Lesions Mimicking Small Vestibular Schwannomas

Matthias Scheich¹ Rudolf Hagen¹ Desiree Ehrmann-Müller¹ Brigitte Bison²
Thomas Günthner-Lengsfeld² Camelia-Maria Monoranu³ Hans-Ullrich Völker³

¹Department of Oto-Rhino-Laryngology, Plastic, Aesthetic and Reconstructive Head and Neck Surgery, University Hospital of Würzburg, Würzburg, Germany

²Department of Neuroradiology, University Hospital of Würzburg, Würzburg, Germany

³Department of Neuropathology, University of Würzburg, Würzburg, Germany

Address for correspondence Matthias Scheich, MD, Department of Oto-Rhino-Laryngology, Plastic, Aesthetic and Reconstructive Head and Neck Surgery, University Hospital of Würzburg, Josef-Schneider-Straße 11, 97080 Würzburg, Germany (e-mail: scheich_m@ukw.de).

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Abstract

Objective Most tumors of the internal auditory canal and cerebellopontine angle (CPA) are vestibular schwannomas (VSs). Preoperative diagnosis is based on typical clinical symptoms and radiological findings. In rare cases, histopathology can, however, show different results.

Design This is a retrospective chart and database review.

Setting The study was conducted at a tertiary skull base referral center at a university hospital.

Participants A total of 207 consecutive cases of VS surgery via the middle cranial fossa approach performed between December 2005 and January 2015 were reviewed.

Main Outcome Measures The main outcome measures were definitive histologic findings in 198 specimens, analysis of preoperative magnetic resonance imaging (MRI) and computed tomography.

Results Histopathology revealed three meningiomas and two cases of lipochoristomas. Clinical presentation was typical for VS in all five cases. In preoperative MRI, all tumors were suspected to be VSs. Retrospective analysis of the preoperative imaging did not lead to a modification of the diagnosis. Intraoperative findings showed increased adherence of the tumor to the adjacent tissue in two of the five cases.

Conclusion CPA lesions other than VSs are unusual but have to be taken into account. In very small tumors, imaging still remains difficult.

Keywords

- histology
- vestibular schwannoma
- imaging
- small tumor
- middle cranial fossa approach

Introduction

Vestibular schwannomas (VSs) are rare slow-growing benign neoplasms of the cerebellopontine angle (CPA) and the internal auditory canal (IAC). They originate from the Schwann cells of the vestibular portion of the eighth cranial nerve.¹ The annual incidence is calculated to be 2 per 100,000.² Due to increased availability of modern diagnostic tools (auditory brainstem response [ABR], magnetic resonance imaging [MRI]), the number of incidental findings has consistently risen.^{2,3} The

diagnosis is usually based on the combination of typical clinical symptoms and imaging findings. Early detection of VS allows the patient to benefit from modern and improved management strategies. Today patients can choose among microsurgery, radiotherapy, and wait-and-scan protocols. Fortunately, malignancy is scarce and more often associated with typical radiological findings so that nonsurgical procedures without histology are possible. Nevertheless, only surgery provides the ability of histopathological examination of tumor tissue to confirm the final diagnosis.

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Materials and Methods

Patient Data

Out of a series of 207 consecutive patients treated in our department between December 2005 and January 2015 for suspected VS, 198 histologic specimens were available for further examination. In the missing cases, no material was sent to the pathology department or sections were actually not available for further review. We extracted patient data from the records and reviewed preoperative imaging.

There were 97 male and 101 female subjects aged 19 to 78 years (mean 52). All patients had undergone microsurgery via the middle cranial fossa (MCF) approach. Tumor stages were T1 (intracanalicular) in 82 cases and T2 (extrameatal part without contact to the brain stem) in 116 patients. We compared the 5 cases of non-VS to the 193 confirmed VS cases and tried to analyze the differences in preoperative findings.

