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Semicircular canal dehiscence

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In the evaluation of patients with suspected superior (or posterior or lateral) semicircular canal dehiscence syndrome, imaging plays a crucial corroborative diagnostic role. The variable audiologic and vestibular signs and symptoms may be attributed to a third-window phenomenon, directly visible on computed tomography as a defect in the superior or posterior semicircular canals. While the diagnosis must incorporate computed tomography findings with the signs, symptoms, and clinical testing, imaging also provides anatomical information on dehiscence size and location that may help explain the specific clinical presentation and aid in surgical planning.

Introduction

Superior semicircular canal dehiscence syndrome is a relatively recently described entity in which imaging plays a fundamental diagnostic role. Minor[1] correlated computed tomography (CT) findings with clinical symptoms to identify a syndrome of vertigo induced by sound or pressure or both that is attributed to a third-window mechanism of labyrinthine pressure alterations. The initial clinical symptoms may overlap with other pathologies, and imaging complements the clinical evaluation with auditory and vestibular testing to confirm the diagnosis, exclude other differentials, and aid the presurgical planning. Studying the relationship between imaging and clinical findings also aids the investigation of the pathophysiology of semicircular canal dehiscence.

Signs and symptoms

A variety of vestibular, auditory, or both auditory and vestibular symptoms are attributed to defects in the osseous covering of a semicircular canal. Signs and symptoms include vertigo induced by sound or pressure or both or nystagmus (Tullio phenomenon), conductive hearing loss, autophony, oscillopsia, chronic disequilibrium and motion intolerance, hyperacusis, and gaze-induced nystagmus.[1] Patients may have an apparent conductive hearing loss that may initially mimic otosclerosis, though further testing can show additional features that distinguish these conditions.[2]

Normally, sound-induced pressure waves in the scala vestibuli are transmitted through the scala media and basilar membrane, to the scala tympani, which is relatively compliant owing to the presence of the round window. The dehiscence, or third window, increases the compliance of the perilymph of the semicircular canals, resulting in transmission of the pressure wave to the semicircular canals, motion of the vestibular endolymph, and vestibular symptoms. Dissipation of the pressure wave into the vestibule also produces conductive hearing loss. Improved bone conduction is seen at low frequencies, which may also be due to changes in the impedance difference between the scala vestibule and scala tympani across the basilar membrane.[2] These changes are reversible following patching of the dehiscence.[2-4]

Shunted pressure from air conduction produces an ampullofugal deflection of the cupula and a perception of motion manifested as dizziness and findings of nystagmus. Sound- or pressure-induced vertical-torsional eye movements lie along vectors that predict the specific dehiscent semicircular canal.[5-8] The degree of air-bone gap (ABG)

