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OTOPATHOLOGY REPORT

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Temporal bone histopathology: Superior semicircular canal dehiscence

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Abstract

Objectives: To present a histopathological case of a 91-year-old woman who was diagnosed with superior semicircular canal dehiscence postmortem.

Methods: The patient was a registered donor with the National Temporal Bone Donor Program at the NIDCD National Temporal Bone, Hearing and Balance Pathology Resource Registry. Computed tomography imaging was performed on each temporal bone. The temporal bones were decalcified with ethylenediaminetetracetate and embedded in celloidin, and tissue sections were stained with hematoxylin and eosin. Horizontal sections were taken through the left temporal bone, and vertical sections were taken through the right temporal bone.

Results: Histopathological sections taken through the right temporal bone demonstrated no bone between the membranous wall of the superior semicircular canal and the middle fossa dura. There was no histopathological evidence of superior semicircular canal dehiscence in the left temporal bone; however, a small dehiscence would not be identified on horizontal sections. Microcavitations were observed in the common crus of the left temporal bone.

Conclusion: This reports describes the case of a woman who was diagnosed with superior semicircular canal dehiscence postmortem. The presence of microcavitations in the temporal bone is consistent with osteoclastic activity, which may play a role in the development of superior canal dehiscence.

KEYWORDS

Superior semicircular canal dehiscence, histopathology

1 | INTRODUCTION

Superior semicircular canal dehiscence syndrome was first described by Minor et al in 1998.[1] Subsequent investigations demonstrated the syndrome consists of auditory symptoms, vestibular symptoms, or both. Auditory manifestations include autophony, aural fullness, hyperacusis, and an air-bone gap on audiometry. Vestibular symptoms include pressure-induced vertigo (Hennebert's sign), sound-induced vertigo (Tullio phenomenon), or chronic vertigo.

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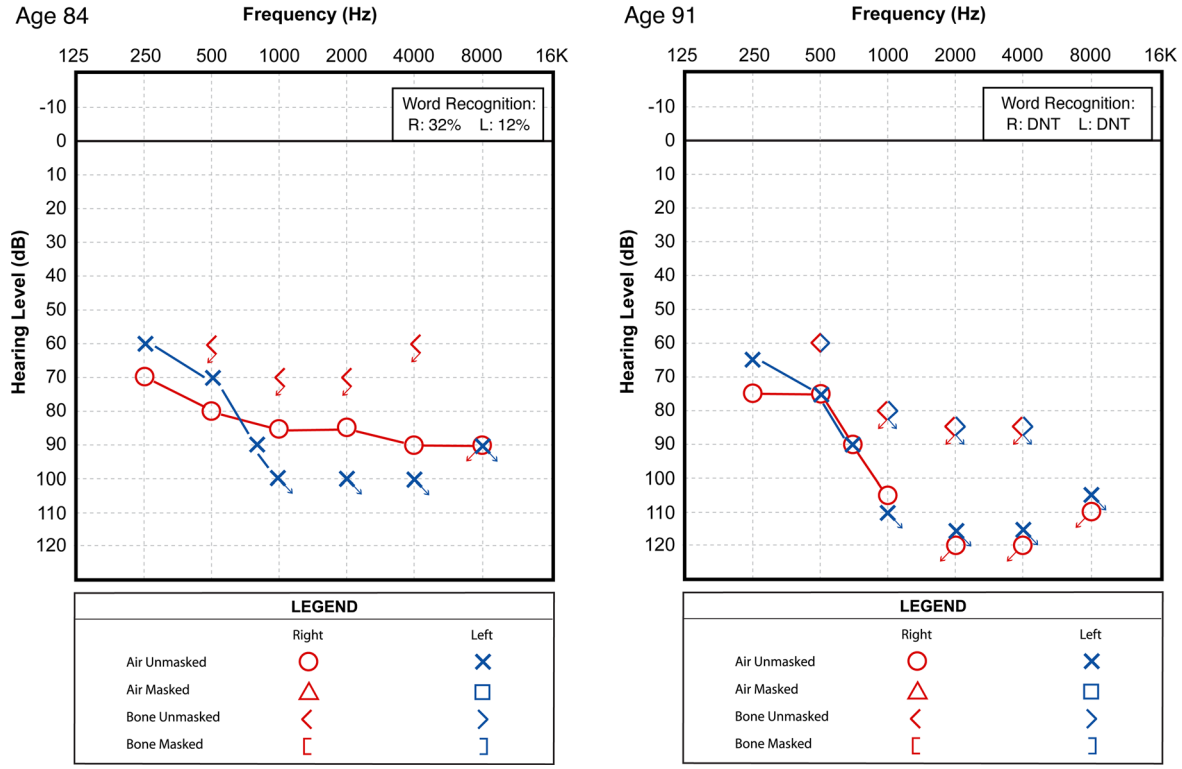

