Exostoses and Osteomas of the Internal Auditory Canal

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ABSTRACT

Objectives: 1) Describe time course of symptom resolution after surgical management of IAC exostoses; 2) Define radiologic and histologic criteria to differentiate exostoses from osteomas of the IAC.

Study Design: Case series and literature review.

Setting: Ambulatory care center and outpatient surgical center

Patients: Three patients with internal auditory canal bony growths were studied.

Intervention: One patient underwent surgery to remove exostosis on the right, and two patients were managed non-surgically.

Results: Patient 1 presented with disabling vertigo and was found to have bilateral exostoses with nerve impingement on the right. Following right-sided exostoses removal via retrosigmoid craniotomy, the patient had complete resolution of her symptoms. However, this occurred over a protracted time course of one year. Patient 2 presented with bilateral pulsatile tinnitus and occasional vertigo and was found to have bilateral IAC exostoses. Patient 3 had unilateral IAC osteoma and presented with hearing loss and tinnitus. Due to the mild nature of their symptoms, these patients were managed non-surgically. We also describe radiologic and histologic criteria to differentiate exostoses from osteomas of the IAC.

Conclusion: In our patient with severe vertigo due to IAC exostoses, surgical removal resulted in complete symptom resolution, although over a protracted time course. We note that IAC osteomas can be differentiated from exostoses by the presence of bone marrow (radiographically) or the presence of fibrovascular channels (histologically).

INTRODUCTION

Although common in the external auditory canal, internal auditory canal (IAC) exostoses and osteomas are rare clinical entities that are often confused with one another and grouped together. In the medical literature, there have been only four cases of IAC exostoses, 15 reported cases of unilateral osteomas and two bilateral cases of IAC osteomas (2-11). Here we report two cases of bilateral exostoses and one case of an osteoma, and discuss the clinical presentation, histopathology, imaging, and management of these rare lesions.

CASE 1

A 65-year old Caucasian female presented to our clinic with an 18-year history of chronic dyssequilibrium which progressed significantly in the past year. Otological examination revealed normal tympanic membranes bilaterally. There was no facial weakness or visual or gait disturbance. The neurological exam was benign. Pure tone audiometry revealed symmetrical mild sensorineural hearing loss with 76% speech discrimination bilaterally (Figure 1). Electronystagmography showed no abnormalities on central or peripheral testing. Temporal bone computed tomography (CT) revealed bilateral smoothly-bordered, broad-based bony growths of the anterior and posterior internal openings of the IAC (Figure 2). The diagnosis of bilateral IAC exostoses was made. There was no evidence of superior semicircular canal dehiscence. No generalized bony disease or syringomyelia were detected. T2-weighted magnetic resonance imaging (MRI) showed right-sided exostoses to be more severe than left, with resulting compression of the nerve bundle (Figure 3). The patient opted to proceed with surgery to remove the more severe right-sided exostoses.

The patient underwent a retrosigmoid craniotomy. Exploration of the right IAC showed significant exostoses posteriorly and superiorly with compression of the VII and VIII nerve complex. With use of diamond burrs, the exostoses were removed, revealing "onion-skinned" laminated bone. The nerve bundle was decompressed. Bone chips were sent for histopathologic exam and showed benign laminated bone consistent with the previous diagnosis (Figure 4).

Figure 1. Audiogram showing bilateral mild sensorineural hearing loss. Mild conductive hearing loss is due to ear wax.

Figure 2. Axial CT temporal bone scan showing multiple, broad-based bony growths consistent with exostoses (arrows) at the porus of the IAC (a. right, b. left).

Figure 3. T2 FSF MRI showing impingement of the right nerve bundle (arrows) due to compression by the exostoses.

At time of discharge, the patient experienced no immediate relief of her presenting symptoms. In the months following surgery, she experienced slow and steady improvement. At 3 months, she remained 100% cane-dependent and at 6 months improved to 75% cane-dependence. One year after surgery, the patient reported that she was cane-free and was no longer suffering from dyssequilibrium. Furthermore, she was able to resume balance-intensive physical activities such as tai chi. Thereafter, the patient was advised to follow-up every 6 months. She has remained asymptomatic on the right side and has never been symptomatic from the left-sided IAC exostoses.

