Incidence and Clinical Significance of Superior Semicircular Canal Dehiscence

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Introduction

Superior semicircular canal dehiscence (SSCD) syndrome was first described by Minor et al. in 1998. Dehiscence of bone overlying the superior semicircular canal has been associated with sound or pressured induced vertigo and oscilloscopia. Hyperacusis, bone-conducted sounds and conductive hearing loss has also been demonstrated in these patients. Tullio phenomenon and Hennebert sign have also been associated with hyperacusis, perilymphatic fistula, and Meniere’s disease.

In SSCD syndrome, it is thought that the presence of a “third window” in addition to the round and oval windows leads to these symptoms. Maneuver’s that cause a positive pressure gradient from the middle ear to the labyrinth such as Valsalva against pinched nostrils and positive pressure in the external auditory canal lead to excitation of the SSC. This third mobile window allows dissipation of this pressure and endolymphatic flow away from the ampulla resulting in an excitatory amplitudolographic deflection of the SSC cupula. The true incidence of persons with symptomatic SSCD is currently unknown. One study of 1000 cadaveric temporal bones revealed that a dehiscence of bone that overlies the superior canal was present in approximately 0.5% of temporal bone specimens. In an additional 1.4% of the specimens, the bone was markedly thin (≤0.1 mm) compared with the normal bone. Martin et al. reported a 3.67% incidence of SSCD on high resolution computed tomography (HRCT) amongst 272 patients presenting with isolated unilateral or bilateral conductive or mixed hearing loss.

In this study, we aimed to determine the incidence of SSCD amongst all comers at our institution who underwent HRCT of the temporal bone. In addition, we sought to correlate the presence of SSCD with the presence of vestibular signs and symptoms. We hypothesize that the incidence of SSCD in the population is much greater than those suffering from true SSCD syndrome.

Methods

This retrospective chart review was approved by our institutional review board. All temporal bone HRCT scans performed from October 2009 to June 2010 were enrolled in the study. These images were analyzed by our neuroradiologist (S.K.) for the presence of SSCD. The radiologist was blinded to the prior history and clinical indication for the HRCT scan. Patients with a history of cholesteatoma, mastoidectomy, malignancy, or temporal bone trauma were excluded.

A thorough chart review was performed on each patient. Specific otologic symptoms associated with SSCD were collected and recorded in a database. These included: Tullio phenomenon, Hennebert sign, autophony, hyperacusis to bone-conducted sounds and oscilloscopia. In addition, the presence of vertigo, tinnitus, aural fullness and nystagmus was recorded. Audiogram, VEMP, and ENG testing were also recorded when they had been performed.

Results

A total of 160 HRCT scans were performed during the above time frame. Sixty-nine patients met our inclusion criteria for a total of 138 temporal bones analyzed. After thorough review by our neuroradiologist, a total of 7 temporal bones in 5 patients were found to have SSCD. Two patients were found to have bilateral dehiscence while three were unilateral. Therefore, the incidence of SSCD in our study population is 5%.

A thorough chart review was performed on all patients included in the study. Of the patients identified with SSCD, 4/5 (80%) presented with tinnitus and were found to have mixed hearing loss on audiometry. Only one patient presented with classic signs/symptoms of SSCD including Tullio phenomenon and Hennebert sign. This patient also reported tinnitus and aural fullness on the left. This patient’s audiogram revealed a left mixed hearing loss. This patient was found to have a left SSCD. Our incidence of SSCD syndrome was therefore 1.4%.

Discussion

Since its description by Minor in 1998, SSCD as a clinical entity has been investigated in many ways. There remains controversy regarding the appropriate imaging techniques for accurate diagnosis. In addition, the true incidence of SSCD remains unknown. Our results demonstrating a 5% incidence compare favorably to recent radiographic studies. Williamson et al. identified radiographic evidence of SSCD in 39 of 442 temporal bones analyzed. Out of the 272 patients studied, 1.4% of temporal bones studied were identified as having extreme thinning (≤0.1mm) of the bone overlaying the SSC. This study is thought to be the closest representation to the actual incidence of SSCD as it is well documented that the radiographic incidence of dehiscence appears to be much higher. The inability to accurately identify bone on high-resolution CT imaging with a thickness of less than 0.324 mm is likely responsible for this disparity and the over diagnosis of SSCD.

The clinical significance of SSCD is still not completely understood. It is suspected that the prevalence of SSCD is much greater than the number of those actually suffering from SSCD syndrome. Amongst the 5 patients identified in our study with radiographic SSCD, only one presented with the classic features as described by Minor. More interesting is the finding of tinnitus and mixed hearing loss in 80% of our patients with SSCD. Martin et al. studied 272 patients with a normal tympanic membrane who suffered from isolated unilateral or bilateral conductive or mixed hearing loss. Out of the 272 patients, 3.67% of them were found to have SSCD. The third labyrinthine window theory of Minor is the most common theory accepted to explain the conductive or mixed hearing loss encountered in patients with a SSCD. As demonstrated by Martin, isolated conductive or mixed hearing loss is a relatively common clinical entity and a small percentage of these patients will be found to have SSCD as the underlying cause. Consideration should be made to obtain HRCT imaging on all patients presenting with conductive or mixed hearing loss of otherwise unknown etiology to rule out SSCD.

Conclusions

The incidence of SSCD amongst our patient population was found to be 5%. The majority of patients with SSCD presented with tinnitus and mixed hearing loss. Although uncommon, a high index of suspicion is necessary to diagnose SSCD in patients with common otologic complaints of unknown origin.

References