

Case Report

Superior Semicircular Canal Dehiscence: Demonstration with Three-Dimensional Multidetector Ct and Mr Imaging

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Keywords

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Abstract

Introduction: Semicircular canal dehiscence (SCD) syndrome has been defined as a bone defect in the roof of the superior, posterior and lateral semicircular canals. We reported that three-dimension CT and MR imaging of superior SCD is rare in english literature.

Case presentation: A 6-year-old boy patient presented with hearing loss and left facial nerve paresis since birth. CT and MR images demonstrated superior SCD causing congenital hearing loss.

Discussion: Multidetector CT with multiplanar reconstruction and three-dimension volume rendered imaging is an excellent imaging method to detect this defect. MR imaging should be preferred in the demonstration of the cranial nerves and relationship of SCD with the epidural space in more detail. In conclusion, superior SCD should be considered for differential diagnoses in patients presenting with congenital hearing loss.

CASE PRESENTATION

A 6-year-old boy patient presented with hearing loss and left facial nerve paresis since birth. There was no family history. The vestibular sign was not detected. When baby was 2 months, he had been operated because of the left mastoid bone mass which was pathologically proven to be a benign fibrous histiocytoma. Audiometry indicated moderate to severe mixed type hearing loss in the left ear. Electroneuromyography showed findings associated with chronic neuropathy of 7. Cranial nerve which is cause of facial palsy. The patient ongoing hearing loss after the simple mastoid bone operation was evaluated with a 4-detector-row computed tomography (CT) scanner (Aquilion; Toshiba Medical Systems, Tokyo, Japan). CT images revealed the superior semicircular canal dehiscence (SCD) (Figure 1) and postoperative changes of mastoid bone (Figure 2) on the left side. Three-dimensional (3D) volume rendering (VR) images demonstrated superior SCD in more detail (Figure 3). Magnetic resonance (MR, 1.5 Tesla, Toshiba Medical Systems, Tokyo, Japan) imaging was done to evaluate 7. and 8. cranial nerves.

These cranial nerves were normal. However, left superior SCD neighboring to the epidural space was detected in coronal T2-weighted turbo spin echo images (Figure 4). The diagnosis of superior SCD was corrected with operation and learned that improvement of hearing loss in the one-year follow-up.

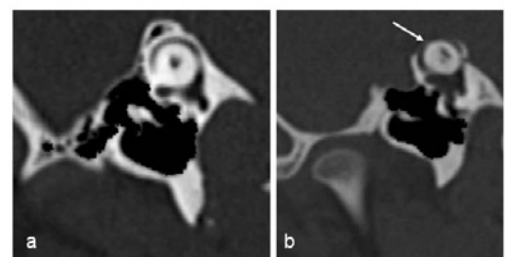


Figure 1 Sagittal oblique CT scans of the temporal bones show normal bone roof on the right side (a), and dehiscence (arrow) of the superior semicircular canal on the left side (b).

Semicircular canal dehiscence syndrome has been defined as a bone defect in the roof of the superior, posterior and lateral semicircular canals [1,2]. It was firstly described by Minor et al. [3] and is a cause of vertigo triggered by sound or pressure changes in the middle ear cavity or in the cranium [4]. Clinical manifestations of SCD is characterized by vestibular and auditory findings such as vertigo, rotatory and vertical nystagmus, tinnitus and hearing loss [2,5]. The etiology of SCD is unknown. Carey et al. [6] suggest that dehiscence arises from a defect in postnatal development. Nadgir et al. [1] study showed that superior SCD is more commonly an acquired rather than congenital or developmental condition. They thought that the increasing prevalence of superior SCD with age may be related to systemic bony demineralization increased with age. It may be also arise from repetitive microtrauma and surgery. Generally, the incidence of SCD varies in different series. Stimmer et al. [2] reported SCD in 9.6% of temporal bones. They found superior, posterior and lateral SCD in 8%, 1.2% and 0.4% of temporal bones, respectively. Mondina et al. [7] reported as 8% and Cloutier et al. [8] found as 4% on temporal bone CT. The diagnosis of SCD is rarely made together with clinical and audiometric findings. Radiographic imaging of the temporal bones with CT plays a significant role in the diagnosis of these patients [1,5]. CT images can also be used to evaluate the extent of the dehiscence area, which is a criterion for operability. Dehiscences of greater than 3 mm are considered as operable [9].

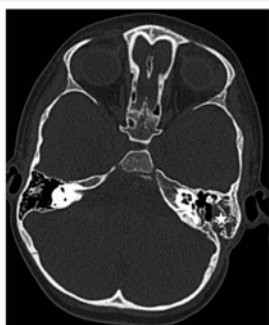


Figure 2 Axial CT scan of the temporal bones shows postoperative changes of the mastoid bone (star) on the left side.

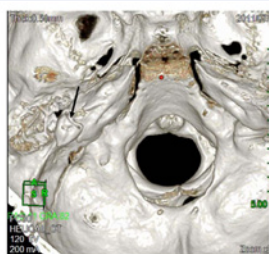


Figure 3 3D volume rendering image shows the superior semicircular canal dehiscence (arrow) in more detail.

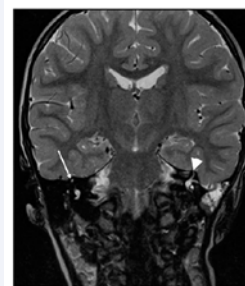


Figure 4 Coronal T2-weighted turbo spin echo image shows normal bone roof (arrow) on the right side and superior semicircular canal dehiscence neighboring to the epidural space (arrowhead) on the left side.

To our knowledge, congenital hearing loss has not been previously reported as associated with superior SCD. In conclusion, superior SCD should be considered for differential diagnoses in patients presenting with congenital hearing loss. Knowledge of the presence and location of SCD is highly important in preoperative surgical planning. Multidetector CT with multiplanar reconstruction and 3D VR imaging is an excellent imaging method to detect this defect. MR imaging should be preferred in the demonstration of the cranial nerves and relationship of SCD with the epidural space in more detail.

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