Meningiomas of the Internal Auditory Canal: Two Case Reports

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ABSTRACT

Meningiomas represent 3 to 12% of the tumors that involve the cerebellopontine angle and internal auditory canal (IAC). Intracanalicular meningiomas, however, are rare. Only 10 well-documented cases have been reported in the English literature. The differential diagnosis includes acoustic neuromas, facial nerve neuromas, hemangiomas, lipomas, and meningiomas. We report two new cases of intracanalicular meningioma. Both patients had unilateral tinnitus. In case 1, the patient also experienced sensorineural hearing loss with unilateral tinnitus. In case 1, the lesion appeared hyperintense on T1-weighted and hypointense on T2-weighted magnetic resonance images. In case 2, the patient had an enhancing lesion in the right IAC. The tumors were moderately cellular, and meningeal whorls and meningothelial inclusions were present. Immunohistochemical staining revealed the tumors to be positive for epithelial membrane antigen (EMA) and negative for S-100 protein. The radiologic findings that help distinguish a meningioma from other lesions are reviewed.

KEYWORDS: Meningioma, internal auditory canal, acoustic neuroma

Almost 10% of all intracranial tumors arise in the cerebellopontine angle (CPA) and internal auditory canal (IAC).^{1,2} Acoustic neuromas and meningiomas represent 90% and 3 to 12%, respectively, of the tumors involving these sites. Intra-

canalicular meningiomas are rarely diagnosed.¹⁻¹⁴ We report two new cases of intracanalicular meningiomas and discuss the diagnostic and therapeutic issues related to these tumors and their site of origin.

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CASE REPORTS

Case 1

The physical examination of a 50-year-old man with a 2-year history of bilateral hearing loss was unremarkable. Only a mild bilateral sensorineural hearing loss could be detected. After 2 months, his symptoms worsened. Pure-tone audiometry documented a more evident hearing loss on the right, and the patient complained of an annoying tinnitus on the right. Impedance testing showed a type "A" tympanogram and normal stapedial reflex. The latencies of waves I, II, III, and V were delayed on recordings of auditory potentials. Magnetic resonance imaging (MRI) showed a mass located in the IAC that appeared hyperintense and hypointense on T1- and T2-weighted images, respectively. The tumor was 1.1 cm $\times$ 0.5 cm and did not extend into the CPA (Figs. 1 and 2).

A retrosigmoid approach combined with partial labyrinthectomy was planned to remove the lesion and to preserve hearing function. The tumor reached the fundus of the meatus, and the diffi-

culty of removing the tumor obliged us to a trans-labyrinthine approach to achieve complete tumor removal. The patient's immediate postoperative facial function was grade II according to the House-Brackmann grading system. During the next 48 hours, facial-nerve function deteriorated to grade III. Two months after surgery, facial-nerve function improved to grade I. The tumor was moderately cellular. Several meningeal whorls and nuclear pseudoinclusions were present. On immunohistochemical staining, the tumor was S-100 protein negative. Histologic analysis identified a meningioma. The patient has remained disease free 1 year after surgery.

Case 2

The physical examination of a 56-year-old woman with a 6-month history of tinnitus in the right ear was unremarkable. Her physical examination was unremarkable. Pure-tone audiography was normal. MRI with gadolinium enhancement showed an enhancing lesion in the right IAC with minimal

Figure 1 Right intracanalicular meningioma appears hyperintense on axial T1-weighted magnetic resonance image.

Figure 2 Right intracanalicular meningeoma appears hyperintense on coronal T1-weighted magnetic resonance image.

extension into the CPA. The tumor was 1.2×0.5 cm (Fig. 3).

A retrosigmoid approach was performed. Intraoperatively, the lesion was compressive and adherent to cranial nerves VII and VIII, but the tumor was removed totally. The patient's preoperative

hearing function was preserved with only a slight (20 dB) hearing loss at 4000 and 8000 Hz. Immunohistochemical staining revealed the tumor to be epithelial membrane antigen (EMA)-positive and S-100 protein negative. Histologically, the lesion was diagnosed as an intracanalicular meningeoma (Figs.

Figure 3 Right intracanalicular meningeoma appears hypointense on axial T2-weighted magnetic image.

Figure 4 Specimen from an intracanalicular meningioma shows several meningeal whorls and meningotheelial inclusions in the tumor cells (hematoxylin and eosin, $\times 10$).

