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Histopathologic characteristics of internal auditory canal diverticula

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Abstract

Hypothesis: We hypothesize that internal auditory canal (IAC) diverticula occur independent of otosclerosis as demonstrated by temporal bone histopathology.

Background: Diverticula at the anterior-inferior aspect of the internal auditory canal have been described histologically in the setting of cavitary otosclerosis. Recent radiographic studies show the prevalence of IAC diverticula that is higher than what can be accounted for by cavitary otosclerosis alone.

Methods: We examined hematoxylin and eosin temporal bone histopathology slides with otosclerosis involving the internal auditory canal. We also examined bones from normal hearing subjects with normal histologic findings. Temporal bones were included if donors were >18 years of age at time of death and adequate horizontal cuts were available to evaluate the area of interest.

Results: IAC diverticula were found in 33 of 47 (70%) temporal bones with IAC otosclerosis and in 5 of 20 (25%) normal temporal bones. The difference in mean pure tone averages (PTA) in the normal temporal bones with (PTA 7.3 +/− 7) and without (PTA 8 +/− 2) diverticula was not statistically significant (p = 0.86)

Conclusion: Internal auditory canal diverticula which have been previously demonstrated to occur in the setting of cavitary otosclerosis can also occur independent from otosclerosis. Subjects with diverticula but without other temporal bone pathology have normal hearing thresholds.

Introduction

Internal auditory canal (IAC) diverticula are focal outpouchings from the normal linear contour of the anterior-inferior aspect of the IAC. Historically, IAC diverticula have been described in the setting of otosclerosis. IAC diverticula have been described

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histopathologically in large series of otosclerotic human temporal bones[1,2] and in case reports in the setting of cavitary otosclerosis.[3-5] Radiographic case reports of IAC diverticula in the setting of otosclerosis have also been published.[6,7] In 2012, Hoeberigs et al. found a radiographic relationship between otosclerosis severity and IAC diverticula in a series of 92 patients.[8] IAC diverticula are also the most common site of cavitary otosclerosis identified on CT imaging.[9]

In 2017, Pippin discovered a 5% prevalence of IAC diverticula in a cohort of patients who underwent high resolution CT (HRCT) of the temporal bone. In this series, 84% of temporal bones with IAC diverticula lacked radiographic evidence of fenestral or retrofenestral otosclerosis.[10] The size of IAC diverticula does not statistically correlate with degree of hearing loss, and diverticula have been shown to have a 5% prevalence in a control population who underwent HRCT for sinus pathology (Hinrich Staecker MD, PhD, personal communication).

These radiographic studies suggest that a preponderance of IAC diverticula occur independent of otosclerotic foci. Moreover, the prevalence of IAC diverticula is higher than what can be accounted for by cavitary otosclerosis alone. We hypothesize that IAC diverticula occur independent of otosclerosis as demonstrated by temporal bone histopathology.

Materials and Methods

The Institutional Review Board (IRB) of UCLA approved this study (IRB protocol #10-001449). All methods used in this study were in accordance with NIH and IRB guidelines and regulations. Appropriate informed consent was obtained from each patient before inclusion in the collection. The National Institute on Deafness and Other Communication Disorders grant 1U24DC015910-01 (AI) provided funding this study.

Human temporal bone harvest and embedding in celloidin were performed as previously described.[11] The celloidin block was cut into 20-micron sections of which every tenth was mounted and stained with hematoxylin and eosin (H&E).

The House-UCLA HTB database was used to identify bones from patients who had otosclerotic involvement of the IAC. The database was then queried to identify bones from patients with normal histologic findings and normal hearing.

Temporal bones were included if donors were >18 years of age at time of death and adequate horizontal cuts were available to evaluate the area of interest. Bones were excluded for ipsilateral abnormal histopathologic findings including otosclerosis, vestibular schwannoma, glomus tumor, Scheibe deformity, cochlear hydrops.

Microscopic analysis was performed and special attention was given to the anterior-inferior aspect of the IAC. In the normal subgroup, slides were excluded if pathologic processes were identified histologically.

In accordance with previous studies,[8-10] an IAC diverticulum was defined as a non-vascular outpouching from the normal linear contour of the IAC wall identified on horizontal sections. A subtle wall curvature was not categorized as a diverticulum.

Historical charts were reviewed to collect pure tone average (PTA) and word recognition scores (WRS) for patients with normal temporal bones. We report the 4-frequency pure tone average (PTA) using 0.5, 1, 2, and 3 kHz as recommended in 1979 by the American Medical Association (AMA),[12] which was later validated by Dobie,[13] and remains the preferred method for reporting hearing outcomes.[14] Of those subjects with normal temporal bone histopathology and diverticula, 3/5 had PTA data and 1/5 had WRS data. Of those subjects with normal temporal bone histopathology without diverticula, 12/15 had PTA data and 4/12 had WRS data. Student's t-test was used to compare the PTA scores. Due to insufficient data, WRS were unable to be compared.

