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HISTOPATHOLOGY OF THE TEMPORAL BONE IN A CASE OF SUPERIOR CANAL DEHISCENCE SYNDROME

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Abstract

Objectives—To describe the histopathologic findings in the temporal bones of a patient with who had been diagnosed during life with superior canal dehiscence (SCD) syndrome.

Methods—The patient was diagnosed with SCD at the age of 59. She became a temporal bone donor, and died of unrelated causes at the age of 62. Both temporal bones were prepared in celloidin and examined by light microscopy.

Results—She developed bilateral aural fullness, pulsatile tinnitus and difficulty tolerating loud noises after minor head trauma at age 53. Symptoms were worse on the right. She also had valsalva-induced dizziness and eye movements, as well as sound-induced dizziness (more prominent on the right), consistent with SCD. Audiometry showed a small air bone gap of 10 dB in the right ear. VEMP testing showed an abnormally low threshold of 66 dB on the right, and CT scan showed dehiscence of the superior canal on the right. Histopathology of the right ear showed a 1.4×0.6 mm dehiscence of bone covering the superior canal. Dura was in direct contact with the endosteum and the membranous duct at the level of the dehiscence. No osteoclastic process was evident within the otic capsule bone surrounding the dehiscence. The left ear showed thin but intact bone over the superior canal. Both ears showed focal microdehiscences of the tegmen tympani and tegmen mastoideum. The auditory and vestibular sense organs on both sides were normal. No endolymphatic hydrops was observed.

Conclusions—The findings were consistent with the hypothesis put forth by Carey and colleagues that the SCD may arise from failure of postnasal bone development, and that minor trauma may disrupt thin bone or stable dura over the superior canal.

Keywords

temporal bone histopathology; otopathology; superior canal dehiscence; aural fullness; tinnitus

INTRODUCTION

The syndrome of dehiscence of bone overlying the superior semicircular canal was first described by Minor and colleagues ¹. That original report and subsequent papers by many investigators have shown that the manifestations of superior canal dehiscence (SCD) may be vestibular, auditory or both ^{2–9}. Vestibular manifestations include sound-evoked vertigo (Tullio phenomenon), pressure-induced vertigo (caused by coughing, straining, sneezing as

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well as by pneumatic otoscopy, i.e. Hennebert's sign), and chronic disequilibrium. Auditory manifestations include hearing loss manifested as an air-bone gap on audiometry, autophony for one's own voice, aural fullness, and hyperacusis in that patients may hear their own pulse, footsteps or eye movements.

Carey and colleagues¹⁰ surveyed 1,000 temporal bones from 596 adults contained in the collection at John's Hopkins Hospital and found five specimens with dehiscence of the superior canal, of which one showed SCD at the middle fossa floor, while four showed a dehiscence where the canal was in contact with the superior petrosal sinus. The diagnosis of SCD in these individuals was made after life by examination of their temporal bones, and the limited clinical records available suggested that these individuals were not symptomatic during life.

We had the opportunity to examine the histopathology of the temporal bones in a patient who was symptomatic and had a diagnosis of SCD made during life.

MATERIAL AND METHODS

The patient that is the subject of this case report was diagnosed with SCD syndrome at the age of 59. She registered as a donor with the National Temporal Bone Donor Program of the NIDCD National Temporal Bone, Hearing and Balance Pathology Resource Registry. She died of unrelated causes at the age of 62, and both temporal bones were removed 44 hours postmortem. They were processed for light microscopy by fixation in formalin, decalcification using ethylenediaminetetracetate, embedding in celloidin, serial sectioning in the vertical plane at a section thickness of 20 microns, and staining of every 20th section using hematoxylin and eosin¹¹. The study was approved by the institutional review board of the Massachusetts Eye and Ear Infirmary.

RESULTS

History and examination findings

The patient was seen at age 59 by author MT for complaints of bilateral aural pressure and tinnitus since experiencing minor head trauma against a car frame at age 53. She had difficulty tolerating loud noises, and her own voice reverberated in her ears. Chewing was heard very loudly inside her head. All these symptoms were worse on the right. She also had a sense of pounding in her right ear. She did not complain of dizziness or vertigo, nor did she hear her own breath sounds. Her past otologic history was noncontributory. Otoscopy revealed normal external auditory canals and tympanic membranes on both sides. Audiometry (Fig. 1) showed a mild high frequency sensorineural hearing loss bilaterally, with a small air bone gap of 10 dB in the right ear (bone thresholds were unmasked). Speech discrimination was 100% in both ears. Tympanometry and ENG were normal. Vestibular evoked myogenic potential (VEMP) testing showed an abnormally low threshold of 66 dB on the right and a low normal threshold of 78 dB on the left. CT scan of the temporal bones using 0.8 mm axial cuts was interpreted as showing definite SCD on the right and a probable SCD on the left. As the patient was not having vestibular complaints, conservative management was advised, and she underwent regular otologic monitoring.