Figure 4. Hematoxylin and eosin staining of temporal bone chip from patient 1 showing onion skinned laminated bone (arrows) consistent with the diagnosis of exostoses.

CASE 2

A 55-year old female presented with an 8-month history of slowly progressive bilateral pulsatile tinnitus and occasional episodes of imbalance and vertigo, lasting only seconds, associated with head turning in either direction. She denied any significant otologic history. The patient had a normal head and neck exam. Pure-tone audiometry displayed a mild symmetric sensorineural hearing loss at 2000 Hz. The patient had a speech reception threshold of 10dB on the right and 15dB on the left with a speech discrimination score of 100% bilaterally. The Dix-Hallpike maneuver was negative bilaterally. CT scans displayed exostoses bilaterally at the porus of the IAC (Figure 5).

Figure 5. Axial CT scan (a. right, b. left) showing broad-based nature of bony growths. In this case, there is the presence of bone marrow, but this is continuous with the bone marrow of the petrous apex, and not the bony growth.

A year later, the patient's tinnitus had become louder and he also experienced some "jolt-like" noises in the right ear. However, repeat imaging showed no changes in the size of the IAC lesion. The head and neck exam was normal. Tympanic membranes were intact bilaterally. There was no facial asymmetry. Pure tone audiometry showed severe to profound sensorineural hearing loss with very poor speech discrimination on the right. On the left, there was severe sensorineural hearing loss from 1000 Hz and greater with excellent speech discrimination (Figure 7). Temporal bone imaging showed a right-sided osteoma originating from the porus, causing narrowing of the IAC to 2 mm.

Two years after his initial presentation, the patient's condition has remained stable. Although he has some complaints of increased right-sided tinnitus, he continues to display no signs of facial nerve compression. Audiometry is unchanged. Follow-up temporal bone CT has shown no changes in the size of the right-sided osteoma. The patient was advised to follow-up annually. In the case that he develops symptoms secondary to facial nerve compression, surgery to remove the osteoma will be considered.

CASE 3

A 70-year old Caucasian male presented to a colleague with a chief complaint of a “plugged” right ear and right-sided tinnitus. He was diagnosed with right-sided labyrinthitis and treated with steroids. A year later, he followed up in our clinic with continued tinnitus and hearing loss. MRI and CT scans showed an osteoma of the right IAC (Figure 6).

Figure 6. Axial CT scan through the temporal bone displaying a bony plug in the right IAC (arrow). Coronal CT scan showing bone growth in the right IAC (arrow). 3D CT reconstruction.

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In the case of Patient 1, it is likely she had exostoses for several years and only experienced significant vertigo in the setting progressive impingement on the nerve bundle, advanced age, and inability to further compensate for vestibular nerve compression. Patient 3 also presented with symptoms secondary to eight nerve compression, manifested as hearing loss and tinnitus. Patient 2 is unusual in that her only presenting symptom was tinnitus.

In practice, most cases of IAC exostoses and osteomas are not surgically treated and are not readily accessible for tissue biopsy. In the absence of a tissue diagnosis, radiographic appearance is fundamental in differentiating between various bony lesions of the IAC. The radiographic appearance of an exostosis is that of a smoothly bordered, broad-based bony growth protruding into the IAC. Exostoses are often multiple and bilateral. Osteomas, on the other hand, are usually solitary, pedunculated lesions extending in the IAC space, with the remainder of the canal having no resection. Furthermore, we propose that osteomas can be differentiated from exostoses by radiographic evidence of bone marrow which would be equivalent to the histopathologic finding of fibrovascular channels.

Treatment of IAC exostoses and osteomas depends on the severity of symptoms occurring due to nerve compression. Observation alone is recommended for asymptomatic individuals. In the cases of Patients 2 and 3, repeat imaging of the IAC bony growths has shown any size progression, and their presenting symptoms have remained stable. Surgical excision of the lesion to decompress the nerve complex before irreversible damage is the treatment of choice for severely symptomatic patients, such as those with progressive hearing loss or vertigo, or those at risk of complete canal occlusion. In the first case we presented, surgical removal of the exostoses was eventually curative of the patient’s presenting symptoms; however, it occurred gradually over a period of one year. The protracted time course required for her recovery likely reflects chronic compression of the neurovascular bundle.

REFERENCES