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Radiographic Features of Superior Semicircular Canal Dehiscence in the Setting of Chronic Ear Disease

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Abstract

Objective—To determine if radiologic chronic otitis media (COM), both with and without cholesteatoma, is associated with superior semicircular canal dehiscence (SSCD).

Study Design—Retrospective review of consecutive high-resolution computed tomography (HRCT) scans of the temporal bone.

Setting—Tertiary care medical center

Patients—Two-hundred consecutive patients undergoing HRCT of the temporal bone beginning January 1st, 2012.

Intervention—Imaging was evaluated by three reviewers (two neuroradiologists and one neurotologist). All scans were assessed for the presence of SSCD, cholesteatoma, chronic otomastoiditis, tegmen dehiscence, and for abnormalities of the cochlea, vestibule, facial nerve, and temporal bone vasculature.

Main Outcome Measure—Ears with COM associated with chronic otomastoiditis or cholesteatoma were compared to those without COM with respect to the presence of SSCD or other temporal bone abnormalities. Statistical analysis was performed to assess for differences between the groups studied.

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Results—194 patients (388 ears) were included. Cholesteatoma was identified in 48 ears (12.4%) and chronic otomastoiditis in 62 ears (16%). Ten ears with cholesteatoma had ipsilateral SSCD and 8 ears with chronic otomastoiditis had ipsilateral SSCD. In 340 ears without either cholesteatoma or chronic otomastoiditis, SSCD was found in 18 (5.3%). SSCD was found to occur significantly more often in patients with ipsilateral radiologic cholesteatoma. No cases of SSCD were associated with cochlear, facial nerve, or vascular abnormalities.

Conclusion—Our findings suggest that COM with cholesteatoma is associated with the presence of SSCD, although the nature of this association is unclear.

INTRODUCTION

Superior Semicircular Canal Dehiscence (SSCD) is defined as an absence of bony covering of the membranous labyrinth of the superior semicircular canal. In some patients, this finding is associated with a constellation of symptoms including autophony, aural fullness, sound and pressure-induced vertigo, tinnitus, and conductive hearing loss (1). The prevalence of SSCD is unknown and varies depending on the detection modality utilized. In cadaver temporal bone specimens, the prevalence of SSCD has been reported to be 0.5%, with an additional 1.4% of specimens displaying an abnormally thin tegmen overlying the superior semicircular canal (2). The prevalence of SSCD on high resolution computed tomography (HRCT) imaging is generally accepted to be higher than that in cadaver studies and ranges from 3-12% (3,4,5,6,7,8). These differences are attributed to limitations of HRCT imaging, including an inability to identify very thin layers of bone overlying the semicircular canal and to partial volume averaging effects (5).

The etiology of SSCD is currently not well understood, and both congenital and acquired processes have been implicated in its pathogenesis. Evidence supporting a congenital etiology includes the observations that SSCD syndrome exists in children (9) and that SSCD has been identified in patients with congenital anomalies such as malformations of the labyrinth (10) and brainstem (11). However, the findings that SSCD is associated with increased age (7), osteopenia (12), and trauma (2) support an acquired cause of SSCD. Another postulated mechanism of the development of SSCD relates to abnormal postnatal middle fossa bone growth during development. This theory is supported by the observation of Carey et al. that temporal bones with thinning over the superior semicircular canal or with dehiscence of the canal display bony architectural patterns similar to those observed in infant temporal bones in which the adult middle fossa plate configuration has not yet formed (2). In this proposed mechanism, the symptoms of the syndrome associated with SSCD are thought to develop after a second insult (trauma, increased intracranial pressure, osteoporosis) occurs that results in violation of this abnormally thin and susceptible bone overlying the superior semicircular canal, thus leading to a functional third window effect (2).

Although these several mechanisms have been proposed to explain the development of SSCD in some subjects, to date no single mechanism has been confirmed to account for all cases of SSCD. It is therefore possible that other processes that act on the structure of the temporal bone could predispose individuals to the development of SSCD. For example, it is well documented that chronic otitis media (both with and without cholesteatoma) results in characteristic changes to the architecture of the temporal bone, including a decrease in mastoid pneumatization and volume (13, 14), formation of new sclerotic bone (15), and changes in position of the sigmoid sinus (16, 17). It is currently not known if SSCD accompanies these changes in temporal bone structure caused by chronic otitis media (COM). The aim of this study is to determine if patients with radiological findings of COM (with or without cholesteatoma) have a higher incidence of SSCD than patients without COM.