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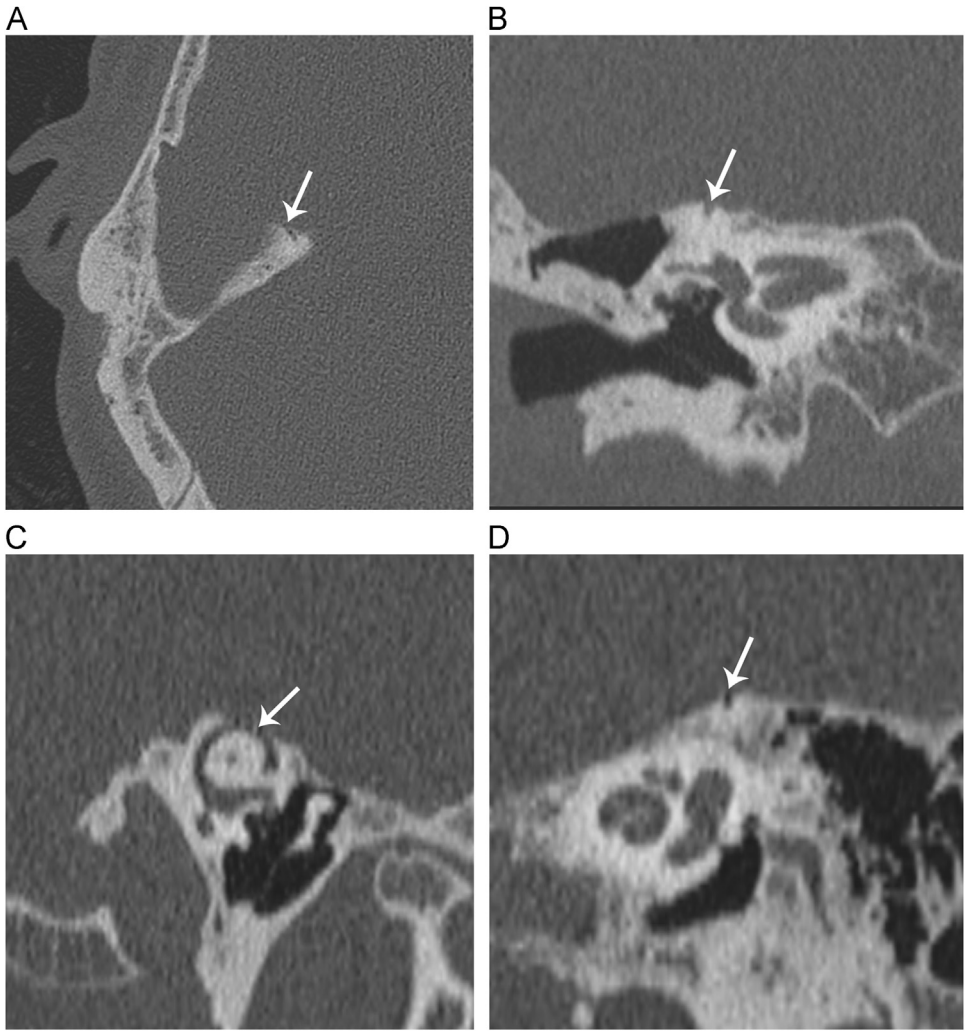

Figure 1 Superior semicircular canal (SSC) dehiscent to the cranial fossa. (A) Axial CT through the level of the SSC shows a focal dehiscence at the anterior aspect of the arcuate eminence (arrow). (B) Coronal CT reformation shows SSC dehiscence (arrow). (C) Poschl plane reconstruction, which displays the SSC nearly in its entirety, better depicts the extent of the dehiscence (arrow). (D) Stenvers plane reconstruction, perpendicular to the SSC, also helps confirm dehiscence (arrow).

may be related to the length of the dehiscence,[5] location of the dehiscence,[3,9-11] and the cross-sectional area of the dehiscence.[3,9,12,13] CT studies may help guide investigation of these effects.

Etiology

Although the etiology of semicircular canal dehiscence is still unknown, several authors favor a congenital etiology due to failure of postnatal bone development.[14] Patients with canal dehiscence may have generally reduced osseous thickness of the temporal bone with a thinner calvarium along the squamousal portion of the temporal bone[15] and a greater association with tegmen defects.[16,17] Dehiscence is frequently bilateral or associated with thinned bone over the contralateral semicircular canals.[14] The increased CT incidence of semicircular canal dehiscence in children younger than 2 years compared with adults supports a possible congenital basis with discontinuation of bone maturation or deposition, but it also suggests that the presence of dehiscence does not correlate with the presence of symptoms.[18] The development of symptoms later in life implies that congenitally thin or dehiscent bone or non-compliant dura requires an acquired, second event, such as trauma or increased cerebrospinal fluid pulsations from elevations in intracranial pressure, to induce symptomatic dehiscence or dural compliance.[14] Other authors have proposed an acquired etiology, noting the increased incidence of superior semicircular canal dehiscence with age, suggesting a process of systemic demineralization increasing with age or repetitive microtrauma.[19]

Incidence

Based on a review of temporal bone pathology specimens, superior semicircular canal dehiscence may be found in 0.7% of individuals and 0.5%-0.6% of temporal bone specimens, with an additional 1.3% of individuals and 1.4%