Superior Semicircular Canal Dehiscence Syndrome in a Woman: A Case Report

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Abstract

Background: Superior semicircular canal dehiscence (SSCD) syndrome is a disease that occurs very rare. Clinical presentation consists of Tullio's phenomenon and Hennebert's sign. This case presentation is atypical and clinician should be aware. **Purpose**: to present the rare case of Superior semicircular canal dehisence (SSCD) **Case presentation:** A 65-year-old woman with complaints of dizziness from 2 years ago and her recurrence. The audiogram showed bilateral sensorineural deafness with a mean hearing threshold of 58 decibels (dB) of the right ear and 48 dB of the left ear. There is also an air-bone gap in the right ear at low frequencies (250 Hz). The tympanogram revealed a type A result in both ears. Positive results were obtained on the examination of Romberg sharpened, positive Fukuda step to the right, and the Gans test. CT scan showed dehiscence of the right superior semicircular canal. Therapy in patients was given conservative therapy because the participants refused to undergo surgery. **Conclusion:** SSCD has a specific symptom of Tullio's phenomenon and CT scan shows dehiscence of the superior semicircular canal.

Keyword: *Superior semicircular canal dehiscence, Tullio's phenomenon, vertigo*

Introduction

Superior semicircular canal dehiscence (SSCD) syndrome was a disorder in which symptoms of vertigo induced by loud noises (Tullio's phenomenon) or intracranial pressure (Hennebert's sign) due to dehiscence of the bone lining the superior semicircular canal[(1)]. The prevalence of developing SSCD was 3% on the computed Tomography (CT) scan survey of the temporal bone[(2)]. Tullio's phenomenon and Hennebert's sign are characteristic sign because of the formation of a third window in the superior semicircular canal. The third window appears due to dehiscence of the superior semicircular canal. Sound stimulation and intracranial pressure on this third window cause excitation or inhibition of the superior semicircular canal leads to symptoms of vertigo. Bone conduction will lead to hyperacusis, leading to symptoms of autophonia[(3)].

Temporal CT scan of SSCD can show dehiscence of the semicircular canal. The CT scan is about 0.5 mm thick to see the structure of the inner ear. Clinical symptoms combined with appropriate CT scan images can increase SSCD specificity by 99%[(4)]. Audiometric examination of SSCD can give results of conduction hearing loss, especially low frequencies and tympanometry examination yields normal values[(5)]. Based on the description above, we are interested in reporting a case of SSCD that we had in a 65 year old woman.

Case Presentation

A 65-year-old woman with complaints of vertigo since 2 years ago has disappeared. Vertigo is not triggered

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by change in head position. The patient has undergone medical rehabilitation therapy but the complaints persist and when she hear loud sounds feel uncomfortable and dizzy. Hearing loss since 1 year ago in both ears. Previous medical history is hypercholesterolemia. The patient has seen several doctors and finally to a neurosurgeon, then a Magnetic Resonance Imaging (MRI) examination is recommended before being referred to a specialist in otorhinolaryngology head and neck surgery.

On both ears Rinne's examination and Weber's examination were no lateralization and Bing's examination was positive in both ears. The audiogram showed bilateral sensorineural deafness with hearing threshold pure tone average of 58 decibels (dB) of the right ear and 48 dB of the left ear. There is also an air-bone gap in the right ear at low frequencies (250 Hz). The tympanogram revealed a type A result in both ears. The acoustic reflex was not continued because the patient experience vertigo during examination. When the acoustic reflex was performed, nystagmus appeared but the direction could not be determined because the patient asked to stop the examination. The valsalva test was not performed in these patients because of vertigo.

Vestibular examination was carried out including the Romberg test, the Romberg sharpened test, the Fukuda step test, the Gans test, and the Dix Hallpike test. Positive results were obtained on the examination of Romberg sharpened, positive Fukuda step to the right, and the Gans test. Negative results were obtained on the Romberg, Dix-Hallpike examination and the coordination examination in the form of dysdiadokinesia and finger nose tests. In conclusion, the results of the vestibular examination showed that Vertigo with non-Benign Paroxysmal Positional Vertigo (BPPV) type.

A CT scan of the temporal bone with suspicion of SSCD was performed on the patient. CT scan showed dehiscence of the right superior semicircular canal. The left semicircular canal appeared normal. The cochlea and vestibule appeared normal. Bilateral normal internal acoustic canal. Both patent of vestibular aqueduct and facial nerve. Osicles and scutum were normal, no erosion found. The foramen ovale and rotundum were normal bilaterally (Figures 1 & 2). Therapy in patients was given conservative therapy because the participants refused to undergo surgery.