MATERIALS AND METHODS

This study was reviewed and approved by the University of Wisconsin Institutional Review Board. The Department of Radiology, Division of Neuroradiology database was searched to identify 200 consecutive patients undergoing High-Resolution Computed Tomography (HRCT) of the temporal bone over a 14 month period beginning January 1, 2012. All scans were performed with 0.625 mm collimation. Images were reviewed by three different evaluators (two Neuroradiologists and one neurotologist). All evaluators were blinded to the indication for the CT imaging and to the clinical history of the patient. Exclusion criteria included severe motion artifact and failure to include all standard HRCT views, including coronal, axial, sagittal, Stenver, and Poschl views. The Stenver view is an oblique coronal reconstruction that is parallel to the petrous portion of the temporal bone. A Poschl view is an oblique coronal reconstruction that is perpendicular to the petrous portion of the temporal bone. Scans were also excluded if severe trauma, extensive surgery, or a unique medical condition (fibrous dysplasia) limited the ability to assess the structures in question. Patients who underwent prior mastoid surgery were not excluded from the study.

All consecutive scans were reviewed bilaterally for the presence or absence of SSCD, cholesteatoma, chronic otomastoiditis, geniculate ganglion dehiscence, tegmen dehiscence, and for the presence of other temporal bone abnormalities (vascular, facial nerve, vestibular, and cochlear anomalies). Cholesteatoma was defined as a nondependent middle ear soft tissue attenuation associated with bony erosion of the scutum (Figure 1). Chronic otomastoiditis was classified as any ear with evidence of sclerotic mastoid air cells with or without associated soft tissue opacification and absence of bony erosion (Figure 2). Both cholesteatoma and chronic otomastoiditis, as defined, were considered to each be manifestations of chronic otitis media. Furthermore, cholesteatoma and chronic otomastoiditis were assessed separately and were not considered mutually exclusive. It was therefore possible for patients to have a diagnosis of both cholesteatoma and chronic otomastoiditis if individual radiologic criteria for each of these conditions was met. The location of canal dehiscence (ascending limb, apex, descending limb, or involvement of two contiguous regions) was documented when dehiscence was identified (Figure 3). If two adjacent subsites of the superior semicircular canal were dehiscent, the location of the dehiscence was classified as “> 1 subsite” for that ear. No patients had involvement of two non-contiguous subsites.

Statistical analysis was performed to assess for differences between groups of patients with and without SSCD with respect to the radiologic presence of cholesteatoma, chronic otomastoiditis, tegmen dehiscence, and the other findings listed above. Analysis was conducted using SAS 9.2 (SAS Institute, Cary NC) software. A p-value < 0.05 was considered significant in two-tailed statistical tests.

RESULTS

A total of 200 HRCT scans were reviewed. Six patients were excluded (one with severe temporal bone trauma, one in which motion artifact limited review of the imaging findings, one subject in which all standard HRCT views were not performed, one subject who underwent prior lateral temporal bone resection, one subject who underwent resection of a glomus tumor, and one subject with fibrous dysplasia of the temporal bone) due to inability to assess the available imaging for all middle and inner ear structures included in the study. A total of 194 patients (388 ears) were included in the study. A total of 21 patients (28 ears) were identified to have SSCD, seven of them bilaterally. Therefore, SSCD was identified in 10.8% of patients and 7.2% of ears.

When considering the incidence of SSCD in ears with and without cholesteatoma, 48 ears were determined to demonstrate cholesteatoma on radiographic imaging. Ipsilateral SSCD was identified in 21% of these ears (Figure 4). In contrast, only 5.3% of ears without cholesteatoma demonstrated SSCD (Figure 5). This finding was statistically significant ($p = 0.0011$). A similar difference in the incidence of SSCD in ears with and without chronic otomastoiditis was observed. Figure 6 shows that SSCD was observed in 12.9% of ears with ipsilateral chronic otomastoiditis. In contrast, SSCD was only observed in 6.1% of ears without evidence of chronic otomastoiditis. Although this trend was found to be present, it was not found to be statistically significant ($p = 0.25$).