4 and 5). Grade I facial function was maintained. The patient has had no recurrence after 2 years of follow-up.

DISCUSSION

The cases presented here illustrate that meningiomas can arise solely within the IAC. Before the advent of MRI, meningiomas usually were detected when they were quite large and involved the IAC only secondarily.^{4,9} Clinically, these tumors manifest during the fourth to sixth decades and occur more frequently in women than men, with a 5:2.3 ratio.^{9,13,15} Their etiology is unknown, but meningiomas may be associated with neurocutaneous syndromes such as neurofibromatosis and damage to the NF2 gene located on chromosome 22.^{9,16,17} Meningiomas are well-encapsulated tumors and may be coarsely or finely nodular. Their consistency is usually firm or rubbery.^{3,4,9,10} Histologically, the tumor has meningeal whorls and meningotheelial inclu-

sions.⁹ Immunohistochemical analysis is devoted to the detection of EMA and S-100 neuronal protein. Histologic analysis is essential for differential diagnosis between meningiomas and acoustic neuromas. Meningiomas are EMA positive, whereas schwannomas are S-100 neuronal protein positive.⁹

Although these tumors tend to invade dura and bone, they are considered benign. The recurrence rate of meningiomas, however, can be high.^{3,4,9,10} They originate from the arachnoid villi, which are found in high concentration at cranial nerve exit foramina, thus explaining their occurrence in the CPA and IAC.^{1-4,9,10,15} Usually meningiomas have been diagnosed when they are large and typically have a broad base of attachment along the posterior surface of the petrous bone.⁴

Only 10 well-documented cases of solely intracanalicular meningiomas have been reported (Table 1).^{3,4,9-12} Their clinical manifestations have related primarily to compression and reactive edema rather than to parenchymal involvement.⁴ Consequently, these tumors tend to become symptomatic when their size increases or when they arise in a re-

Figure 5 Specimen from an intracanalicular meningioma shows several meningeal whorls and meningotheelial inclusions in the tumor cells (hematoxylin and eosin, $\times 25$).

stricted space, such as the bony portion of IAC.^{4,9,10} In these situations, the tumors become symptomatic early. Of the 10 well-documented cases of intracanalicular meningiomas, eight patients (80%) had unilateral sensorineural hearing loss alone or associated with tinnitus, fullness, vertigo, and facial palsy. One patient (10%) complained of tinnitus only, and another patient (10%) became symptomatic with vertigo and aural fullness. Our two cases were similar. The first patient experienced sensorineural hearing loss, and the second had unilateral tinnitus.

Pure-tone audiometry showed unilateral sensorineural hearing loss in eight (80%) of the reported patients and was normal in two patients (20%). Of the latter, one patient's audiometric response to speech was altered. Auditory evoked responses were recorded from only three patients, all of whom exhibited a deterioration in latency and an altered waveform morphology. Our experience supports these findings. The patient in case 2 did not exhibit specific characteristics. The diagnosis in case 1 was delayed because of the initial symmetric bilateral hearing loss.

Magnetic resonance imaging is the imaging modality of choice for the detection of small tumors (<2 mm) in the IAC.^{1-4,9} The differential diagnosis includes other pathologies that can originate in the IAC: acoustic neuromas, meningiomas, arachnoid cysts, hemangiomas, and lipomas.^{3,6} Based on the typical appearance of these lesions on MRI (Table 2), hemangiomas and lipomas can be diagnosed correctly before surgery. Hemangiomas appear hyperintense on both T1- and T2-weighted images, whereas lipomas appear hyperintense on T1- and hypointense on T2-weighted MRIs.³ Arachnoid cysts appear similar on MRI T1- and T2-weighted images, but their fluid-filled structure can be detected.³ Acoustic neuromas represent the most difficult differential diagnosis. Although meningiomas may arise eccentric to the IAC and may be hypointense on T1- and hyperintense on T2-weighted images and associated with less enhancement than neuromas, the rate of misdiagnosis results is high.^{3,9} Because MRI is insufficient to differentiate meningiomas from neuromas, these lesions must be diagnosed by histologic analysis.

