Prevalence of Internal Auditory Canal Diverticulum and Its Association with Hearing Loss and Otosclerosis

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ABSTRACT

BACKGROUND AND PURPOSE: Focal low-attenuation outpouching or diverticulum at the anterolateral internal auditory canal is an uncommon finding on CT of the temporal bone. This finding has been described as cavitary otosclerosis in small case reports and histology series. The purpose of this study was to establish the prevalence of internal auditory canal diverticulum and its association with classic imaging findings of otosclerosis and/or hearing loss.

MATERIALS AND METHODS: Temporal bone CT scans of 807 patients, obtained between January 2013 and January 2016, were retrospectively reviewed to identify internal auditory canal diverticula and/or classic imaging findings of otosclerosis. Clinical evaluations for hearing loss were reviewed for patients with internal auditory canal diverticula and/or otosclerosis.

RESULTS: Internal auditory canal diverticula were found in 43 patients (5%); classic otosclerosis, in 39 patients (5%); and both findings, in 7 patients (1%). Most temporal bones with only findings of internal auditory canal diverticula (91%) demonstrated hearing loss, with 63% of this group demonstrating sensorineural hearing loss. The hearing loss classification distribution was significantly different ($P<.01$) from that in the classic otosclerosis group and in the group with both diverticula and otosclerosis.

CONCLUSIONS: Internal auditory canal diverticula are not uncommon on CT examinations of the temporal bone and most commonly occur without classic imaging findings of otosclerosis. These lesions are associated with sensorineural hearing loss, and referral for hearing evaluation may be appropriate when present.

ABBREVIATIONS: CHL = conductive hearing loss; IAC = internal auditory canal; SNHL = sensorineural hearing loss

A focal low-attenuation notch or diverticulum within the temporal bone continuous with the internal auditory canal (IAC) is an unusual finding on imaging studies of the temporal bone. Several histologic and imaging case reports refer to this finding as a form of cavitary otosclerosis and even suggest that the presence is associated with advanced disease.

Otosclerosis is an osteodystrophic disorder of the otic capsule, resulting in abnormal resorption of endochondral bone and deposition of abnormal vascular bone. Otosclerosis usually appears in the third-to-fifth decades of life, and most commonly affects women. Clinical otosclerosis is present in <1% of the population, though it has been reported in up to 11% of the population on histology performed at postmortem examination. On CT of the temporal bone, otosclerosis commonly appears as lucent or hypodense bone surrounding the otic capsule, often limited to the region anterior to the oval window. This process results in either conductive hearing loss (CHL) due to fixation of the stapes footplate or mixed conductive and sensorineural hearing loss (SNHL) due concomitant otic capsule involvement. Otosclerosis presenting with only SNHL in the absence of CHL is rare and is often called “cochlear otosclerosis.”

Establishing the significance of the IAC diverticulum or notch is important for both the radiologist and referring physician in guiding clinical management. Determining the relationship of this lesion to classic imaging findings of otosclerosis could also be helpful in the understanding of otosclerosis and the spectrum of clinical presentations. Therefore, the purpose of this study was the following: 1) to determine the prevalence of IAC diverticula at our institution, and 2) to explore potential associations with otosclerosis and hearing loss in patients identified with an IAC diverticulum.
The prevalence of findings in the study population and associations between hearing groups were evaluated with Wilcoxon rank sum, Kruskal-Wallis, Fisher exact, and χ² analyses performed with SAS software, Version 9.4 (SAS Institute, Cary, North Carolina). A P value of < .05 was used to determine statistical significance.

**RESULTS**

A total of 810 eligible temporal bone CT examinations were identified during the 3-year study period. Three studies were excluded because of nondiagnostic images due to motion or artifacts. Patient demographics are described in Table 1. Of the 807 patients included in the study, IAC diverticulum was identified in 5% (n = 43) of examinations. The appearance ranged from a small notch to full outpouching, with all diverticula located along the anterior IAC medial to the cochlea (Fig 1). Sixty percent of patients (n = 26) demonstrated bilateral diverticula. Classic imaging findings of otosclerosis were also present in 5% (n = 39) of patients. Sixty-nine percent of patients with classic imaging findings of otosclerosis (n = 27) demonstrated bilateral findings. One percent (n = 7) of all eligible patients demonstrated imaging findings of both otosclerosis and IAC diverticula with 71% (n = 5) of this group demonstrating bilateral otosclerosis and IAC diverticula (Fig 2). Demographics of patients with IAC diverticula, both with and without otosclerosis, are described in Table 2.