FIGURE 1 Audiometric evaluation at 85 years and 91 years of age

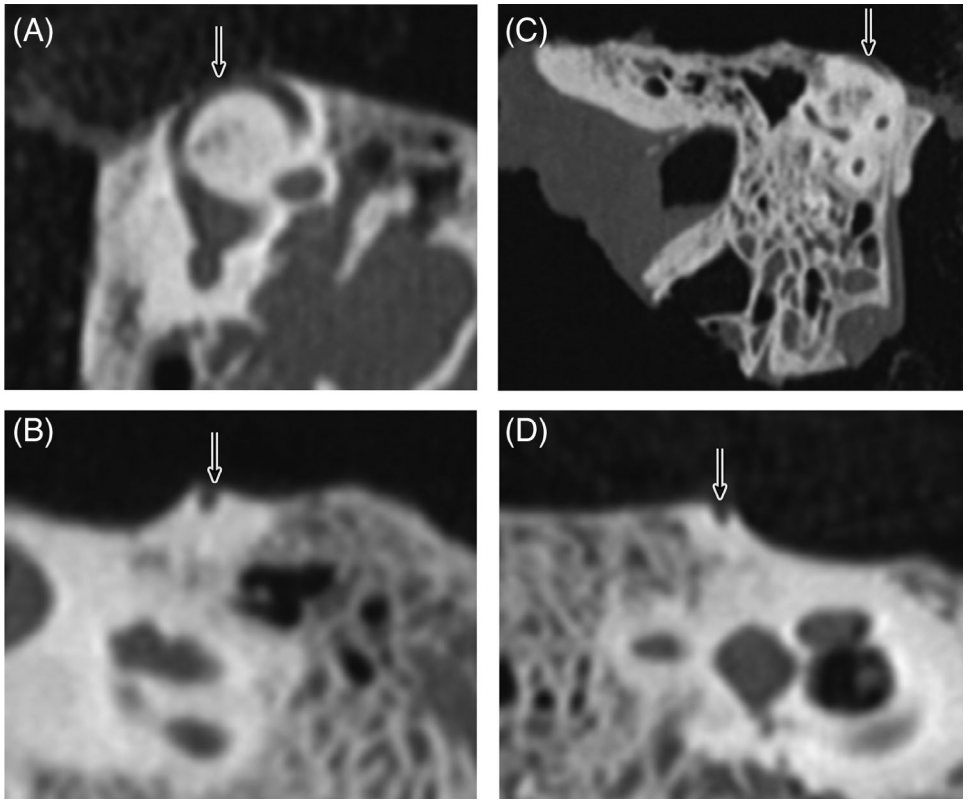

FIGURE 2 Computed tomography scans of the temporal bone: A, Pöschl view of the right ear; B, Stenver view of the right ear; C, Pöschl view of the left ear; and D, Stenver view of the left ear. These scans demonstrate canal dehiscence of the right superior semicircular canal (arrow), and dehiscence of the left semicircular canal. On the left, the bony partition between the superior semicircular canal and the middle fossa dura appears to be thinned but intact (C)

