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Osteoma of the Internal Auditory Canal Mimicking Vestibular Schwannoma: Case Report and Review of 17 Recent Cases

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Osteoma of the internal auditory canal (IAC) is an uncommon benign bone tumor. Its imaging features may be similar to other IAC lesions, such as vestibular schwannomas that are benign and usually slow-growing but sometimes life-threatening tumors. Thus, detecting IAC lesions and differentiating osteoma from other IAC lesions are both important clinically. We report a case of misdiagnosis of an IAC osteoma as an IAC schwannoma based on magnetic resonance (MR) imaging using the three-dimensional constructive interference in steady state (CISS) sequence instead of T1-weighted MR imaging with gadolinium. We also review 17 cases of IAC osteomas reported in the past 22 years. A 61-year-old female was admitted to our department with IAC lesion incidentally discovered by the CISS sequence. The lesion was diagnosed as an IAC schwannoma, and was followed up annually under "wait and scan" management. Follow-up T1-weighted MR imaging with gadolinium showed no enhancement of the tumor, and additional computed tomography (CT) of the temporal bone showed a solitary pedunculated bony lesion, resulting in the diagnosis of IAC osteoma. The CISS sequence is useful for detecting small IAC lesions, such as vestibular schwannomas. However, the CISS sequence has limitations for qualitative diagnosis and can misdiagnose osteomas as schwannomas. Use of the CISS sequence without T1-weighted MR imaging with gadolinium for the screening of a lesion of the IAC and cerebellopontine angle should consider the possibility of IAC osteomas, and temporal bone CT or T1-weighted MR imaging with gadolinium should be performed when an IAC lesion is detected.

Keywords: constructive interference in steady state; diagnosis; internal auditory canal; magnetic resonance imaging; osteoma
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Introduction

Osteoma is a type of slow-growing benign bone tumor which is commonly observed in the temporal bone, especially in the mastoid cortex and external auditory canal (Graham 1979; Ramsay and Brackmann 1994). However, osteoma of the internal auditory canal (IAC) is very rare (Ramsay and Brackmann 1994; Baik et al. 2011), and is usually discovered incidentally because the tumor remains asymptomatic (Liétin et al. 2010). Computed tomography (CT) is the most effective diagnostic modality that can reveal apparent dense bony lesion that is sometimes missed by magnetic resonance (MR) imaging (Davis et al. 2000). In the last decade, MR imaging has become a popular diagnostic tool, and T1-weighted imaging with gadolinium (Gd) enhancement has been considered the golden standard for detecting IAC lesions, especially vestibular schwannomas (also known as acoustic neuromas) that are usually slow-growing benign tumors but can severely affect neurological function and become fatal (Allen et al. 1996; Zealley et al. 2000). Three-dimensional constructive interference in steady-state (CISS) MR imaging is a slow version of fully refocused steady-state sequences (Chavhan et al. 2008) and provides a high spatial resolution image with clear contrast between fluid and other structures, and is reported to have high sensitivity for detecting IAC lesions and cerebellopontine angle (CPA) lesions without additional contrast agents (Casselman et al. 1993; Curtin 1997). Therefore, the CISS sequence is frequently used for the diagnosis and follow up of IAC and CPA lesions combined with T1-weighted MR imaging with Gd, and several reports have recently suggested that only CISS imaging may be suitable for the regu-

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lar follow up of vestibular schwannomas (Ozgen et al. 2009; Bayraktaroglu et al. 2011). However, no reports discuss the availability and limitations of the CISS sequence for diagnosing IAC osteomas.

In this case report, we describe an uncommon case of osteoma of the IAC that was misdiagnosed as IAC schwannoma based on the findings of CISS MR imaging instead of T1-weighted MR imaging with Gd. We also review 17 cases of IAC osteomas reported in the past 22 years and discuss their characteristics.

Clinical Report

A 61-year-old female was admitted to the neurology department at a regional center hospital with complaints of tinnitus and aural fullness of the left ear lasting for 2 months. Screening MR imaging detected no pathological lesions in the brain. She was referred to the otolaryngology department at the same hospital, and diagnosed with left sensorineural hearing loss. Thin-slice MR imaging (IAC/CPA protocol) including the CISS sequence was performed (MAGNETOM Avanto 1.5T, Siemens), and CISS imaging detected a small lesion in the right IAC (unaffected side, Fig. 1). Her left ear hearing loss resolved spontaneously after MR imaging, and she was referred to our university hospital for assessment of an IAC tumor of the right ear.