Seventy-five of 807 patients (n = 123 temporal bones) demonstrated findings of IAC diverticula, otosclerosis, or both and were further evaluated for hearing loss. These individual temporal bones were divided into 3 groups: IAC diverticulum only, otosclerosis only, and both IAC diverticulum and otosclerosis (Table 3). In the IAC diverticulum-only group, there were 36 patients with 57 affected temporal bones. Audiogram results were unavailable or excluded due to a prior operation for 14 temporal bones in this group. Of the 43 temporal bones with only an IAC diverticulum and hearing evaluation, 9% (n = 4) demonstrated no hearing loss, 63% (n = 27) demonstrated SNHL, and 28% (n = 12) demonstrated a mixed hearing loss. No temporal bones in this group demonstrated CHL.

In the classic otosclerosis-only group, there were 33 patients with 54 affected temporal bones. Most (81%, n = 44) temporal bones had fenestral otosclerosis only, while 4% (n = 2) had co-
chlear otosclerosis only and 15% (n = 8) had both fenestral and cochlear otosclerosis. Audiogram results were unavailable or excluded for 15 temporal bones in this group. A total of 18% (n = 7) demonstrated no hearing loss, 26% (n = 10) demonstrated SNHL, 23% (n = 9) demonstrated a mixed hearing loss, and 33% (n = 13) demonstrated CHL.

In the IAC diverticulum and otosclerosis group, there were 7 patients with 12 affected temporal bones. Fenestral otosclerosis was present in all 12 temporal bones, with 3 temporal bones demonstrating concomitant cochlear otosclerosis. Audiogram results were unavailable or excluded for 5 temporal bones in this group. A total of 14% (n = 1) demonstrated no hearing loss, 29% (n = 2) demonstrated SNHL, 43% (n = 3) demonstrated a mixed hearing loss, and 14% (n = 1) demonstrated CHL.

Comparison of the 3 groups demonstrated a statistically significant difference in hearing loss patterns between each group. These findings suggest that SNHL is more commonly associated with IAC diverticulum than with traditional imaging findings of otosclerosis.

DISCUSSION
The prevalence and clinical significance of IAC diverticula are not well-established in the current literature. In this large retrospective case review, IAC diverticula were present in 5% of our study population and more frequent (n = 43) than classic findings of otosclerosis (n = 39). The appearance of diverticula varied from a small notch to a larger outpouching and was always located at the anterior IAC near the medial margin of the dense otic capsule. IAC diverticula were more commonly an isolated finding and rarely occurred with concurrent classic findings of otosclerosis. Compared with patients with isolated otosclerosis (median age, 52 years), patients with IAC diverticula were significantly older at median ages of 62 years with additional findings of otosclerosis and 61 years without additional findings of otosclerosis (P < .01).

Most (91%) patients with an isolated IAC diverticulum without additional imaging evidence of otosclerosis had hearing loss with more than half (63%) demonstrating pure SNHL. The pattern of hearing loss in isolated IAC diverticula was also found to be significantly different from hearing loss patterns in the classic imaging appearance of otosclerosis with or without an additional finding of an IAC diverticulum. These findings suggest that IAC diverticula are a clinically important finding on temporal bone CT and have important hearing loss implications.

