Dehiscences of the semicircular canals as discrete third window lesions of the inner ear

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Abstract The aim of this study was to assess the role of high resolution multi-detector CT in diagnosis of different dehiscences of the semicircular canals (SCC).

Material and Methods: This study was conducted on 20 patients within one year interval. All the patients had given clinical diagnosis of dehiscences of the SSC by clinical examination and/or audiogram. Sixty-four MDCT was used in assessment of all patients.

Results: Congenital defects of the SCC were encountered in 9 cases (45%); acquired defects were encountered in 11 cases (55%). Among the congenital defects, superior and posterior SCC were involved in 6 and 3 patients (30 and 15%) respectively. Congenital defects of lateral SCC were not encountered in this study. Cholesteatoma was the most common single etiology causing defect within the SCC.

Conclusion: Radiological diagnosis of SCC dehiscences depends on the use of high resolution MDCT. Ultrathin reconstruction with MPR is mostly sufficient in accurate display of the defects. Primary congenital dehiscences do occur in the superior SCC but also less commonly within the posterior canal. In this study, lateral canal dehiscence was only acquired. Secondary semicircular canal dehiscences could be caused by cholesteatoma and temporal bony aggressive mass lesions.

1. Introduction

Congenital dehiscences of the semicircular canals were first described by minor et al. (1998), a syndrome related to the dehiscence of bone overlying the superior semicircular canal (SSC), including vertigo, oscillopsia, and/or dysequilibrium resulted from sound, changes in middle ear pressure, and/or changes in intracranial pressure in these patients. (1) Generally it is a rare condition, and usually symptomatic in adults. (2) Developmental theory is suggested by the fact of bilateral involvement of superior semicircular canals (3,4).
The dehiscent bone acts as a “third mobile window” (4,5). Normally, the oval and round windows are the only two openings in the hydraulic system of the inner ear, while semicircular canals represent a hydraulically closed system. If there is creation of a “third window”, the brain interprets motion of the endolymph as movement of the body, and the patient feels dizzy (6). Symptoms and clinical findings also include: (1) “Tullio” phenomenon representing vertigo in response to loud sound (3,6). (2) Conductive hearing loss: the third window dissipates energy and thus diminishes the amplitude of the pressure wave in the cochlea. The patient interprets this decrease in acoustic energy as a diminished sound volume (6). (3) Hennebert’s sign that represents nystagmus induced by pressure in the external auditory canal (7). (4) Audiogram shows a characteristic low to mid-frequency air-bone gap, due to shunt of perilymph through the semicircular canal away from the cochlea (4,7,8). (5) “Conductive hyperacusis” or “autophony”, attributed to elevated bone threshold, has enabled patients to hear the sound of their moving eyes (4,7,8).

Dehiscence of the lateral semicircular canal, on the other hand, may be acquired and has been described in association with cholesteatoma or fenestration procedure, made to treat otosclerosis (9). Pre-operative identification of this defect is critical, as cholesteatoma matrix is usually decided to be left during surgery as a protective cover, thus evading profound SNHL (10). It is essential for the radiologist to diagnose such lesions in both pre and post surgical settings especially in cases of recurrent manifestations and exaggerated hearing loss.

High resolution CT is the study of choice in radiological diagnosis of semicircular canal dehiscence. CT diagnosis of semicircular canal dehiscences depends on the demonstration of a tiny defect in the bony wall of the superior semicircular canal, in a patient with given clinical diagnosis by examination and/or audiogram.

The aim of this study was to assess the role of high resolution multi-detector CT in diagnosis of different dehiscences of the semicircular canals.

2. Materials and methods

This prospective study was conducted on 20 patients within one year interval (from June 2011 to June 2012). All the patients had given clinical diagnosis of dehiscences of the semicircular canals by clinical examination and/or audiogram. Beside conductive hearing loss, all the patients had clinical manifestations of inner ear pathology as sound induced vertigo (Tullio phenomenon), oscillopsia and/or disequilibrium. Hennebert’s sign were clinically diagnosed in 60% of cases. All the audiogram showed the characteristic low to mid-frequency air-bone gap.

Sixty-four multi-detector CT machine (Light speed, GE, USA) was used in assessment of all patients. Before imaging, the patient was informed about the investigation and instructed not to move during scanning. The patients were in supine position. The head was strapped to the head rest and positioned as symmetrically as possible. A lateral scout view was taken and used for planning the axial images. The scanning covered all components of the temporal bone. Axial images were taken without any angulations (Tilt 0). The protocol was 120 mA, 130 kV, 1.2 mm slice thickness and large field of view (FOV).