Histopathology

All tissue specimens were routinely stained with hematoxylin and eosin (HE). Complementary immunohistochemistry usually includes S-100 and Ki-67 to confirm VSs as well as epithelial membrane antigen (EMA) to prove or exclude meningiomas. In some cases, we also used vimentin, pancytokeratin, CD56, EvG, actin, desmin, Ladewig, Kluver-Barrera, and periodic acid-schiff (PAS).

Imaging

In all cases, imaging was discussed in an interdisciplinary skull-base conference with a neuroradiologist prior to decision making. All tumors were suspected to be VS. The imaging of the non-VS cases was additionally analyzed by two different neuroradiologists.

Tumor size was measured in three spatial dimensions with the tumor length defined as the largest diameter across the whole tumor including the IAC. Tumor volume was calculated as a triaxial ellipsoid.

Compliance to Ethical Standards

The study was approved by the local ethics committee.

Table 1 Preoperative characteristics

	Tumor stage	Tumor volume (mm ³)	Tumor length (mm)	Age (y)	Sex
193 vestibular schwannomas	80 T1 113 T2	212	10	52	96 m 97 f
Meningioma 1	T2	220	12	70	f
Meningioma 2	T2	136	13	62	f
Meningioma 3	T2	440	15	50	f
Lipochoristoma 1	T1	25	4	38	m
Lipochoristoma 2	T1	42	5	52	f

Results

Final Histological Findings

In 193 tumors, histopathology confirmed the diagnosis of a VS. Five non-VS cases were diagnosed as three cases of meningioma (► Fig. 1a) and two cases of lipochoristoma (► Fig. 1b, c).

Preoperative Clinical Findings

The group of 193 VS patients consisted of 80 T1 and 113 T2 tumors with a mean tumor volume of 212 mm³ (range: 10–1,347 mm³; median: 151 mm³) and a mean tumor length of 10 mm (range: 4–25 mm; median: 10 mm). The five non-VS tumors were two cases of T1 and three cases of T2 tumors with a median volume of 136 mm³ (25–440 mm³) and a median tumor length of 12 mm (4–15 mm) (► Table 1).

There were 96 men and 97 women in the group of VS patients, with a mean age of 52 years (range: 19–78 years; median: 51 years), whereas the non-VS patients (1 man, 4 women) had a mean age of 55 years (range: 38–70 years; median 52 years).

The VS patients complained of hearing loss for an average of 26 months ($n = 125$ patients), tinnitus for 25 months ($n = 107$ patients), and dizziness for 14 months ($n = 90$ patients) before surgery. In the non-VS group, three patients complained of hearing loss for 19 months, one patient suffered from tinnitus for 36 months, and three patients had dizziness for 13 months. In both groups, there were no cases of preoperative facial nerve symptoms.

Fig. 1 Final histopathological findings: (a) meningioma (EMA ×400); (b) lipochoristoma with few adipocytes (HE ×25); and (c) lipochoristoma with plenty of adipocytes (HE ×25).

Fig. 2 Meningioma 2: (a) axial MRI (T1): intra + extrameatal tumor of the right side (arrow). (b) Axial MRI (T1CE): intrameatal part of the tumor. (c) Axial MRI (T1CE): extrameatal part of the tumor. (d) Coronal CT without calcification in the IAC/CPA. CPA, cerebellopontine angle; CT, computed tomography; IAC, internal auditory canal; MRI, magnetic resonance imaging; T1CE, T1 with contrast enhancement.

We could not find any differences between the five non-VS patients compared with the 193 VS patients in audiological testing (pure tone audiometry, speech discrimination test, AAO-HNS grade, ABR) or in facial nerve function (House-Brackman score).