She complained of mild occasional imbalance, vertigo, and tinnitus of right ear on admission. Her past medical history was unremarkable. Otological examination revealed both tympanic membranes were normal. No signs of facial palsy or facial spasm were found. Neuro-otological examinations, including pure tone audiometry (PTA), auditory brainstem response (ABR), and caloric test, were performed. PTA revealed sensorineural hearing loss (60 dB) only at 4,000 Hz in the right ear (Fig. 2). ABR showed decreased threshold in the right ear without latency elongation. The caloric test was normal in both ears. Finally, she was diagnosed as having an IAC schwannoma, and was followed up annually under "wait and scan" management.

One year after the initial presentation, her condition remained stable. Follow-up T1-weighted MR imaging with Gd showed no change in tumor size, and no enhancement of the tumor (Fig. 3), suggesting that the tumor was not any type of schwannoma or meningioma. Imaging by CT of the temporal bone showed a solitary pedunculated hyperdense lesion on the anterior wall of the right IAC orifice, resulting in diagnosis of osteoma of the IAC (Fig. 4A and C). Additional neuro-otological examinations, including the short increment sensitivity index (SISI) test, distortion product otoacoustic emissions (DPOAEs) test, speech audiometry, and automatic audiometry, were performed at 3 years after the initial presentation. The SISI score was 100% at 4,000 Hz and DPOAEs were absent in the right ear, indicative of cochlear hearing loss. The results of speech audiometry and automatic audiometry did not show any apparent laterality of hearing. Her symptoms remained unchanged for 3 years, and she was advised to return for annual follow-up examinations.

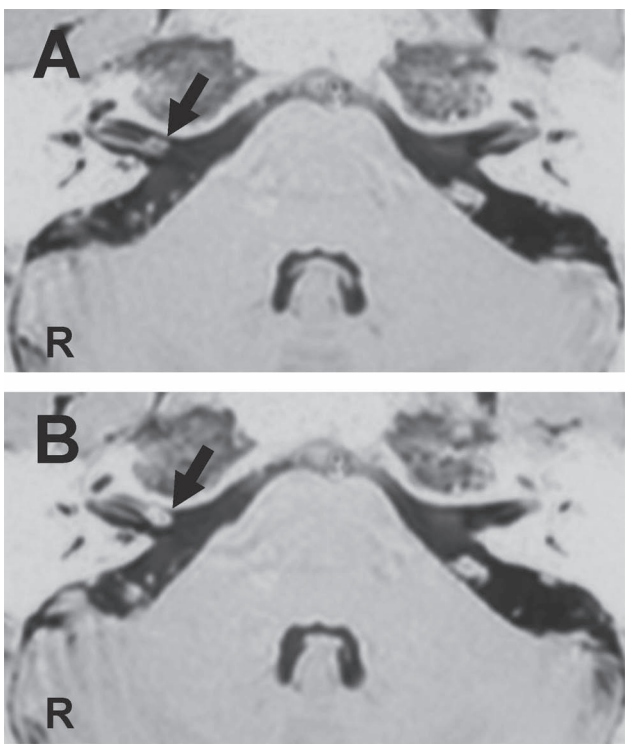

Fig. 1. High-resolution CISS images of the IAC.
(A, B) Ultrathin T2-weighted magnetic resonance images using the constructive interference in steady state (CISS) sequence show a small mass in the right internal auditory canal (IAC) (arrows). R, right.

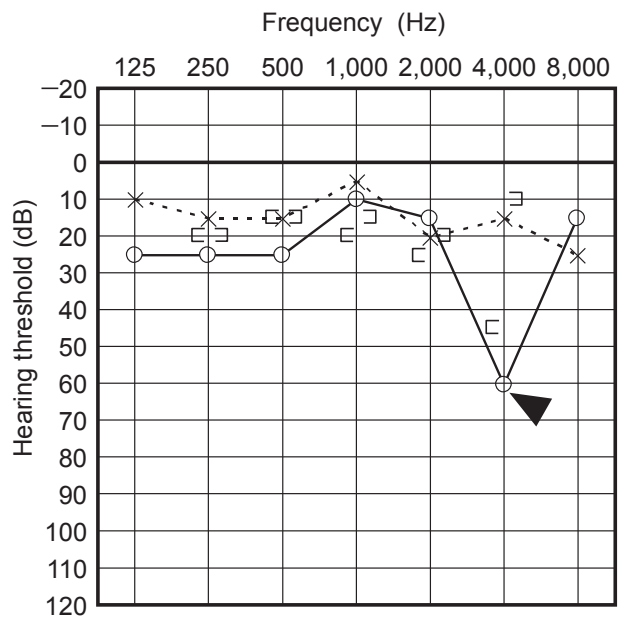

Fig. 2. A pure tone audiogram from the first examination.
An audiogram shows unilateral mild sensorineural hearing loss only at 4,000 Hz in the right ear (arrowhead).