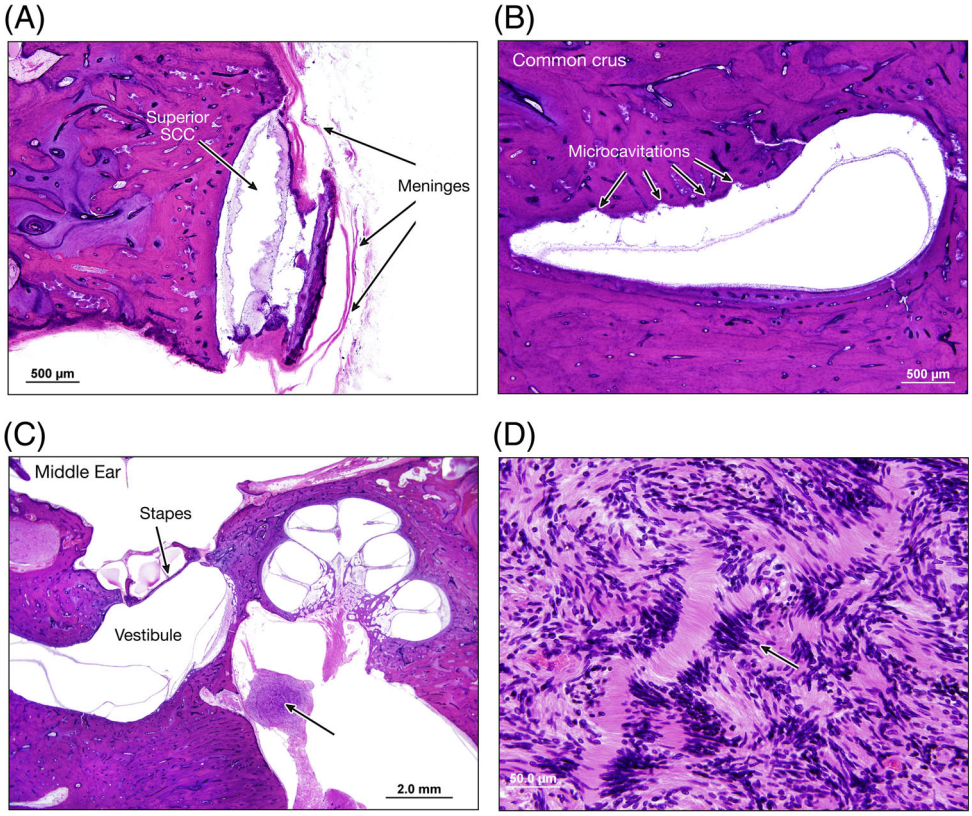

FIGURE 3 Histopathologic findings—horizontal sections through the left temporal bone. A, High-power view of the semicircular canal and meninges near the arcuate eminence. There is no obvious dehiscence of the superior semicircular canal but it would not be possible to fully assess this on horizontal sections. The disruption of the bony cap of the superior canal is due to processing artifact. B, High-power view of the common crus, demonstrating microcavitations. C, Incidentally discovered inferior vestibular nerve schwannoma (unlabeled black arrow). D, Higher power view of the vestibular schwannoma demonstrating spindle-shaped elongated cells in an Antoni A pattern (black arrow)