Audiometric data demonstrated more advanced hearing loss in patients with isolated IAC diverticulum than previously reported with presbycusis or age-related hearing loss. Presbycusis or age-related hearing loss is defined as SNHL caused by the aging process without a confounding etiology, such as metabolic disorder, trauma, or noise injury.10 Presbycusis may play a partial role in SNHL in the diverticula group, given that the median age of the diverticula-only group was 61 years. However, the average pure tone average was 53.5 dB in the 27 temporal bones with isolated IAC diverticulum, which is consistent with moderate-to-severe hearing loss and was greater than expected with presbycusis alone.10,11 Moreover, the average pure tone average in the 27 isolated diverticula with SNHL was like that in the 10 temporal bones
IAC diverticula are described in small case reports as rare entities reflective of cavitary otosclerosis.1-4 Beyond the hearing loss implications of cavitary otosclerosis, Makarem et al5 also described the clinical significance of the lesion for potential cochlear implant malpositioning into the diverticulum. The consistent location of all diverticula in this series along the anterior IAC matches that in prior temporal bone histology series descriptions of cavitary otosclerosis.1,2,11 The occurrence of outpouching or notches was like classic CT findings of otosclerosis in this study, both groups representing 5% of all patients. Therefore, the prevalence of IAC diverticula is likely not as rare as previously implied. Moreover, the findings in this series do not support previous reporting of an IAC diverticulum as a manifestation of severe otosclerosis due to its high frequency as an isolated finding and the occurrence of the lesion in patients without hearing loss. Previous case reports do not directly address the significance of IAC diverticula as an isolated finding without additional classic CT findings of otosclerosis. This study suggests that isolated IAC diverticula are related to hearing loss in a different pattern than the classic CT presentation of otosclerosis.

Classic otosclerosis invariably involves the anterior stapes footplate near the fissula ante fenestram and presents with a component of CHL.5,14 SNHL in otosclerosis is less well-understood and may be due to production of local inflammatory cytokines leading to atrophy and concomitant dysfunction of the spiral ligament.15,16 SNHL is usually present with CHL and manifests as a mixed hearing loss classification. Temporal bones in this study that had classic imaging findings of otosclerosis more commonly presented with CHL, and temporal bones with both classic imaging findings of otosclerosis plus diverticula more commonly presented with mixed hearing loss as expected. No hearing loss was identified in 7 temporal bones with findings of otosclerosis (18% of temporal bones with otosclerosis). These temporal bones had imaging indications for otosclerosis or hearing loss with a unilateral clinical presentation of hearing loss and bilateral imaging findings. Otosclerosis presenting with isolated SNHL without a conductive component is believed to be rare and not well-understood.5,17-19 IAC diverticula may represent an imaging manifestation of otosclerosis with an isolated SNHL, because most patients with diverticula in this study presented with isolated SNHL (63%). A smaller portion of isolated IAC diverticula demonstrated a mixed pattern of hearing loss (28%). These IAC diverticula may result in not only SNHL due to spiral ligament dysfunction but also CHL due to abutment to the scala vestibuli, resulting in a third-window phenomenon.2,4,16,20 Another potential explanation of mixed hearing loss with CT findings of isolated diverticula may be otosclerosis involvement of the footplate below the resolution of routine CT temporal bone examinations.

There were several limitations to this study. Its retrospective nature limited the information available for review. Namely, clinical data were not available for all patients with IAC diverticula or classic otosclerosis. Small sample sizes, especially in the combined IAC diverticulum and otosclerosis group, limited statistical analysis for comparing grouped populations of CT findings. This study also had a component of selection bias that may have overestimated the prevalence of findings on CT due to referral patterns. MR images of the temporal bone were also not reviewed in this study, despite MR imaging being a frequently ordered examination to evaluate SNHL. With most patients with IAC diverticula demonstrating SNHL, the actual prevalence of IAC diverticula may have been higher on high-resolution MR imaging compared with CT.