Analysis of Preoperative Imaging

Two independent neuroradiologists reviewed all available preoperative images. The first meningioma patient had presented with three previous external MRIs. The fourth MRI was done with 1.5 Tesla in our neuroradiology department in 2004, but at that time, only film copies were archived. We

found a typical tumor with extrameatal extent and strong dental artifacts. We were not able to distinguish between VS and meningioma with the films of that of last MR. All images of this patient had been discussed in four skull-base conferences and were diagnosed to be a VS. The second meningioma patient (→Fig. 2) presented with two consecutive external MR images within 6 months that showed a growing tumor. T1 images with contrast enhancement (T1CE) demonstrated a tumor without dural tail. The temporal bone computed tomography (CT) did not show any calcification of the lesion and no hyperostosis or hyperostotic spurs were visible. The third meningioma patient (→Fig. 3) also presented with typical

Fig. 3 Meningioma 3: (a) coronal MRI (T1CE): intrameatal part. (b) Axial CT without calcification in the IAC/CPA. (c) Coronal MRI (T1CE): extrameatal part. CPA, cerebellopontine angle; CT, computed tomography; IAC, internal auditory canal; MRI, magnetic resonance imaging; T1CE, T1 with contrast enhancement.

Fig. 4 Lipochoristoma 1: (a) axial MRI (T1CE): small intrameatal tumor (arrow). (b) Coronal MRI (T1CE fat saturated): small intrameatal tumor. (c) Axial MRI (T1CE fat saturated): small intrameatal tumor. MRI, magnetic resonance imaging; T1CE, T1 with contrast enhancement.

signs of an intra- and extrameatal VS on the external MRI. T1CE images showed no dural tail, and CT scans did not detect calcification or bone alterations as previously described.

The first patient with a lipochoristoma (►**Fig. 4**) had received two external MR scans within 6 months, which were also demonstrated as being typical for VS. Retrospective analysis showed that the T1 images without contrast were too thick (5 mm) and only axial planes were available. The small intrameatal tumor was clearly visible on the T1CE images with fat saturation.

The second patient with a lipochoristoma (►**Fig. 5**) also had thick T1 precontrast and T2 slices (6 mm) in external imaging. The T1CE slices showed a small-enhanced intracanalicular tumor. This tumor was also still bright on T1CE images with fat saturation.

Intraoperative Findings

According to the surgeons report, there were no abnormalities during surgery in three of the non-VS cases. In one lipochoristoma and in one meningioma, extensive adherence of the tumor to the surrounding neurovascular tissue was described.

Discussion

In this series of 198 histologic results of small tumors of the CPA, five specimens (2.5%) were found not to be a VS as suspected preoperatively. Neoplasms of the CPA account for ~8 to 10% of all intracranial tumors. Most tumors (81–91%) of the CPA are VS.^{4–7} Small tumors of the IAC typically present with hearing loss, tinnitus, and dizziness,^{8,9} which were also

Fig. 5 Lipochoristoma 2: (a) axial MRI (T1CE): small intrameatal tumor, lateral part (arrow). (b) Axial MRI (T1CE): small intrameatal tumor, medial part (arrow). (c) Coronal MRI (T1CE fat saturated): small intrameatal tumor (arrow). (d) Axial CT without calcification in the IAC/CPA. CPA, cerebellopontine angle; CT, computed tomography; IAC, internal auditory canal; MRI, magnetic resonance imaging; T1CE, T1 with contrast enhancement.

Table 2 MRI protocol

Entire endocranium FLAIR/T2 axial 5 mm T1 with CE axial 5 mm
Temporal bone T1 with and without CE axial ≤ 3 mm T1 with CE coronal ≤ 3 mm High-resolution T2 (e.g., CISS) with CE
Additional T1CE fat-saturated coronal or axial ≤ 3 mm if fat is suspected DWI in suspicion of cholesteatoma MR angiographic sequences if vascular lesions are detected T1CE 3D dataset depending on individual circumstances

Abbreviations: CE, contrast enhancement; CISS, constructive interference in steady state; FLAIR, fluid attenuation inversion recovery; DWI, diffusion-weighted images; MRI, magnetic resonance imaging; 3D, three-dimensional.

the leading complaints in our 193 confirmed VS patients for an average period of 12 to 23 months prior to surgery.

