAC	CPA, IAC, C, V, all SCC, ME, EAC	Yes	Oval Window Bone erosion at the level of the cribliform plate	Missing data	S + H Two surgeries, first an endaural incision with histological confirmation then a retroauricular approach a year later
15	Storrs [14]	1974	W, 70	SNHL, vertigo Sudden SNHL	No	IL	C, V, ME	Yes	Round and oval windows	Missing data	S + H
16	Storrs [14]		M, 63	Fluid effusion in ME	No	IAC	CPA, IAC, C, V, SCC, ME	Yes	Round window	Missing data	S + H. Exploratory myringotomy, then surgery with translabyrinthine removal of the tumour

W woman, *M* man, *SNHL* sensorineural hearing loss, *CPA* cerebellopontine angle, *IAC* internal auditory canal, *IL* intralabyrinthine, *C* cochlea, *V* vestibule, *SCC* semicircular canal, *ME* middle ear, *EAC* external auditory canal, *FU* follow-up, *S + H* surgery + histologically confirmed diagnosis

In our series, the tumour invaded the middle ear through the round window, which is also coherent with previously documented case studies. Eight cases (62%) of round window extension have been described [4–6, 10, 12, 14], giving an overall frequency for round window extension of 69% ($n = 11$). In two cases, by Storrs et al. and Huy et al., the tumour extended in the middle ear through the oval window [7, 13]. In one case, reported by Stoney et al., the lysis was so extended that the initial site of middle ear invasion could not be clearly determined [11]. Lysis of the walls of the otic capsule were also observed by Woolford et al. and Mafee et al., especially in large tumours extending from the cerebellopontine angle to the external auditory canal [9, 14].

Discussion

Middle ear extension of an internal auditory canal or intralabyrinthine schwannoma only occurred after the tumour (partially or completely) filled the labyrinth. The most frequent route from the inner ear to the middle ear appears to be through the round window ($n = 11$, 69%). A probable (hypothetical) explanation for this pattern of extension through the round window rather than the oval one, may be the lower resistance presented by the round window (membranous) compared to the oval window (closed by the footplate), which may represent a stronger mechanical barrier [13, 14, 16]. Another possible explanation for this pattern of spread, is that of an extension by contiguity, as the scala tympani, which is the most common location of intralabyrinthine schwannomas, ends at the round window [2]. In patients with a history of previous middle/inner ear surgeries, a hypothesis could be made that the tumour might have used the surgical defect as a route of extension. Sone et al., disagreed and favoured the round window extension as a preferred route for the middle ear extension of their tumour, even though it had a history of surgical resections [6]. As the otic capsule is very hard, the other middle ear extensions, which did not involve the windows, only occurred through tumour-related defects (secondary to lysis) of the walls of the labyrinth [4].

Conclusion

Middle ear extension of a vestibular schwannoma is a rare occurrence, which requires a careful examination of the round and oval windows on MR imaging, especially as surgery could be one of the therapeutic options. Middle ear extension of vestibular schwannomas only occurred through the labyrinth, after it was filled completely ($n = 14$) or partially ($n = 2$) by the tumour. The round window appears to be the main route (69%) of extension of vestibular schwannomas towards the middle ear.

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Compliance with ethical standards

Conflict of interest The authors have no conflict of interest.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent This is a review article of cases from the literature.

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