All images were prospectively reconstructed at 0.625 mm; using soft tissue and high-resolution bone filter. The reconstructed axial images were transferred to Advantage 4.4 GE, USA workstation for manipulation of data. Then multi-planar reformation (MPR) as well as minimum intensity projection (Min. IP) was generated. Additional reformations in the planes of longitudinal oblique (Stenver’s view) and transverse oblique (Pöschl view) were done but reserved for equivocal or confusing cases. All the medical ethics were respected.

In CT diagnosis of semicircular canal dehiscence, we depend on the demonstration of the tiny defect in the covering bony wall of semicircular canals. Even very thin sheet of intact covering bone excluded the diagnosis.

3. Results

Twenty patients were included in this study within a year period. Nineteen are adults (95% of patients); age ranges from 21 to 52 years. Only one pediatric patient (5% of patients) was a 12 year old girl.

We classified the patients into four groups: (Table 1)

1- First group: It included six cases of superior semicircular canal dehiscence, 4 of them had bilateral superior semicircular dehiscence. (Fig. 1) All of them were adults (27–50 years). Two cases had unilateral superior semicircular dehiscence. (Fig. 2)

2- Second group: It included three cases of posterior semicircular canal dehiscence. One case had bilateral involvement (Fig. 3) and two cases of unilateral involvement. (Fig. 4)

3- Third group: It included secondary canal dehiscence due to cholesteatoma. This includes 9 cases. 7 cases showed defect within the bony covering of the lateral semicircular canal, one case with defect within superior semicircular canal and remaining one with defect within both lateral and superior canals. (Figs. 5 and 6)

4- Fourth group: It includes cases with secondary canal destruction by mass lesions. This included two cases only: one case had left sided glomus jugulo-tympanicum, and the other case had left temporal bone deposit from parotid adenoid cystic carcinoma. (Fig. 7)

| Table 1 Distribution of patients in different groups. |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
| Groups                          | Number of cases | Percent | Number of cases | Percent |
| Congenital defects              | Superior semicircular canal dehiscence (first group) | 6      | 30%             | 9      | 45%             |
|                                | Posterior semicircular canal dehiscence (second group) | 3      | 15%             |
| Acquired defects                | Secondary canal dehiscence due to cholesteatoma (third group) | 9      | 45%             | 11    | 55%             |
|                                | Secondary canal destruction by mass lesions (fourth group) | 2      | 10%             |
| Total                          | 20               | 100%          | 20               | 100%          |
In our study the congenital defects of the SCC were encountered in 9 cases (45%), secondary acquired defects were encountered in 11 cases (55%).

Among the congenital defects, superior SCC was the most commonly involved, seen in 6 patients (30%). Posterior SCC defects were seen in only three cases (15%). Congenital defects of lateral SCC were not encountered in this study. Two thirds of patients with congenitally defective superior SCC had bilateral involvement (4 out of 6). On the other hand only one case of bilateral defective posterior SCC was diagnosed versus two cases of unilateral involvement (Table 2).

![Fig. 1](image1.jpg)

Fig. 1 A 31 year old male patient with clinical diagnosis of superior semicircular dehiscence syndrome. Right ear shows bony defect displayed by oblique reformation (double window view) (a). Coronal plane also shows the defect in (b). The left ear shows similar findings in (c and d).

![Fig. 2](image2.jpg)

Fig. 2 A 45 year old male patient referred for clinical suspicion of superior semicircular dehiscence syndrome. Coronal (a) and sagittal oblique reconstruction in (b) nicely displays the defect. This involves the dome and adjoining portion of the anterior limb.
Cholesteatoma was the most common single etiology causing defect within the semicircular canals. It was the primary responsible pathology in 9 cases (45%) among them, lateral semicircular canal suffered in 8 cases (40%). Superior semicircular canal suffered in only two (Table 3).

Malignancy was responsible for defects within the SCC in only 10% of total cases (2 patients).

4. Discussion

The prevalence of SSCD is not accurately estimated, however it is a rare condition, and the largest case-series published to date contains 65 cases seen over a 9-year period (2). Findings from post mortem temporal bone survey by Carey et al. reflect that approximately 2% of individuals already have a dehiscence or extremely thin bone overlying the superior semicircular canal. They suggested that the bilateral nature of thinning and the presence of mature lamellar bone on the margins of thin areas point toward a post natal developmental cause. However, as SCC syndrome first develops in adult, they also suggested a second event as closed head injury or intracranial pressure changes destabilize dura over a pre-existent dehiscence (3).

This developmental theory was also supported by Suryanarayanan, who described tegmen thinning and multiple defects.