On MRI, VSs typically show T1 isointensity and T2 high signal intensity with strong enhancement after gadolinium injection. On heavily T2-weighted images, they replace the fluid signal of CSF. In CT scans, VSs are isodense or slightly hypodense and may present a widening of the IAC.

Histopathology of VSs usually shows two distinct patterns in HE staining: Antoni A and B. Additional immunohistochemistry to confirm the diagnosis shows a low proliferation index of Ki-67 and a strong expression of S-100.

Meningioma Cases ($n = 3$)

Meningiomas comprise 15 to 20% of all intracranial neoplasms. They typically occur parasagittal/falcine and only in rare cases in the CPA.¹⁰ Nevertheless, meningiomas represent 3 to 12% of the CPA cases and are known to be the second most nonschwannoma tumor in the CPA.^{4,5,7,11} All of our meningiomas consisted of an intra- as well as an extrameatal part (stage T2). Meningiomas limited to the IAC are rare. Bohrer and Chole¹² presented one intra- and one intra/extrameatal meningioma and extracted four more purely intracanalicular cases from the literature prior to 1996.

Unlike Mallucci et al,¹³ who stated that patients with meningiomas presented with different symptoms compared with VS patients, we found hearing loss in two of the three cases and dizziness in all three cases. This can easily be

explained by the close vicinity of the tumors to the vestibulo-cochlear nerve in the IAC.

Meningiomas are, similar to VSs, isointense on T1 MRI with strong enhancement after gadolinium injection, but differ with isointensity on T2-weighted MR images. They can typically show increased thickness of the dura, presenting as a dural tail (so-called meningeal tail sign), which was not present in our three cases. Typical calcifications in CT scans are found in 20 to 50% of the meningiomas. Hyperostosis of the adjacent bone or a hyperostotic spur is possible as well, but none of these signs was visible in our three cases. This difficulty in distinguishing between VS and meningioma has also been described in a case report by Grauvogel et al in 2010,¹⁴ in which they demonstrated that preoperative imaging could not differentiate that a tumor was a coexistence of a VS and a meningioma.

Histopathological appearance of meningiomas can be variable. Within the group of benign meningiomas (WHO class I), there are even nine different subtypes described. If complementary immunohistochemistry shows no expression of S-100 but a strong expression of EMA, a meningioma can be diagnosed.

Lipochoristoma Cases ($n = 2$)

Lipomatous tumors of the IAC are very rare CPA tumors ($<1\%$)^{5,15,16} and it is considered that they are congenital malformations.¹⁶ Recent literature has found the term lipochoristoma to be more accurate than lipoma because these tumors consist of adipose cells between nerve fibers mixed up with fibrous tissue.⁴

On MRI, lipochoristomas usually present isointense to fat with high signal on T1 and intermediate signal on T2. They do not enhance gadolinium and usually show a signal loss on fat-suppressed images. There are several case reports of IAC lipochoristomas. In most cases, fat saturation sequences led to the correct diagnosis and conservative treatment was offered.^{15,17,18} In both of our lipochoristoma cases, only thick T1 slices were available and the tumor was classified to be isointense. We might have identified them to be hyperintense, if T1 thin slice images would have been available. Since both tumors in our series showed an uptake of contrast and were still visible in fat suppression, they were misdiagnosed as a VS. Bacciu et al¹⁹ also reported on an uptake of contrast agent in four of eight lipomas of the CPA which led to the misdiagnosis of VS. The uptake of contrast agent and a persisting signal in fat suppression is not typical for lipomas (that typically solely consist of adipocytes), but it may be explained by the varied proportions of fat in relation to the surrounding nerve fibers in our lipochoristoma cases.