Results

Twenty-six adult subjects with IAC otosclerosis were identified with 47 temporal bones meeting inclusion criteria. In this group, 33 IAC diverticula were identified, of which 24/33 existed in bilateral temporal bones of the same subjects. Five were unilateral without a contralateral diverticulum. Four were unilateral with contralateral side unavailable for review. Mean age at death of patient with an IAC diverticulum in the setting of otosclerosis was 69 years. Twenty diverticula were found on the right and 13 on the left.

Thirty adult subjects with normal temporal bones were initially identified, with 20 temporal bones meeting inclusion criteria. Five IAC diverticula were identified in this group. Contralateral histopathologic findings are listed in Table 1. The mean age at death of patients with an IAC diverticulum with otherwise normal temporal bones was 70 years. Two patients' causes of death were unknown, one died of intracerebral hemorrhage following contralateral translabyrinthine resection of a vestibular schwannoma, and one died of mucormucyosis that did not affect either temporal bone as confirmed by histopathological analysis. Two diverticula were found on the right and 3 on the left.

Normal temporal bones with diverticula were associated with a mean PTA of 8 +/− 2 decibels (dB) and mean WRS 96%. For this group, mean time between last audiogram and death was 12 years. Normal temporal bones without diverticula were associated with a mean PTA of 7.25 +/− 7 dB and a mean WRS of 98% +/− 4%. For this group, mean time between last audiogram and death was 3.3 years. The mean PTAs of normal temporal bones with and without diverticula were not significantly different ($p = 0.86$).

On histopathologic review, several examples of otosclerotic foci at the typical site for an IAC diverticulum without a diverticulum were found. Figure 1.

Discussion

We identified IAC diverticula in 5/20 (25%) of normal temporal bone specimens, donated by subjects with normal ipsilateral hearing. This finding validates our hypothesis that IAC diverticula can occur independent of otosclerosis. In addition, we discovered that when otosclerosis involves the IAC, there is a concomitant IAC diverticulum in 70% of cases in this series.

Contemporary studies estimate the histologic prevalence of otosclerosis to be near 2.5%.[15] In large otosclerosis temporal bone series, IAC diverticula prevalence has been found to range from 4.9%[1] to 18.75%.[2] These rates can be extrapolated to estimate the prevalence of IAC diverticula in the setting of otosclerosis to be 0.12 – 0.47%. These data lead us to propose that most IAC diverticula occur in the setting of a normal temporal bone. Pippin et al showed a radiographic prevalence of IAC diverticula to be 5%, with only 0.8% prevalence of concurrent otosclerosis and IAC diverticula.[10] Mihal et al recently found a 4.7% prevalence of IAC diverticula. This study also compared unilateral diverticula to their contralateral control side to show that IAC diverticula are not associated with a worse pure tone average or worse recognition score.[16]

Given the preponderance of IAC diverticula independent of otosclerosis, we suspect that IAC diverticula may be congenital. Bast and Anson described endochondral otic capsule development in detail.[17] By the 8th gestational week, a cartilaginous model of the otic capsule has formed from the mesenchyme. Fourteen ossification centers appear and fuse to complete otic capsule ossification. An endosteal layer, an endochondral layer, and a periosteal layer develop within the otic capsule. IAC diverticula are known to occur at the edge of the otic capsule. Perhaps IAC diverticula represent a small cleft at the fusion between the otic capsule periosteum and the cancellous petrous apex bone. IAC diverticula may be developmentally similar to the fissula ante fenestram, fossula post fenestram, and the hypoattenuated focus in the anterior otic capsule, all of which are thought to be relics of otic capsule development.[18,19] The congenital nature of IAC diverticula should be further explored radiographically and histopathologically.

The primary limitation of our study is the small sample size. We do not feel that the true prevalence of IAC diverticula in normal temporal bones can be inferred from this study. In our laboratory, only 20 normal temporal bones met criteria for review. A sample size of 508 would be required to produce a two-sided 95% confidence interval with a width equal to 4% (+/− 2%) when the sample percentage is 5%. Although we cannot infer prevalence from the current study, we have demonstrated the occurrence of diverticula in histologically normal temporal bones from subjects with normal hearing. A second limitation is that there is a requisite interval between last audiogram and date of death for these patients who donated their temporal bones. The length of this interval will affect the degree to which the last audiogram accurately reflects the patient’s hearing status at death.

In conclusion, IAC diverticula can occur in normal temporal bones independent of otosclerosis. This finding implies that radiographic evidence of an IAC diverticulum in and of itself is not necessarily a harbinger of pathology.