Table 1 Summary of Clinical Findings Associated with Reported Intracanalicular Meningiomas and Their Surgical Outcome

Author	Age/ Sex	Symptoms	Surgical Approach	Outcome
Singh et al. ¹¹	14/M	Hearing loss, facial palsy	Middle fossa	Total facial n paralysis
Brookler et al. ¹²	70/F	Vertigo, tinnitus, hearing loss	Translabyrinthine	VII normal
Langman et al. ¹⁰	54/M	Hearing loss	Translabyrinthine	VII normal
Bohrer and Chole ³				
Case 1	52/M	Tinnitus	Middle fossa	VII and VIII normal,
Case 2	66/F	Vertigo, hearing loss	Translabyrinthine	VII normal
Zeitouni et al. ⁹	60/F	Fullness, disequilibrium	Translabyrinthine	VII normal
Caylan et al. ⁴				
Case 1	42/M	Fullness, hearing loss, vertigo	Translabyrinthine	VII normal
Case 2	46/M	Hearing loss	Translabyrinthine	VII normal
Dinh et al. ¹³	53/F	Vertigo, tinnitus, hearing loss hemifacial spasm	Suboccipital	VIII normal, partial facial palsy
Martinez Devesa et al. ¹⁴	48/F	Hearing loss, tinnitus, disequilibrium	Translabyrinthine	Facial nerve palsy
This report	50/M	Hearing loss,	Translabyrinthine	VII normal
	56/F	tinnitus	Retrosigmoid	VII and VIII normal

Any intrameatal lesion should undergo biopsy intraoperatively to ensure appropriate treatment.

Surgery is influenced by both the challenging differential diagnosis and the high recurrence rate of meningiomas. Reported recurrence rates range between 7 and 20% for intracranial meningiomas that were completely removed.^{18,19} In contrast, the reported recurrence rate for acoustic neuromas is 0.8 to 1%.²⁰⁻²⁴ These data confirm the importance of obtaining an intraoperative biopsy.

When hearing function is already lost, ablative techniques are the first treatment choice.

Treatment when hearing is normal or impaired is controversial. In these cases, we suggest starting with a conservative approach, such as the retrosigmoid or middle fossa approach, and obtaining an intraoperative histologic examination to identify the lesion. The translabyrinthine and retrosigmoid approaches extended to the IAC with removal of the vestibule and semicircular canals if the tumor reaches the fundus afford satisfactory results with very low risk of recurrence. The surgeon must not hesitate to sacrifice hearing function if incomplete removal of a meningioma is detected or suspected.

Table 2 Magnetic Resonance Imaging Signal Characteristics of Intracanalicular Tumors

Tumor	T ₁	T ₂	Gd Enhancement
Neuroma	Hypoiso intense	Isohyperintense	Yes
Meningioma	Hypoiso intense	Hyperintense variable	Yes
Arachnoid cyst	Hypointense	Hyperintense	No
Hemangioma	Hyperintense	Hyperintense	Variable
Lipoma	Hyperintense	Hypointense	No

Gd, gadolinium.

Figure 6 Proposed algorithm for the treatment of internal auditory canal (IAC) meningiomas.

Treatment of a patient with unilateral hearing is also controversial. Because the surgical risk of compromising the patient's remaining hearing is high,²⁵ close observation with serial MRI every 6 months is advisable. Radiosurgery is another option, but the risk of incurring a trigeminal deficit, facial weakness, or a subjective reduction or loss of hearing (40%) contraindicates this particular modality as the first choice in this particular condition.^{26–28} With evidence of tumor growth, radiosurgery may play an alternative role to microsurgery. When tumor growth induces serious neurologic deficit, surgery is recommended, but the need to preserve unilateral hearing favors conservative procedures as much as possible (Fig. 6).

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Commentary

This is an interesting case report of two patients with intracanalicular meningiomas. The internal auditory canal is a rare location for meningiomas: only 10 cases have been documented in the English surgical literature. Certainly by viewing preoperative magnetic resonance images alone, clinicians would presume that these patients had acoustic neuromas primarily because of the intracanalicular location of the lesions and the characteristic symptoms of hearing loss and tinnitus. As the authors note, the clinical significance of histologic identification is quite important because the meningiomas do not invade the nerve but have a higher recurrence rate than neuromas because they may involve the adjacent dura or even bone. The authors note the importance of obtaining intraoperative samples for frozen-section specimens. Simply noting a spindle cell tumor on frozen section may be insufficient. This report serves to remind us of the importance of obtaining a clear differential diagnosis of cerebellopontine masses.

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