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**Fig. 3** Female patient with bilateral posterior semicircular canal dehiscence. Axial CT scans show the defect on right and left side (a, and b respectively). Oblique reformations with minimum intensity projection reconstruction are also displayed for right and left ears in (c and d) respectively.

**Fig. 4** A 21 year old female patient complaining of right sided pulsatile tinnitus. CT showed dehiscence of the posterior semicircular canal on the right side, as shown in axial scan (a), and oblique reformation (b). No other abnormalities were detected in the left ear.
the so called “honey comb tegmen”) in three patients with superior SCC dehiscence (4). However this theory has been debated in the literature, Nadgir et al. tried to correlate increasing prevalence with age to systemic bony demineralization with age, or repetitive cumulative microtrauma, supporting the impression of acquired rather than developmental condition (5).

In their article, Belden et al. pushed their machines to the very limits of resolution in an effort to improve the accuracy of diagnosis, and avoids the partial volume averaging effect (11). This can be made with ultrathin 0.3 mm incremental reconstruction (12). With such technique, high-resolution temporal-bone CT scans can achieve a sensitivity of 100%, specificity of 99%, positive predictive value of 93%, and negative predictive value of 100% in the identification of superior SCC dehiscence (12). We used 64-multidetector CT scanner, with ultrathin reconstruction. We also pushed our machine to its maximum resolution. We agreed with others (11–18), in that MPR reconstruction was mostly sufficient for the evaluation of SCC dehiscences and additional reformations in the planes of Stenver and Pöschl views do not change the radiologic diagnosis and should be reserved for equivocal or controversial cases.

Our study included 6 cases of SCCD, 4 of them are bilateral, and all of them had nearly mirror image defects. Isolated dehiscence of the posterior semicircular canal was also reported, but less commonly occurring than the superior ones. Three out of our patient had defects of posterior semicircular canals: 1 bilateral and two unilateral.

Dehiscence of the lateral semicircular canal has only been described in association with cholesteatoma or fenestration procedure for otosclerosis (9). Pre-operative identification of this defect is critical, as cholesteatoma matrix is usually decided to be left during surgery as a protective cover, thus evading profound SNHL (10). Semicircular canal destruction was also reported in case reports of multiple destructive lesions of the temporal bone including jugular fossa, external auditory canal and petrous apex mass lesions (19).

Differential diagnosis for SCD includes other entities of CHL in the presence of a normal-appearing tympanic membrane. This includes two types of pathologies, middle and inner-ear CHL pathologies. Middle ear pathologies include ossicular discontinuity, malleus head fixation, tympanosclerosis, Paget’s disease, and osteogenesis imperfecta. Inner-ear conductive hearing loss pathologies present with CHL in the absence of any abnormality of the middle-ear conductive mechanism with preserved acoustic reflexes and normal tympanometry. These include enlarged vestibular aqueduct syndrome, and other inner-ear malformations (of which the X-linked stapes gusher syndrome is most important) (8).

To be mentioned, Otosclerosis, also leads to air-bone gap, with intact ear drum. Intact stapedial reflex can be a differentiating point, being lost in otosclerosis and intact in dehiscence of semicircular canal (8).
Fig. 6 A 30 year old male patient, with cholesteatoma of mastoidectomy bowel. Coronal CT scan showed opacification with labyrinthine fistula at lateral semicircular canal in (a). This is shown also in axial plane in (b). Associated tympanogenic labyrinthitis ossificans is seen as bony encroachment of the basal turn of cochlea as shown in (c, arrow). Co-existing bony defect of tympanic segment of facial nerve canal is marked by arrow in (d).

Fig. 7 A 52 year old male patient with history of resected adenoid cystic carcinoma of the left parotid gland. Axial CT scan in (a) shows ill defined small recurrence behind the mandibular ramus (arrow). Bone window in (b) shows residual lytic destruction of the mandibular ramus by tumoral invasion. Axial Scans in (c) shows temporal bone deposit with enhancing soft tissue components (arrows). Bone window in (d) shows lytic destruction at the site of metastatic deposits, and lytic destruction of the otic capsule at the lateral wall of the lateral semicircular semicircular canal.
5. Conclusion

Radiological diagnosis of semicircular dehiscences depends on the use of high resolution MDCT. Ultrathin reconstruction with MPR reconstruction is mostly sufficient in accurate display of the bony defects. Primary congenital dehiscences do occur in the superior semicircular canal and less commonly within the posterior canal. In this study, lateral canal dehiscence was only acquired. Secondary semicircular canal dehiscences are mostly caused by cholesteatoma but could be also caused by temporal bony aggressive mass lesions.

References

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