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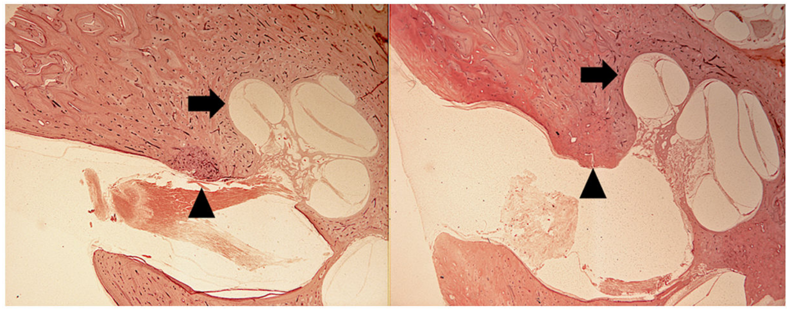

Figure 1a, 1b.
100x magnification H&E stains. Each of these slides demonstrate an otosclerotic focus abutting the IAC at the anterior, inferior aspect of the IAC—the typical site for a diverticulum. Arrow- basal turn of cochlea. Arrowhead- focus of otosclerosis.

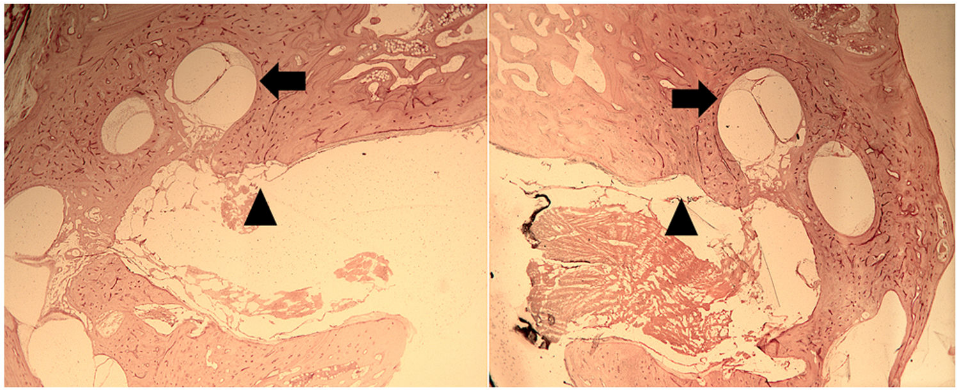

Figure 2a, 2b:
100x magnification H&E stains. Temporal bones #2, #3 from Table 1 demonstrate bilateral IAC diverticula in the same patient who has normal hearing and no histopathologic findings bilaterally. Arrow- basal turn of cochlea. Arrowhead- IAC diverticulum.

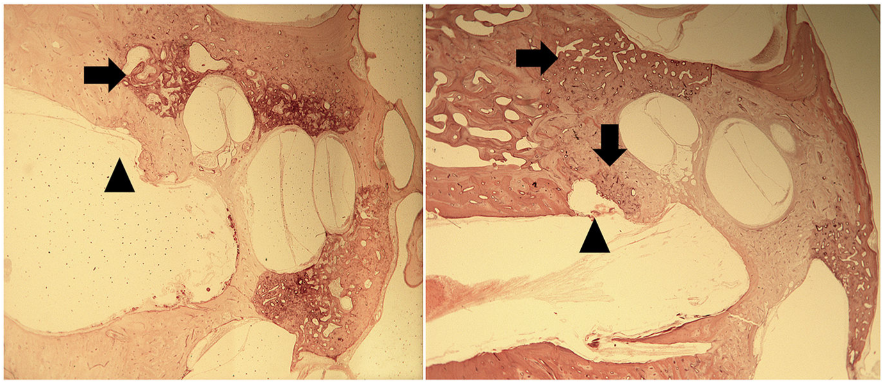

Figure 3a, 3b.

100x H&E stains. Two examples of IAC diverticula in the setting of retrofenestral otosclerosis that abuts the IAC. Arrow- retrofenestral otosclerosis. Arrowhead- IAC diverticulum.

Table 1:
Contralateral Histopathologic findings

List of contralateral histopathologic findings for the five normal temporal bones that demonstrated IAC diverticula.

Temporal bone number	Contralateral histopathologic findings
1	• Inadequate slides to evaluate region of interest • Vestibular schwannoma
2	• Normal temporal bone #3 with IAC diverticulum
3	• Normal temporal bone #2 with IAC diverticulum
4	• Coexisting IAC diverticulum (excluded from series due to ipsilateral pathology) • Vestibular schwannoma
5	• No IAC diverticulum • Scheibe deformity, absent organ of Corti, cochlear hydrops with rupture, deformed saccule, deformed tectorial membrane