Discussion

Osteomas of the IAC are very rare and usually asymptomatic (Ramsay and Brackmann 1994; Liétin et al. 2010; Baik et al. 2011); thus, the clinical and pathological characteristics are not fully understood. A previous review of 13 cases of IAC osteoma reported from 1882 to 1993 identified several clinical characteristics as follows: mean age of 57 years; female-to-male ratio of 5.5:1, suggesting a particular predilection for postmenopausal women; and 42% of the patients had no symptoms, 42% had hearing loss, 25% had tinnitus, and 33% had vertigo (Clerico et al. 1994). Our present study summarized and reviewed 17 cases of IAC osteoma reported in the past 22 years (1992-2013) (Singh et al. 1992; Ramsay and Brackmann 1994; Coakley et al. 1996; Wright et al. 1996; Boedts et al. 1997; Davis et al. 2000; Vrabec et al. 2000; Kovacić et al. 2001; Gerganov et al. 2008; Liétin et al. 2010; Baik et al. 2011; Kaymakci et al. 2012; Plantone et al. 2013) and excluded the previously reviewed cases by Clerico et al. (1994) (Table 1). Mean age was 44.1 years (male: 44.0, female: 44.2), and female-to-male ratio was 1.4:1. Only 2 of the 17 cases were bilateral. Six cases were identified incidentally (12%), including our case which was asymptomatic in the affected side at detection, 7 cases had subjective or objective hearing loss (41%), 10 cases had tinnitus and/or aural fullness of the affected side (59%), 11 cases had vertigo and/or dizziness (including past medical history) (65%), and 2 cases had facial palsy (including past medical history) (12%). Several causes have been proposed for IAC osteoma, such as trauma, infection, inflammation, hormones, and constant vascular irritation (Clerico et al. 1994; Wright et al. 1996; Baik et al. 2011), but no apparent risk factors were observed in this review. These results suggest that the rate of incidentally diagnosed cases continues to be high, the predilection for postmenopausal women is not so remarkable, and the risk factors for IAC osteoma remain unclear. Further experience with cases of IAC osteoma will be needed to reveal the nature of this rare tumor.

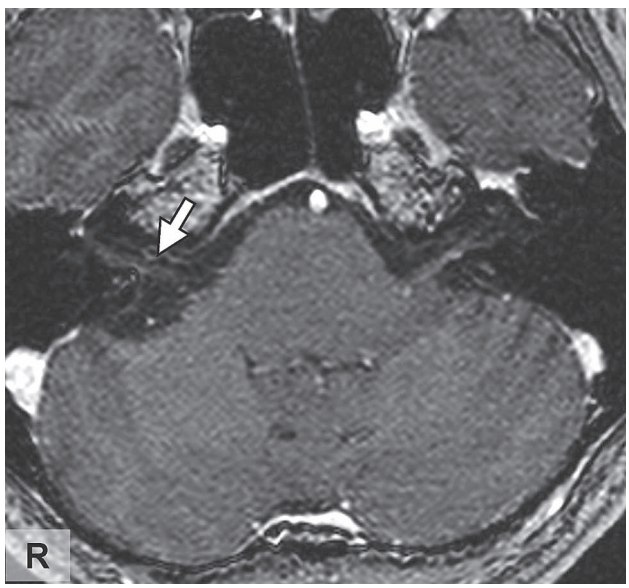

Fig. 3. T1-weighted MRI with gadolinium of the IAC.
A T1-weighted magnetic resonance (MR) image with gadolinium shows no enhancement of the lesion in the right internal auditory canal (IAC) (arrow). R, right.