In looking at the presence of SSCD in contralesional ears, the incidence of SSCD in ears contralateral to those with only cholesteatoma was found to be 12.5% while the incidence of SSCD in ears contralateral to those with both SSCD and cholesteatoma was found to be 30%. This difference was not statistically significant ($p = 0.095$).

When assessing other middle and inner ear features, when the specific region of middle fossa bone overlying the geniculate ganglion was assessed, dehiscence of the geniculate ganglion was identified in 39.3% of ears associated with ipsilateral SSCD, compared to an incidence of a dehiscence of the geniculate ganglion in 11% of ears that were not associated with SSCD. This difference was statistically significant ($p = 0.0002$). When documenting the location of identified superior semicircular canal dehiscence, the apex of the superior canal was the most common dehiscence location (42.9%), followed by the ascending limb (35.7%; Table I). Only three dehiscences (10.7%) were associated with the posterior limb of the superior semicircular canal, one of which was caused by apposition of the superior petrosal sinus (Figure 7). No patients with SSCD demonstrated cochlear, ossicular, facial nerve, or vascular middle or inner ear anomalies.

DISCUSSION

Our findings of a radiologic incidence of SSCD of 10.8% in the study population and an incidence of bilateral dehiscence of 33.3% are in agreement with previous literature which documents the radiologic incidence of SSCD between 3-12% (3,4,5,6,7,8,9) and the incidence of bilateral dehiscence between 25-60% (7,18). Our observations also provide new information regarding SSCD. First, our data indicate that dehiscence of the ascending limb of the superior semicircular canal may be more common than previously thought. Indeed, the finding that the area of dehiscence was localized to the ascending limb in 35.7% of dehiscences is higher than in previous reports (3). Our results also indicate a possible association between dehiscence of the geniculate ganglion and SSCD.

Next, our results demonstrate a statistically significant association between a radiologic diagnosis of cholesteatoma and SSCD. Although the mechanism behind this association remains unknown, several possibilities exist that may explain this observation. One possibility relates to the theory of SSCD development proposed by Carey et al. that SSCD results from a second insult to a thin bony covering over the superior semicircular canal due to a failure of postnatal bone growth in this area (2). Our observation of a relationship between cholesteatoma and SSCD may be evidence that cholesteatoma serves as one such “second insult” on an abnormally thin tegmen in these individuals, although the exact method by which cholesteatoma influences the development of SSCD in this manner is unknown. Our finding that the incidence of SSCD in ears contralateral to those with cholesteatoma is not significantly different from the incidence of SSCD in ears contralateral to both SSCD and cholesteatoma further supports this theory, as it suggests that factors unrelated to processes present in the contralateral ear may be contributing to the development of SSCD in the opposite ear. With further study, it may become more clear if

the formation of SSCD in individuals with thin middle fossa bone is directly influenced by the presence of ipsilateral cholesteatoma.

Another possibility may be that SSCD associated with ipsilateral cholesteatoma may occur by a different mechanism than other cases of SSCD. For example, it is possible that the direct effects of inflammation known to be associated with cholesteatoma may influence the architecture of the temporal bone, thereby inducing formation of SSCD. Cholesteatoma has been associated with increased production of several inflammatory cytokines which have been shown to influence the bony architecture of the middle ear and mastoid (19). For example, cholesteatoma has been shown to result in an increased expression of MIB-1, a marker of cellular proliferation (20). MIB-1 has been implicated in osteitic destruction processes commonly observed in association with cholesteatoma (lateral canal, ossicular, and tegmen erosion), with higher levels of MIB-1 detected in cases of more severe bony damage (21). Cholesteatoma has also been observed to incite expression of MMP-9, an inflammatory cytokine associated with cellular destruction (22). It is conceivable that the presence of these abnormal inflammatory changes occurring in the temporal bone in conjunction with cholesteatoma may influence the architecture of the otic capsule in the region of the superior semicircular canal resulting in dehiscence. It is not clear why the superior semicircular canal would be more prone to these effects than other parts of the otic capsule, however one might posit that the continuous, pulsatile pressure from the overlying middle fossa dura leaves the bone overlying the superior semicircular canal prone to erosion when adjacent inflammation in the temporal bone is present. Our finding that the apex of the superior semicircular canal is the most common location of dehiscence supports this theory, as this region of the superior canal is the subsite nearest to the overlying middle fossa dura. However, this mechanism would not explain other cases of SSCD at locations other than the canal apex that are associated with cholesteatoma. With further study, the association and causality of SSCD in the presence of COM associated with cholesteatoma may become more clear.