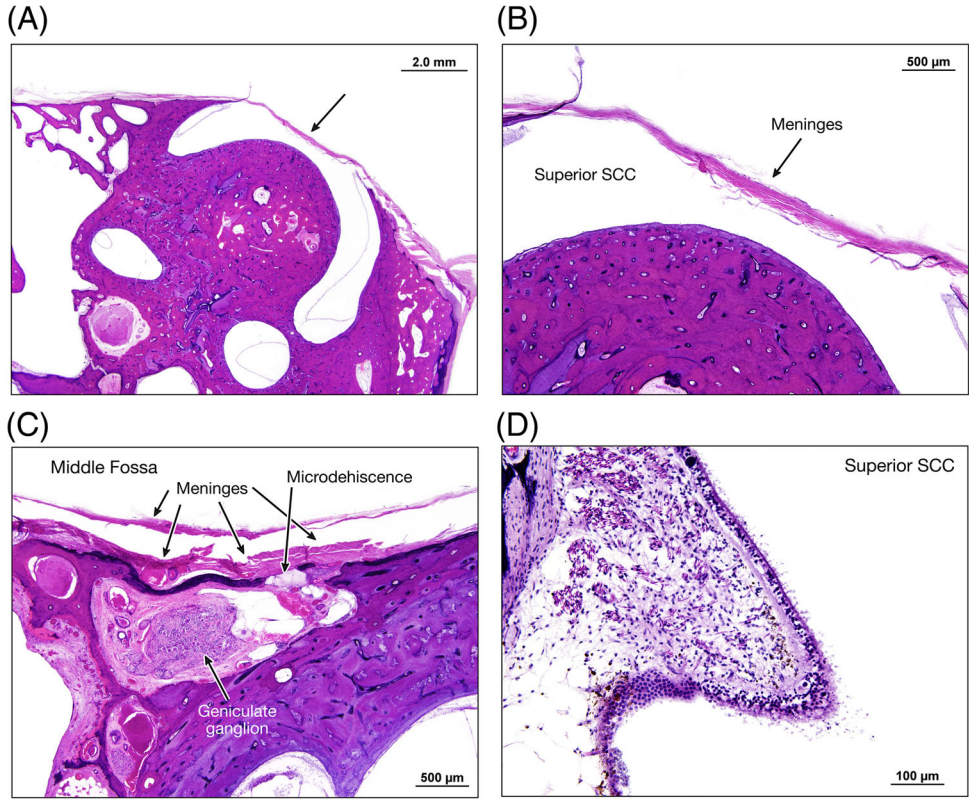

FIGURE 4 Histopathlogic findings—vertical sections through the right temporal bone. A, Low-power view demonstrating dehiscence of the superior semicircular canal (black arrow). B, Higher power view demonstrating the meninges is in contact with the membranous superior semicircular canal. C, A section through geniculate ganglion with a microdehiscence. D, A high-powered view through the superior semicircular canal crista ampullaris showing mild degeneration of the neuroepithelium

We present a histopathologic case of a patient who was diagnosed with superior semicircular dehiscence postmortem.

2 | MATERIALS AND METHODS

The patient was a woman who died of unrelated causes at age 91. She registered as a donor with the National Temporal Bone Donor Program at the NIDCD National Temporal Bone, Hearing and Balance Pathology Resource Registry. Bilateral ear canals were injected with embalming fluid within a few hours of death, and both temporal bones were removed 80 hours following death and placed in fixative. Computed tomography (CT) imaging was performed on each temporal bone, using multi-detector CT with 0.5 mm collimation with the same protocol that is used clinically. Temporal bones were then decalcified with ethylenediaminetetraacetate, and embedded in celloidin. The right temporal bone was sectioned vertically moving from medial to lateral along the trajectory of the internal auditory canal, and the left temporal bone was sectioned in the standard horizontal plane parallel to the middle fossa. Every 10th section was stained with hematoxylin and eosin, and slides were examined by light microscopy.

3 | RESULTS

3.1 | Clinical presentation

The patient is a 91-year-old woman who developed progressive hearing loss beginning in her 60s, which progressed to profound bilateral hearing loss. Audiograms available from ages 85 and 91 years showed progressive bilateral high-frequency sensorineural hearing loss (Figure 1). Tympanograms demonstrated a type A pattern, and acoustic reflexes were absent bilaterally.

The patient suffered from progressively worsening disequilibrium in her 80s, and eventually required a walker and a wheel chair. Her disequilibrium was attributed to a cerebrovascular accident that occurred in her 80s. Of note, the patient reported bilateral tinnitus, and self-reported sound-induced vertigo in the last 10 years of her life. Her past medical history was significant for coronary artery disease, hypertension, hypercholesterolemia, sleep apnea, depression, gastric ulcer, arthritis, and macular degeneration. Medications that the patient was taking regularly at the time of death were atenolol, losartan, hydrochlorothiazide, and warfarin.

Postmortem CT imaging performed prior to histologic preparation demonstrated bilateral superior semicircular dehiscence (Figure 2).

3.2 | Histopathology of left temporal bone

Horizontal sections were taken through the left temporal bone. As such, we were unable to adequately histologically assess for superior semicircular canal dehiscence. In horizontal sections taken at the arcuate eminence, there was no large dehiscence of the superior semicircular canal but a small dehiscence would not be identified on horizontal sections (Figure 3A). Microcavitations were observed in the common crus (Figure 3B). Moderate degeneration of the stria vascularis was observed. No organ of Corti was appreciated in the basal one-half of the cochlear duct. There was moderate degeneration of the vestibular neuroepithelium in the superior, lateral, and posterior semicircular canals, and minimal degeneration in the maculae utriculi and sacculi. Further assessment of the vestibular neuroepithelium was limited by postmortem artifact. There was an incidentally discovered histologically identified inferior vestibular nerve schwannoma (Figure 3C,D). The total corrected spiral ganglion count was 12 327, or 74% of normal for age.