Table 3 MR characteristics of tumors of the internal auditory canal and cerebellopontine angle

	T1 weighted	T2 weighted	Contrast enhancement	Fat suppression
Schwannoma	Isointense	Hyperintense	Strong	No effect
Meningioma	Isointense	Isointense	Strong	No effect
Lipochoristoma	Hyperintense	Intermediate	No	Signal loss

Abbreviation: MR, magnetic resonance.

In a large series of 84 “lipomas” of the IAC and CPA, Bigelow et al¹⁵ reported hearing loss, dizziness, and tinnitus to be the most common symptoms. This corresponds to our findings of preliminary symptoms as well, but as in the meningioma cases, these are typical unspecific symptoms of all growing lesions in the IAC caused by compression of the cranial nerves inside the bony canal.

Histology usually confirms mature fat cells and connective tissue. IAC lipochoristomas typically show an infiltration between nerve fibers,^{12,15,19,20} which makes them more difficult and unfavorable to resect.^{19,21,22} This effect was also noted during surgery in one of our two lipochoristomas, where a strong adherence of the tumor to the surrounding tissue was encountered. This may explain why both lipochoristoma cases showed functional results below the average compared with the meningioma or schwannoma cases.

Other Tumors

Calzada et al⁴ or Brackmann and Bartels⁵ described facial nerve schwannomas which can also occur in the IAC/CPA region in ~1% of the cases. We did not reveal any facial nerve schwannomas by chance intraoperatively during the reported period of 207 consecutive MCF cases. We also did not detect any of the less frequent reported lesions such as cholesteatomas, hemangiomas, or malignomas.

Magnetic Resonance Imaging Recommendations

It is necessary to follow a standardized solid MRI protocol in diagnostics of the CPA. However, even in the “best” imaging series, we cannot always distinguish between VS and other lesions. There is no general recommendation, but we chose to use the specifications in ►Table 2 following the example of De Foer et al.²³ In ►Table 3, we summarize the typical characteristics of IAC tumors in MR diagnostics.

Limitations of the Study

Of course, this is only a retrospective analysis of preoperative imaging. As we operated on small VS in early stages, in some cases, more follow-up imaging may have led to another diagnosis. The study clearly shows that it may have been helpful to be more accurate with “inadequate” external MRI.

Of course, the knowledge of the final histology would have changed the therapy in the lipochoristomas toward observation.

Conclusion

Small lesions in the CPA and the IAC other than VSs are unusual but have to be taken into account. This should at least be kept in mind, especially in cases of radiotherapy or observation without proof of histology. In very small tumors, imaging still remains difficult. This is why patients should be provided with very detailed instructions as to what is expected from external imaging, or you should consider new imaging from “your” neuroradiologist experienced in skull base lesions.

Conflict of Interest

All authors declare that they do not have any conflict of interest.

Note

The study was presented at the 12th Congress of the European Skull Base Society 2016 in Berlin, Germany.