At a subsequent examination, she was noted to have valsalva-induced dizziness and eye movements which could not be attributed to either ear with certainty. With the Barany noisemaker, she had bilateral sound-induced dizziness that was more prominent on the right, but no sound-induced eye movements. The head thrust test was normal. A Weber test (placed on the forehead) lateralized to the right. A tuning fork placed on the medial malleolus of the ankle was heard loudly in her right ear. She continued to have difficulty

tolerating noise, particularly in her right ear, and had pulsatile tinnitus in her right ear. The pulsations could not be modified by jugular venous compression.

She was diagnosed with pancreatic cancer at the age of 61. Despite surgery, radiation and chemotherapy, she succumbed to the malignancy at the age of 62.

Histopathology, right temporal bone (Fig. 2)

The external auditory canal, tympanic membrane, ossicles, middle ear, mastoid, oval and round windows, and otic capsule were normal. The tegmen tympani and tegmen mastoideum showed multiple focal bony dehiscences where mucosa was apposed to dura (Fig. 2A). None of the dehiscences showed an arachnoid granulation or meningocele.

The cochlea was fully developed, showing two and three-quarters turns. Reissner's membrane was in normal position in all turns without endolymphatic hydrops (Fig. 3). Although postmortem autolysis precluded an accurate assessment of cytologic elements within the labyrinth, it appeared that outer and inner hair cells were present in all turns. The stria vascularis showed focal areas of atrophy in the apical half of the cochlea. Cochlear neurons were present in numbers that were considered within the normal range for age.

There was a dehiscence of bone covering the superior semicircular canal (Fig. 2B). The dehiscence measured $1.4 \times .6$ mm, and showed dura apposed to the endosteum and the membranous canal. The fluid spaces within the canal at the level of dehiscence appeared normal. The superior semicircular duct was intact throughout, without any hydrops, distortion, collapse or other morphologic alteration of its wall. The bone of the otic capsule surrounding the dehiscence was extremely thin, but there was no histologic evidence of an active osteoclastic process. The vertical height of the superior canal (its radius in the vertical dimension) was smaller than average.

There were no dehiscences of the bone covering the posterior and lateral canals. The saccule, utricle, and cristae of all three semicircular canals showed good populations of hair cells, supporting cells and innervating dendrites, considered within the normal range for age. There was a normal population of cells within Scarpa's ganglion. The facial canal and nerve were normal. The jugular bulb was large and rose up to the level of the hypotympanum. There was a bony dehiscence between the jugular bulb and the vestibular aqueduct.

Histopathology, left temporal bone (Fig. 2)

The external auditory canal, tympanic membrane, middle ear and mastoid, ossicles and otic capsule appeared normal. The tegmen tympani and tegmen mastoideum showed focal areas of bony deficiency, similar to the contralateral side (Fig. 2C). The dura was apposed to mucosa in these dehiscences without arachnoid granulations or meningoceles. The cochlea, all three cristae and both maculae showed a good population of hair cells and neuronal innervation, similar to the opposite ear. There was no endolymphatic hydrops. The bone covering the superior semicircular canal was thin but intact throughout, measuring 0.1 mm in height at its thinnest point (Fig. 2D). The dura over the thin bone was normal.

DISCUSSION

This patient had fairly typical clinical manifestations of SCD in her right ear during life. Her symptoms began after relatively minor head trauma, which has been described in other patients with SCD¹⁻⁹. The predominant manifestations were auditory, consisting of aural fullness, autophony for her own voice, pulsatile tinnitus and hyperacusis. Although these symptoms were present in both ears, they were clearly worse on the right. Examination also showed valsalva-induced dizziness and eye movements, as well as sound-induced dizziness

in both ears, more so on the right. There was evidence of right ear 'conductive hyperacusis'⁸ as manifested by her ability to hear a tuning fork placed on the medial malleolus of the ankle in her right ear, and the Weber test lateralizing to the right. There was also a slight air bone gap on the right on audiometry, although it should be noted that the bone thresholds were unmasked. The VEMP test showed an abnormally low threshold on the right, and a CT scan was interpreted as showing a right SCD.

Histopathology confirmed SCD in the right ear. Dura was in direct contact with the endosteum and the membranous semicircular duct in the right superior canal at the level of the dehiscence. There were no other obvious abnormalities within the fluid spaces or crista of the superior canal. Light microscopy did not reveal any obvious osteoclastic process within the otic capsule bone surrounding the dehiscence. However, it did show multiple focal micro-dehiscences of the bone of the tegmen. The radius of the superior canal was also smaller than average. The histological findings are consistent with the hypothesis put forth by Carey et al¹⁰ that patients may be predisposed to SCD as a result of failure of postnatal bone development, as opposed to the alternative explanation of bone having developed normally and then eroding over the years. Our findings cannot determine if the dehiscence was present before she sustained the minor head trauma that resulted in symptoms. In another words, it is not known if the head trauma disrupted bone that was thin, or whether it disrupted the stability of dura over a pre-existing dehiscence.

