

An Uncommon Case of Bilateral Posterior and Superior Semicircular Canal Dehiscence Syndrome

RISHIKANT SINHA¹, UPASNA SINHA², PREM KUMAR³, SUBHASH KUMAR⁴, S APARNA⁵

ABSTRACT

Semicircular Canal Dehiscences (SCD) are rare and involvement of Posterior Semicircular Canal (PSCC) and/or multiple dehiscences are even rarer. A dehiscence of any of the semicircular canals may produce various auditory or vestibular symptoms by creating an abnormal communication between the inner ear and nearby structures. We report a rare case of multiple dehiscence with intracranially protruding PSCC and Superior Semicircular Canal (SSCC) beyond the margins of temporal bone with bony roof defects in bilateral PSCCs and SSCCs in a patient with chief complaints of positional vertigo.

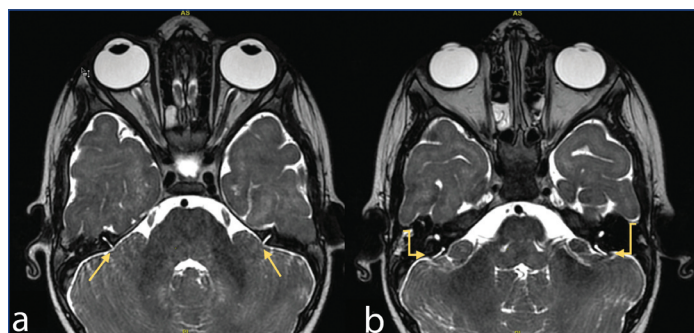
Keywords: High resolution temporal bone, Lateral semicircular canal dehiscence, Magnetic resonance imaging

CASE REPORT

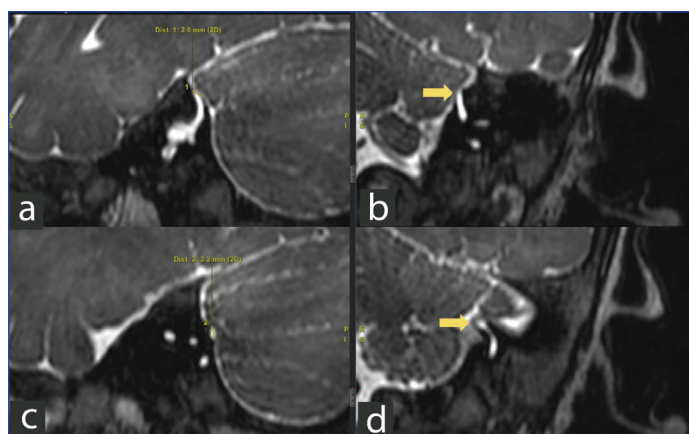
A 19-month-old male child presented to Department of ENT with chief complaints of unable to walk independently, decreased hearing and poor speech since six months. There was history of full term normal vaginal delivery with delayed immediate cry and neonatal jaundice in first 6 months. On examination vitals were stable. Local examinations (Pallor, icterus, cyanosis, clubbing, generalised lymphadenopathy, bilateral pitting pedal oedema) and systemic examinations including cardiovascular, respiratory, per abdominal and gross central nervous system examination were normal. Positional vertigo and positive hennebert sign were seen.

Otoscopic examination of bilateral external auditory canal showed normal tympanic membrane. Brainstem Evoked Response Audiometry (BERA) showed severe to profound hearing loss. Intelligence Quotient (IQ) assessment showed borderline level of intelligence (score of 70).

Patient was advised High-Resolution Computed Tomography (HRCT), MRI, TORCH (Toxoplasma, rubella, cytomegalovirus, herpes simplex) test, echocardiography (ECHO) and counselled for Cochlear Implant (CI). Weaning with trial and speech therapy was also advised. TORCH tests was negative. ECHO was normal. Plain MRI with inner ear protocol showed dehiscent bilateral PSCCs and SSCCs with their extra-axial protrusion along the Cerebro Spinal Fluid (CSF) space. Bony roof defect was seen in the postero-superior aspect of both posterior and superior SCCs, measuring 2.0 mm at left SSCC and 2.2 mm at left PSCC. Defect of 1.8 mm at right SSCC and 2.0 mm at right PSCC was seen [Table/Fig-1,2].



[Table/Fig-1]: Axial FIESTA at the level of superior semicircular canal: (a) and posterior semicircular canal; (b) shows protrusion of SSCCs above the horizontal plane of temporal bone on both sides with bony roof defect as absence of T2 hypointense line between CSF and membranous labyrinth (straight arrows). Protrusion of PSCCs behind the oblique temporal plate of posterior fossa with bony roof defect (elbow arrows).



[Table/Fig-2]: FIESTA sequence at the level of posterior limb of left SSCCs, sagittal (a); coronal (b); and at the level of left PSCCs, sagittal (c); and coronal (d) shows defect in the bony wall (thick straight arrows) with their defect size of 2 mm at left SSCC and 2.2 mm at left PSCC.

HRCT confirmed the presence and size of the bony defects [Table/Fig-3]. Patient underwent surgical plugging of the bony defects via



[Table/Fig-3]: HRCT temporal bone at the level of superior SCC, axial (a); and at the level of posterior SSCC, coronal (b); confirms the focal bony wall defect in bilateral posterior and superior semicircular canal with mild intracranial protrusion (solid yellow arrows).

transmastoid approach. The surgery was uneventful. There was alleviation of symptoms within one week of surgery. The patient was advised to follow-up monthly in Department of ENT.