The relationship of the anterior internal auditory canal wall to the development of a diverticulum or focal otosclerosis is unclear. The IAC diverticulum may be occasionally present in classic otosclerosis early in the disease process when other changes are too subtle to be detected on imaging. An additional consideration is that the IAC diverticulum represents a variant of cochlear otosclerosis with the nidus for resorption of endochondral bone near the junction of endochondral bone of the medial otic capsule and lamellated bone of the IAC, as opposed to the more common

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Table 2: Demographic characteristics by the presence of IAC or otosclerosis

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>IAC Diverticulum Only (n = 36)</th>
<th>Otosclerosis Only (n = 32)</th>
<th>IAC Diverticulum + Otosclerosis (n = 7)</th>
<th>None (n = 732)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median age (range) (yr)</td>
<td>62 (18–91)</td>
<td>52 (18–91)</td>
<td>61 (30–70)</td>
<td>52 (18–96)</td>
<td>&lt;.01a</td>
</tr>
<tr>
<td>Sex (No. [%])</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>.47b</td>
</tr>
<tr>
<td>Male</td>
<td>15 (41.7)</td>
<td>9 (28.1)</td>
<td>3 (42.9)</td>
<td>316 (43.2)</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>21 (58.3)</td>
<td>23 (71.9)</td>
<td>4 (57.1)</td>
<td>416 (56.8)</td>
<td>.02b</td>
</tr>
<tr>
<td>Temporal bone involvement (No. [%])</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unilateral</td>
<td>9 (20.9)</td>
<td>15 (48.4)</td>
<td>5 (71.4)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bilateral</td>
<td>34 (79.1)</td>
<td>24 (51.6)</td>
<td>2 (28.6)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*P value was calculated with the Kruskal-Wallis test.

Table 3: Associations between IAC diverticulum or otosclerosis and hearing loss

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>IAC Diverticulum Only (n = 43)</th>
<th>Otosclerosis Only (n = 39)</th>
<th>IAC Diverticulum + Otosclerosis (n = 7)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing loss (No. [%])</td>
<td></td>
<td></td>
<td></td>
<td>&lt;.01a</td>
</tr>
<tr>
<td>None</td>
<td>4 (9%)</td>
<td>7 (18%)</td>
<td>1 (14%)</td>
<td></td>
</tr>
<tr>
<td>SNHL</td>
<td>27 (63%)</td>
<td>10 (26%)</td>
<td>2 (29%)</td>
<td></td>
</tr>
<tr>
<td>PTA (average) (range) (dB)</td>
<td>53.5 (10.6–120)</td>
<td>55.3 (6.9–115)</td>
<td>97.5 (94.4–100.6)</td>
<td></td>
</tr>
<tr>
<td>CHL (No. [%])</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>13 (33%)</td>
<td>1 (14%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mixed (No. [%])</td>
<td>12 (28%)</td>
<td>9 (23%)</td>
<td>3 (43%)</td>
<td></td>
</tr>
</tbody>
</table>

Note: —PTA indicates pure tone average.

*P value was calculated for comparison of groups with the Fisher exact test.
location of the fissula ante fenestram. In this situation, the diverticula could potentially represent advanced spongiotic bone immediately adjacent to IAC CSF. Hounsfield unit values were not measured to evaluate quantitative density due to the small size of several of the diverticula, limiting accuracy. However, this lesion qualitatively appeared as a low, near-CSF density notch or outpouching in the bone cortex. IAC diverticula are also unlikely to be found in disease processes such as osteoporosis, Paget disease, or osteogenesis imperfecta given their frequency as a small isolated finding. It is also unlikely that an IAC diverticulum reflects a normal anatomic variant, given its association with hearing loss greater than expected for age. While this study cannot determine the etiology of the diverticulum, this lesion does demonstrate a relationship with hearing loss and, most commonly, SNHL.

The results of this study illustrate the clinical importance of identification of IAC diverticula due their association with hearing loss, and radiologists should be aware of this entity. If identified on imaging, otolaryngology consultation and audiometric evaluation should be considered if not previously performed. IAC diverticula can be identified not uncommonly on temporal bone CT and should be mentioned in the radiology report, given their association with hearing loss.

CONCLUSIONS
IAC diverticula are focal CSF attenuation outpouchings at the anterolateral internal auditory canal, present on 5% of CT examinations of the temporal bone in our study population. IAC diverticula are most commonly isolated findings and are associated with a different pattern of hearing loss compared with classic findings of otosclerosis. Identification of this clinically significant finding on CT may warrant audiometric evaluation, given the association with hearing loss, specifically SNHL.

ACKNOWLEDGMENTS
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