References

- 1 Roche PH, Bouvier C, Chinot O, Figarella-Branger D. Genesis and biology of vestibular schwannomas. In: Regis J, Roche PH, eds. *Modern Management of Acoustic Neuroma*. Vol. 21. Basel, Paris, London, New York, Sydney: Karger; 2008:24–31
- 2 Stangerup SE, Tos M, Thomsen J, Caye-Thomasen P. True incidence of vestibular schwannoma? *Neurosurgery* 2010;67(05):1335–1340, discussion 1340
- 3 Stepanidis K, Kessel M, Caye-Thomasen P, Stangerup SE. Socio-demographic distribution of vestibular schwannomas in Denmark. *Acta Otolaryngol* 2014;134(06):551–556
- 4 Calzada AP, Go JL, Tschirhart DL, Brackmann DE, Schwartz MS. Cerebellopontine angle and intracanalicular masses mimicking vestibular schwannomas. *Otol Neurotol* 2015;36(03):491–497
- 5 Brackmann DE, Bartels LJ. Rare tumors of the cerebellopontine angle. *Otolaryngol Head Neck Surg* (1979) 1980;88(05):555–559
- 6 Moffat DA, Saunders JE, McElveen JT Jr, McFerran DJ, Hardy DG. Unusual cerebello-pontine angle tumours. *J Laryngol Otol* 1993; 107(12):1087–1098
- 7 Grey PL, Moffat DA, Hardy DG. Surgical results in unusual cerebellopontine angle tumours. *Clin Otolaryngol Allied Sci* 1996; 21(03):237–243
- 8 Geyer G, Helms J. Subjective and objective symptoms before and after treatment of small acoustic neuromas (in German). *Med Welt* 1985;36:38–41
- 9 Lee SH, Choi SK, Lim YJ, et al. Otologic manifestations of acoustic neuroma. *Acta Otolaryngol* 2015;135(02):140–146
- 10 DeMonte F, Al-Mefty O. Meningiomas. In: Kaye AH, Law ER Jr, eds. *Brain Tumors*. New York: Churchill Livingstone; 1995:675–704
- 11 Kohan D, Downey LL, Lim J, Cohen NL, Elowitz E. Uncommon lesions presenting as tumors of the internal auditory canal and cerebellopontine angle. *Am J Otol* 1997;18(03):386–392
- 12 Bohrer PS, Chole RA. Unusual lesions of the internal auditory canal. *Am J Otol* 1996;17(01):143–149
- 13 Mallucci CL, Ward V, Carney AS, O'Donoghue GM, Robertson I. Clinical features and outcomes in patients with non-acoustic cerebellopontine angle tumours. *J Neurol Neurosurg Psychiatry* 1999;66(06):768–771
- 14 Grauvogel J, Grauvogel TD, Taschner C, Baumgartner S, Maier W, Kaminsky J. A rare case of radiologically not distinguishable coexistent meningioma and vestibular schwannoma in the cerebellopontine angle - case report and literature review. *Case Rep Neurol* 2010;2(02):111–117
- 15 Bigelow DC, Eisen MD, Smith PG, et al. Lipomas of the internal auditory canal and cerebellopontine angle. *Laryngoscope* 1998; 108(10):1459–1469
- 16 Wu SS, Lo WW, Tschirhart DL, Slattery WH III, Carberry JN, Brackmann DE. Lipochoristomas (lipomatous tumors) of the acoustic nerve. *Arch Pathol Lab Med* 2003;127(11):1475–1479
- 17 Filli L, Huber A, Husain NA. Symptomatic lipoma of the internal auditory canal: CT and MRI findings. A case report. *Neuroradiol J* 2014;27(04):479–481
- 18 Mukherjee P, Street I, Irving RM. Intracranial lipomas affecting the cerebellopontine angle and internal auditory canal: a case series. *Otol Neurotol* 2011;32(04):670–675

- 19 Bacciu A, Di Lella F, Ventura E, Pasanisi E, Russo A, Sanna M. Lipomas of the internal auditory canal and cerebellopontine angle. *Ann Otol Rhinol Laryngol* 2014;123(01):58–64
- 20 Christensen WN, Long DM, Epstein JI. Cerebellopontine angle lipoma. *Hum Pathol* 1986;17(07):739–743
- 21 Dazert S, Aletsee C, Brors D, et al. Rare tumors of the internal auditory canal. *Eur Arch Otorhinolaryngol* 2005;262(07):550–554
- 22 White JR, Carlson ML, Van Gompel JJ, et al. Lipomas of the cerebellopontine angle and internal auditory canal: primum non nocere. *Laryngoscope* 2013;123(06):1531–1536
- 23 De Foer B, Carpentier K, Bernaerts A, Kenis C, Casselman J, Offeciers E. Imaging of cerebellopontine angle and internal auditory canal lesions. In: Lemmerling M, De Foer B, eds. *Temporal Bone Imaging*. Berlin, Heidelberg: Springer; 2015:155–218