Bony stenosis of the IAC is easily recognized on CT imaging, and results from various pathologies: bony tumors

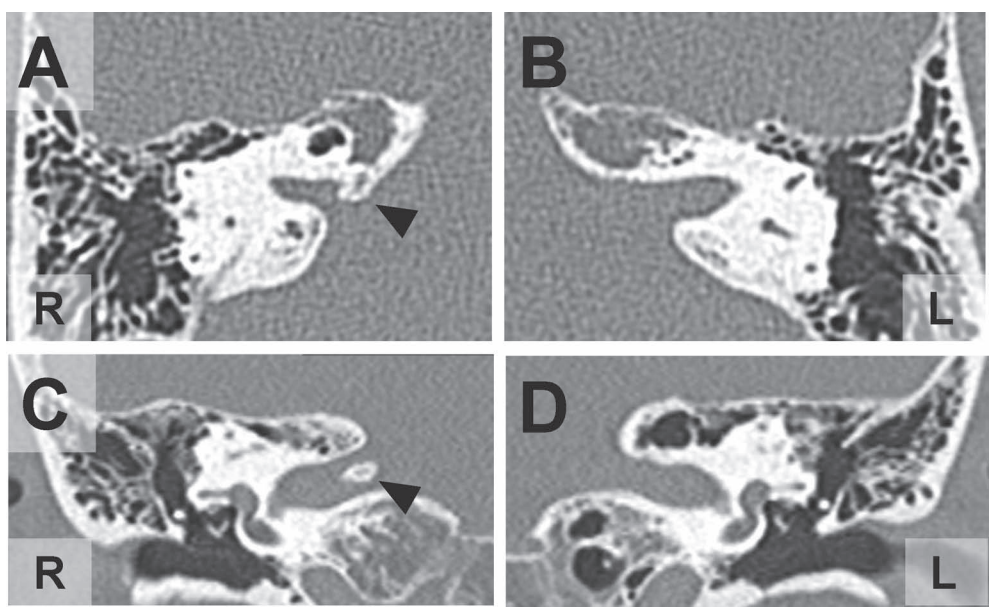

Fig. 4. CT scans of the temporal bone.
Axial (A, B) and coronal (C, D) computed tomography (CT) scans of the temporal bone show a solitary pedunculated bony lesion on the anterior wall at the porus of the right internal auditory canal (arrowheads in A and C). R, right; L, left.

Table 1. Reported cases of osteomas of the internal auditory canal in the past 22 years.

Year	Author	Age	Sex	Side	Incidental discovery	Hearing loss	Tinnitus	Aural fullness	Vertigo/dizziness	Facial palsy	Treatment
1992	Singh V	33	M	R		yes			dizziness		FU
1994	Ramsay HA	30	F	L		yes	yes		vertigo		surgery
1996	Wright A	34	F	R		yes	yes		dizziness		surgery
1996	Coakley DJ	42	M	R			yes		vertigo		surgery
1997	Boedts M	15	M	R	yes	yes (objective)					N/A
2000	Davis TC	44	F	L				yes	dizziness	yes (resolved)	surgery
2000	Davis TC	51	M	R				yes	dizziness		FU
2000	Vrabec JT	31	F	LR	yes						FU
2000	Vrabec JT	61	F	L			yes		vertigo and dizziness		FU
2000	Vrabec JT	59	M	L	yes						FU
2001	Kovacić J	38	M	L			yes		vertigo and dizziness		FU
2008	Gerganov VM	30	F	LR			yes		vertigo		surgery
2010	Liétin B	79	F	L	yes	yes				yes	FU
2011	Baik FM	70	M	R		yes (objective)	yes	yes			FU
2012	Kaymakci M	51	F	R	yes						FU
2013	Plantone D	21	F	L					dizziness		N/A
2014	Suzuki J	61	F	R	yes	yes	yes		dizziness		FU

M, male; F, female; R, right; L, left; FU, follow-up; N/A, data not available.

Table 2. Surgical cases of osteomas of the internal auditory canal.