Of interest are the symptoms that she experienced in her left ear, which also occurred after the episode of minor head trauma. Although not as severe as the symptoms on the right, she complained of aural fullness, autophony for her own voice and pulsatile tinnitus in her left ear. She also experienced vestibular symptoms in response to sound in this ear. Histopathology showed that the bone was thin but intact over the left superior canal, measuring 0.1 mm at the thinnest portion. One may speculate that if the thin bone covering the canal were sufficiently compliant, the area might function as a dehiscence, and that transmission of sound energy through the thin bone might explain some of her symptoms. A bony covering of high enough compliance would be consistent with some simple models of SCD effects on hearing and balance¹².

The lack of observed hydrops is also of interest. It has been noted that SCD may be associated with an abnormal summing potential/action potential (SP/AP) ratio on electrocochleography (ECoG) testing, similar to that described in cases of Meniere's syndrome¹³. Some have used this association to suggest that SCD must be associated with hydrops. While the lack of hydrops in one SCD case does not resolve the issue, we wish to point out that an abnormal SP/AP ratio may be a sign of underlying pathology associated with hydrops rather than hydrops itself. One possibility that may be consistent with both hydrops and SCD is an abnormal distribution of inner-ear static pressures. The presence of the SCD not only opens a window to sound-frequency stimuli, but would also open a window for the equalization of static pressure differences between the inner ear scalae and the cranial cavity. Just as SCD has been shown to mimic a wide range of disorders, such as stapes fixation, perilymphatic fistula and eustachian tube dysfunction, it may also mimic endolymphatic hydrops by evoking a change in the inner ear that is common to changes associated with hydrops.

Although the findings in this case may be helpful in understanding the pathology of SCD, it is difficult to make generalizations based on one case. Examination of the temporal bone in additional cases in the future, both unoperated and postsurgical, should provide more insight into the pathology and pathophysiology of this fascinating disorder.

SUMMARY AND CONCLUSIONS

This report describes a 62-year old patient who developed bilateral aural fullness, pulsatile tinnitus and difficulty tolerating loud noises after minor head trauma at age 53. Her own voice would reverberate in her ears. The symptoms were worse on the right. She also had sound-induced dizziness, as well as valsalva-induced dizziness and eye movements, consistent with SCD. Audiometry showed a small air bone gap of 10 dB in the right ear. VEMP testing showed an abnormally low threshold of 66 dB on the right, and a CT scan showed dehiscence of the superior canal on the right.

Histopathology of the right ear showed a 1.4×0.6 mm dehiscence of bone covering the superior canal. The left ear showed thin but intact bone over the superior canal. Both ears showed focal microdehiscences of the tegmen tympani and tegmen mastoideum. The auditory and vestibular sense organs on both sides were normal. The findings were consistent with the hypothesis put forth by Carey and colleagues¹⁰ that the SCD may arise from failure of postnasal bone development, and that minor trauma may disrupt thin bone or stable dura over the superior canal.

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Figure 1.

Audiometric evaluation at age 59 at the time of the diagnosis of SCD. There was a relatively flat, mild, sensorineural hearing loss bilaterally with a small air bone gap noted on the right. Note that the bone thresholds were unmasked. Speech discrimination was 100% in both ears.

A. RIGHT TEMPORAL BONE

B. RIGHT SUPERIOR CANAL

C. LEFT TEMPORAL BONE

D. LEFT SUPERIOR CANAL

Figure 2.

A. Low power view of vertical section from right temporal bone at the level of the superior semicircular canal. The external auditory, middle ear, ossicles and otic capsule were normal. There were multiple focal bony dehiscences of the tegmen (arrowheads), probably as a result of developmental anatomical variation. There were no instances of arachnoid granulation or meningocele at any of the dehiscences. There was dehiscence of the bone covering the superior canal, which is shown in more detail in B. TM, tympanic membrane.

B. Section through the superior semicircular canal on the right showing bony dehiscence. The dura showed artifactual separation of its layers. There were no abnormalities within the endolymphatic or perilymphatic fluid spaces of the canal. The bone of the otic capsule surrounding the dehiscence appeared normal without any evidence of an osteoclastic process. Inset shows higher power view of dehiscence. Dura was apposed to the endosteum and wall of the membranous duct within the dehiscence. The morphologic appearance of the dura and the semicircular duct was normal. The folded appearance of the dura was artifactual.

C. Vertical section through the left temporal bone at the level of superior canal showing thin bone covering the entire canal. The external auditory canal, middle ear, ossicles, mastoid air spaces and otic capsule were normal. There were multiple asymptomatic focal dehiscences of the tegmen (arrowheads). TM, tympanic membrane.

D. A higher power view of the thin bone over the superior canal on the left side. The dura had been artifactually separated into layers.

Figure 3. Mid-modiolar section from right temporal bone showing absence of endolymphatic hydrops. Reissner's membrane (arrows) is in normal position in all turns.