3.3 | Histopathology of right temporal bone

Vertical sections were taken through the right temporal bone, running perpendicular to the trajectory of the internal auditory canal. This results in histologic sections similar to the Pöschl plane radiographically, that is, parallel to the plane of the superior semicircular canal. No bone was appreciated between the membranous wall of the superior semicircular canal and the middle fossa dura—confirming the presence of a histologic superior semicircular canal dehiscence (Figure 4A, B). There were microdehiscences of the tegmen mastoideum and at the geniculate ganglion (Figure 4C).

In the cochlea, a section through the mid-modiolus demonstrated no organ of Corti in the basal half of the cochlea, and decreased numbers of spiral ganglion cells in the basal turn. There was mild degeneration of the stria vascularis. The total corrected spiral ganglion cell count was 15 021, or 91% of normal for age. The superior, lateral, and posterior semicircular canals, as well as the utricle and saccule, all showed only minimal degeneration of the vestibular neuroepithelium and were well preserved on this side (Figure 4D).

4 | DISCUSSION

This report describes a 91-year old patient who died of unrelated causes, and postmortem, was diagnosed with right superior semicircular canal dehiscence both radiographically and histologically. The right ear showed dehiscence of the bone covering the superior semicircular canal, creating a true “third window” in the labyrinth with the dura adherent directly to the membranous labyrinth. The patient reported sound-induced vertigo for the last 10 years of her life, which in retrospect was likely secondary to SCD. She notably also suffered from general disequilibrium, which was likely multifactorial, but may be in part due to an incidentally discovered left inferior vestibular nerve schwannoma and moderate vestibular hair cell degeneration in the left ear. Notably, the vestibular neuroepithelium showed only minimal degenerative changes consistent with her age throughout all end organs in her right ear, including the dehiscent superior semicircular canal. Her sensorineural hearing loss was primarily due to significant degeneration of the organ of Corti and hair cell loss, which was symmetric bilaterally.

Possible etiologies of superior semicircular canal dehiscence include congenital and acquired causes. Congenital etiologies are supported by evidence that demonstrates the prevalence of SCD is

high in children less than 2 years of age, but decreases over the first decade of life, as well as the fact that there was no evidence of bony remodeling in five cases of dehiscence and 14 cases of near-dehiscence in the case series published by Carey et al.[2] Acquired causes include head trauma and an association between elevated intracranial pressure and SCD.

In the present case, microcavitations of bone suggesting osteoclastic activity were seen at the left ear common crus but not adjacent to the right-sided superior canal dehiscence. The presence of microcavitations in the temporal bone is consistent with osteoclastic activity, which may play a role in the development of SCD.[3] There were also microdehiscences of the mastoid tegmen and over the geniculate ganglion in this case, which is consistent with the hypothesis that superior canal dehiscence may be secondary to failure of postnatal bone development.[2] The visual depiction of the exposed geniculate ganglion is also a reminder to surgeons that care must be taken with elevation of the dura anteriorly over the geniculate ganglion during middle fossa surgery. Of note, similar to this case, a previous histopathologic study of the temporal bone in a patient with superior canal dehiscence syndrome did not find any nearby osteoclastic activity, vestibular hair cell loss or hydrops.[4]

In summary, this report describes a 91-year old woman who had progressive worsening high-frequency bilateral sensorineural hearing loss, progressive worsening disequilibrium resulting in walker and subsequently wheel-chair dependence, and self-reported sound-induced vertigo. Postmortem CT images demonstrated right superior semicircular canal dehiscence, and histopathologic sections confirmed this finding in the right ear. The adjacent labyrinthine bone and membranous labyrinth were normal, and the mild degenerative changes in the superior canal neuroepithelium were consistent with her age. Thus, the dehiscent right superior semicircular canal was not associated with any significant vestibular hair cell loss beyond typical age-related changes. The histologic correlate of the sensorineural hearing loss in this patient was degenerative changes in the organ of Corti and stria vascularis in both ears.

CONFLICT OF INTEREST

None declared.

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