DISCUSSION

SCD is a rare condition that was first described in 1998. Dehiscence involving lateral and posterior semicircular canal or multiple dehiscence are even rarer [1,2]. The dehiscence acts as a "third mobile window" in the bone that causes abnormal communication between the inner ear and subadjacent structures and results in multiple auditory or vestibular symptoms [1].

A study by Stimmer H et al., showed incidence of SCD to be 9.6% in which SSCD was 8%, PSCD was 1.2%, and 0.4% had LSCD [3]. A study by Russo JE et al., showed prevalence of SSCD to be 4.9% and that of PSCD was 1.2% [4]. SSCD occurs more commonly than posterior or lateral SSD [4]. It was hypothesised by Chen WW et al., that the SSCD and PSCD are more common as they are close to dura and are more susceptible to pressure effects, but they didn't explain the prevalence difference of SSCD and PSCD [1]. Some literature have hypothesised that the weight of the brain on the SSCD in the upright position contributes to the increased incidence of SSCD. Most commonly they are idiopathic, other aetiopathogenesis are due to congenital hypoplastic overlying bone, trauma, increased pressure during Valsalva manoeuvre and vascular pulsations or infections causing bony erosions [1,3]. High-riding jugular bulb and fibrous dysplasia are sometimes associated with PSCDs. LSCDs are usually associated with chronic otitis media or cholesteatoma [1].

The pathophysiology is disturbance in the pressure transmission in the labyrinth. The formation of the third mobile window causes the canal to respond to sound and pressure stimuli from outside resulting in decreased cochlear impedance [5].

The presenting symptoms of canal dehiscence are both acoustic and vestibular and include aural fullness, autophony, tinnitus, pulsatile tinnitus, disequilibrium, vertigo, tullio and hennebert phenomenon [1] with aural fullness and autophony being the most common presentation in PSCD [4]. Also, these cases are diagnosed incidentally in asymptomatic patients at HRCT study and surgeries of temporal bone. Our case presented with positional vertigo and showed positive hennebert sign.

An air-bone gap due to increased bone and decreased air conduction is seen in audiometric evaluation, more below 1 kHz of frequency. Vestibular evoked myogenic potential testing is abnormal on the side of pathology and shows that due to reduced impedance, there is increased transmission of acoustic energy at the sacculle.

Other middle ear pathology also produces air-bone gap, tympanometry and acoustic reflexes which are used to rule out middle ear abnormality [1,6].

HRCT temporal bone is the first line imaging modality of choice [7]. Half millimetre cuts have 100% sensitivity and 99% specificity with a Positive Predictive Value (PPV) of 93% and a Negative Predictive Value (NPV) of 100% [7]. Bony roof defects and protrusions of SSCDs are best visualised in Multiplanar Reconstruction (MPR) images reformatted parallel to Pöschl plane and perpendicular to Stenvers plane [1,8]. In present case, HRCT temporal bone was done with slice thickness 0.625 mm, slice increment of 3 mm and 0.625 mm collimation, as it

avoids volume averaging that can mask small bony defects. The key diagnostic features on CT imaging are defect in the bony covering of the semicircular canals [8]. Bony protrusion with or without defect beyond the temporal bone margin, in middle cranial fossa or posterior fossa is also delineated on HRCT with very high sensitivity.

On MRI, sequences such as Fast imaging employing steady-state acquisition (FIESTA)/Constructive Interference in Steady State (CISS)/ True FISP are used to evaluate the inner ear and internal auditory canal abnormalities as they provide high contrast definition between fluid and bone, produce excellent anatomic definition of the membranous labyrinth as they have high signal to noise ratio and MPR imaging capabilities [9]. MRI shows characteristic absence of bony hypointense layer between fluid in the semicircular canal and extra-axial CSF, indicating absence of bony coverage, or dehiscence. A study showed sensitivity of 100% in the FIESTA sequence compared with CT for detecting bony defects. However, they overestimated the presence of a dehiscence resulting in more false positive results and low PPV [10].

Surgical resurfacing, plugging, or capping of the dehiscence by either transmastoid or craniotomy approach have positive outcome in alleviating the symptoms [8]. Present case underwent successful surgical capping of the bony defect with resolution of the symptoms.

CONCLUSION

SCDs are uncommon occurrences with multiple dehiscences which are even more rare. They cause both vestibular (vertigo, tullio phenomenon, and hennebert sign) and auditory symptoms (autophony and inner ear conductive hearing loss). CT, Valsalva manoeuvre, Vestibular Evoked Myogenic Potentials (VEMP), and auditory testings are useful in diagnosis of a suspected dehiscence. HRCT is the modality of choice. MRI is 100% sensitive. Surgical treatment by various craniotomy approach have positive outcome in alleviating the symptoms. However, true understanding of aetiopathogenesis is essential for ideal management for canal dehiscence.

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PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Radiodiagnosis, All India Institute of Medical Sciences, Patna, Bihar, India.
2. Assistant Professor, Department of Radiodiagnosis, All India Institute of Medical Sciences, Patna, Bihar, India.
3. Professor and Head, Department of Radiodiagnosis, All India Institute of Medical Sciences, Patna, Bihar, India.
4. Associate Professor, Department of Radiodiagnosis, All India Institute of Medical Sciences, Patna, Bihar, India.
5. Academic Junior Resident, Department of Radiodiagnosis, All India Institute of Medical Sciences, Patna, Bihar, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Rishikant Sinha,
Department of Radiodiagnosis, Patna-801507, Bihar, India.
E-mail: drrishikantsinha@gmail.com

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