Year	Author	Age	Sex	Side	Approach	Histology	Resolution of symptoms		
							SNHL	Tinnitus or aural fullness	Vertigo/dizziness
1994	Ramsay HA	30	F	L	middle fossa	N/A	improved	resolved	resolved
1996	Wright A	34	F	R	retrosigmoid	osteoma	improved	N/A	resolved
1996	Coakley DJ	42	M	R	retrosigmoid	osteoma		resolved	resolved
2000	Davis TC	44	F	L	retrosigmoid	osteoma		aural fullness: improved	improved
2008	Gerganov VM	30	F	L	retrosigmoid	osteoma	improved	resolved	resolved

M, male; F, female; R, right; L, left; N/A, data not available; SNHL, sensorineural hearing loss.

within the IAC, such as osteomas and exostoses; generalized enlargements of the temporal bone caused by Paget's disease, fibrous dysplasia, and osteopetrosis; and congenital abnormalities (Clerico et al. 1994; Wright et al. 1996; Baik et al. 2011). IAC osteomas and exostoses are usually slow-growing tumors, and the differential diagnosis is sometimes difficult to establish. These tumors are characterized by several pathological findings that are useful for the differential diagnosis as follows. Osteomas are usually solitary and grossly pedunculated tumors, and microscopic examination reveals fibrovascular channels surrounded by lamellar bone with osteocytes (Graham 1979; Baik et al. 2011). In contrast, exostoses are grossly smooth-bordered and broad-based tumors, and microscopic examination reveals parallel concentric layers of subperiosteal bone, abundant osteocytes, and no fibrovascular channels (Graham 1979; Baik et al. 2011). In addition, exostoses are often multiple and bilateral (Baik et al. 2011). The histological diagnosis of IAC lesions is very difficult to establish based on biopsy sampling, so neuroimaging diagnosis is a practical method. Osteomas may be differentiated from exostoses based on neuroimaging evidence of the presence of bone marrow (Baik et al. 2011), but MR imaging found no bone marrow in our solitary and pedunculated small tumor. Further improvements in imaging technology are needed for effective neuroimaging diagnosis of small IAC tumors.

The differential diagnoses of IAC osteoma with the exception of bony lesions include vestibular schwannoma, facial nerve schwannoma, meningioma, hemangioma, lipoma, and cholesteatoma (Vrabec et al. 2000). With the widespread introduction of imaging devices, the incidental diagnosis of smaller and asymptomatic vestibular schwannomas has increased (Lin et al. 2005). We experienced a case of IAC osteoma misdiagnosed as IAC schwannoma

because the diagnosis was based only on CISS imaging without additional T1-weighted imaging with Gd or CT. The use of MR imaging to diagnose IAC lesions is known to carry the risk of misdiagnosis of IAC bony lesions because of poor visualization of the tumor (Estrem et al. 1993; Davis et al. 2000), but the CISS sequence has not been reported to present potential pitfalls in the diagnosis of IAC lesions without CT. Our experience with the present case indicates several important points as follows: T1-weighted MR imaging with Gd is still essential in some cases to establish the correct diagnosis of IAC lesions; and the CISS sequence is useful for the detection of small IAC lesions in patients for whom contrast material is contraindicated, but consecutive CT should be performed to rule out bony tumors if an IAC lesion is detected.

The treatment of IAC osteomas remains controversial. Periodic follow-up examination was selected in 10 of the 17 cases in this review of IAC osteomas. Surgery was performed in 5 cases (middle fossa approach in one case, retrosigmoid approach in 4 cases) (Table 2). Review of the surgical outcomes of eight IAC osteoma patients, including 4 patients in our study, found that all patients obtained some improvement, but all symptoms were resolved in only 3 patients (Davis et al. 2000). In this study, all patients treated by surgery reported resolved or improved symptoms after surgery (Table 2). The treatment of IAC osteoma is decided according to the severity of the symptoms caused by nerve compression (Baik et al. 2011; Kaymakci et al. 2012), and patients with severe symptoms and/or risk of complete canal occlusion may require surgical excision of the tumor before irreversible damage occurs (Baik et al. 2011). However, it is sometimes difficult to decide whether cochlear lesion or retro-cochlear lesion has caused the symptoms of a patient with IAC osteoma. Our case showed abnormalities in the SISI score and DPOAEs that suggest cochlear hearing loss. Accurate hearing and vestibular tests were not always performed or not described in previous case reports. Therefore, further investigations of detailed hearing and vestibular functions in patients with IAC osteoma are needed to establish the appropriate surgical indications.

In conclusion, IAC osteoma is an uncommon disease. CISS MR imaging is useful for the detection of small IAC lesions, but has a limitation in the specific diagnosis of IAC lesion, because little information is provided in addition to the structure. Therefore, consecutive CT should be performed if IAC lesions are detected by CISS imaging in a patient contraindicated for contrast medium.

Conflict of Interest

The authors declare no conflict of interest.

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