The observations in our study have several important implications to temporal bone surgeons. First, temporal bone surgeons should be aware of the observed association between radiologic cholesteatoma and ipsilateral SSCD as this may alert them to the possible existence of SSCD in these patients, which may help guide preoperative counseling. Secondly, our findings may serve to make the temporal bone surgeon more cautious intraoperatively when dissecting cholesteatoma near the vestibule so as to avoid inadvertent elevation of the dura over the superior semicircular canal. Finally, our observations may assist in the workup of patients with cholesteatoma in whom vestibular symptoms are present and no other identifiable fistulae (such as a lateral canal fistula) can be identified on computed tomography.

Our study has a number of limitations. First, the study is retrospective and included only individuals who underwent HRCT at a tertiary care institution. Second, this work was a radiologic study and did not assess for symptoms of SSCD syndrome in the subjects studied. Therefore it cannot be determined from our data if COM, with or without cholesteatoma, may lead to symptoms of SSCD syndrome. Last, we did not review the clinical charts of the patients in our study as our goal was to determine if there was association between the radiological findings of COM and SSCD. As such we cannot determine if patients with the radiological findings of cholesteatoma or chronic otomastoiditis had ongoing clinical ear disease at the time of the study. Despite these limitations, the association of SSCD with ipsilateral cholesteatoma has not been described previously and suggests a risk factor for the formation of SSCD in some individuals.

CONCLUSION

Our findings suggest that COM with cholesteatoma is associated with the presence of SSCD, although the nature of this association is unclear. An awareness of this association is prudent for the temporal bone surgeon and could assist in preoperative counseling and in prevention of inadvertent injury to the superior semicircular canal and overlying dura in some circumstances.

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Figure 1. Coronal (A) and axial (B) CT images of the left temporal bone demonstrate non-dependent soft tissue within the left external auditory canal that extends into Prussak's space (long arrows) and associated blunting of the scutum (short arrow) which are features of an acquired cholesteatoma.

Figure 2.
An axial CT image of the right temporal bone shows complete opacification of the right middle ear cavity and mastoid air cells which are thickened and sclerotic which are features of chronic otomastoiditis.

Figure 3.
A Poschl view of the left temporal bone illustrates the 3 divisions of the superior semicircular canal: ascending limb (long arrow), apex (arrow head) and descending limb (short arrow). This patient has normal osseous covering of the superior semicircular canal.

Figure 4. Poschl (A) and coronal (B) CT images of the left temporal bone show dehiscence of the apex of the left superior semicircular canal (long arrow) and non-dependent soft tissue with Prussak's space associated with erosion of the scutum (short arrow).

Incidence of SSCD in Ears with and without Cholesteatoma

Figure 5. Diagram depicting characteristics of patients with cholesteatoma with and without superior semicircular canal dehiscence.

Incidence of SSCD in Ears with and without Chronic Otomastoiditis

Figure 6.

Diagram depicting characteristics of patients with chronic otomastoiditis with and without superior semicircular canal dehiscence.

Figure 7.
CT reconstructions of the left temporal bone in the Stenvers(A) and Poschl (B) projections demonstrate dehiscence of the left superior semicircular canal related to abutment of the superior petrosal sinus (arrows).

Table I

Characteristics of Superior Semicircular Canal Dehiscence by Location

Location	Right	Left	Total #	Percent (%)
Ascending Limb	4	6	10	35.7%
Apex	8	4	12	42.9%
Descending Limb	1	2	3	10.7%
> 1 Continuous Location	0	3	3	10.7%