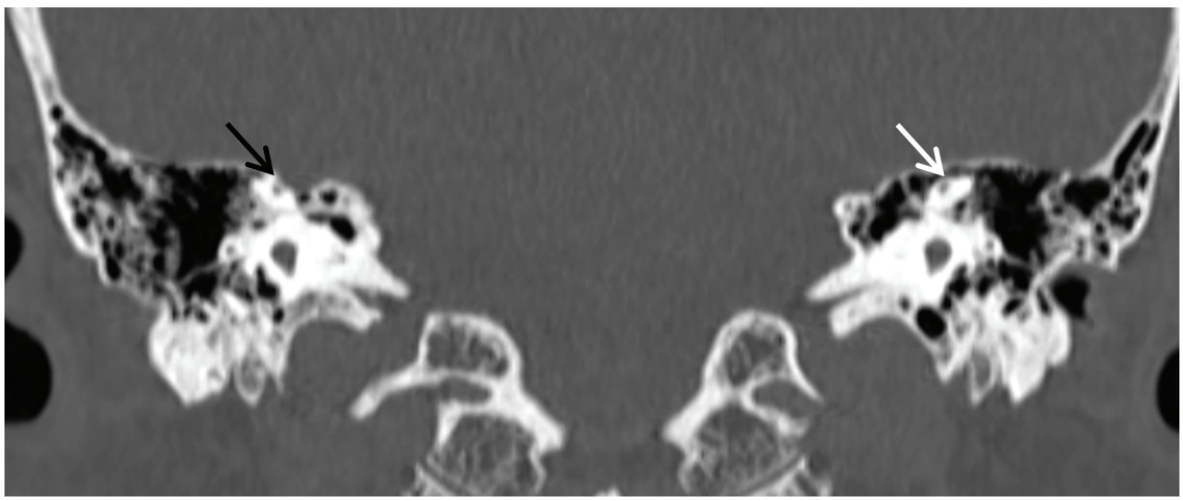

Figure 1. CT scan of the coronal temporal cut shows dehiscence of the right superior semicircular canal (black arrow). Left superior canal appears normal (white arrow).

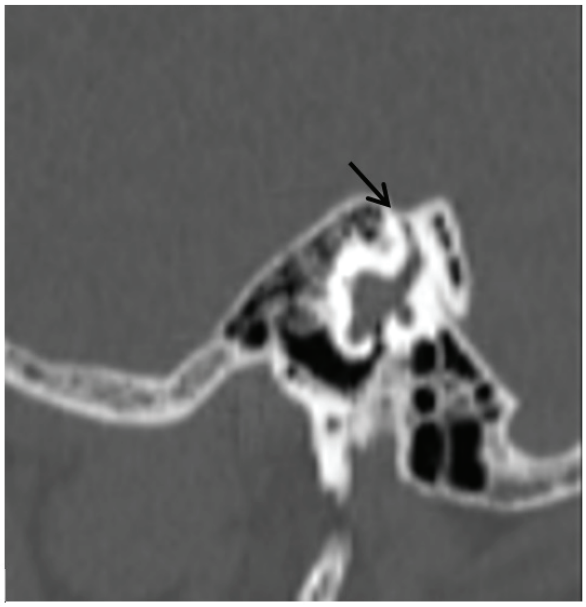

Figure 2. CT scan of the right mastoid showed dehiscence of the right superior semicircular canal (black arrow).

Discussion

SSCD syndrome was a disorder due to dehiscence of the roof of the superior semicircular canal occurs. Symptoms that arise involve hearing and balance problems. In general, the symptoms that arise are divided into two, related to sound and pressure changes. Symptoms associated with sound are vertigo triggered by loud noises (Tullio's phenomenon), autophonia (can hear sounds from one's own body), hyperacusis, fullness in the ears, tinnitus, low frequency conduction deafness, while symptoms related to pressure changes are vertigo when the intracranial pressure increases (Hennebert's sign) eg when coughing, valsalva, and lifting heavy objects[5]. The age of SSCD patients in this case is in accordance with previous studies where many SSCD patients were found to be over 60 years of age[2] and other studies stated that the incidence of SSCD increases with increasing age[6]. In this case there was dehiscence of the superior semicircular canal in the right ear. Dehiscence often occurs unilaterally but can also be obtained bilaterally[7].

The Tullio phenomenon can produce vertigo when hearing loud sounds especially at low frequencies[8]. The Tullio phenomenon was found in 96% of these SSCD sufferers. The symptom description is characterized by vertigo or nystagmus on hearing a loud sound[7]. The symptoms of Tullio's phenomenon are not given enough attention as a result of the patients are misdiagnosed with psychiatric disorders. Vertigo in SSCD can also result from the Valsalva maneuver[9].

Low frequency (250 Hz) air bone gap in the right ear corresponds to hearing loss in SSCD usually conduction hearing loss at low frequencies below 1000 Hz[3]. This conduction hearing loss results from the presence of a third window of the superior semicircular canal. Sound energy that enters the cochlea through the foramen ovale will partially go to the dehiscent semicircular canal, resulting in an increase in the threshold with air conduction. This results in an air-bone gap without pathology in the middle ear[10]. Electronistagmography (ENG) can help diagnose SSCD where nystagmus occurs when tympanometry and valsalva are performed. A 100-110 dB stimulation examination at 125-4000 Hz will show vertical and torsional nystagmus. Fistula examination revealed vertical and torsional nystagmus with a slow phase moving away from the side of lesion, but valsalva examination revealed nystagmus in the opposite direction[11].

The results of temporal CT scan in this patient showed dehiscence of the right superior semicircular canal. Temporal CT scan is the main modality in diagnosing SSCD, especially if the patient is planning for surgery. Normal view of the superior semicicular canal has 3 layers of roof, namely the otic capsule, trabecular bone and bone cortex. These three layers can be clearly seen on a temporal CT scan with 0.5 mm thickness. The recommended CT scan is multi-axial with a parallel image of the coronal, sagittal and axial slices with a thickness of 0.5 mm. Superioriy the picture of dehiscence when viewed from the coronal slice. Temporal reconstruction CT scan with multiple slices can increase the sensitivity and specificity in detecting the presence of SSCD. Dehiscence that appears on one slice can not confirm the presence of dehiscence in other slices[4].

SSCD therapy can be both conservative and surgical. Conservative is aimed primarily at SSCD patients with mild symptoms. Conservative therapy by avoiding increased intracranial pressure such as straining, blowing hard, air travel, lifting heavy weights, or bending over. These activities can worse the vertigo. Surgical therapy has had high success in reducing vestibular symptoms and autophonia. Hearing symptoms rarely improve after surgical therapy[12].

Surgical therapy may involve window reinforcement, resurfacing, and plugging / occlusion techniques. The reinforcement window has a high recurrence rate in the long term and is less effective in reducing symptoms of autophonia. This technique is usually combined with resurfacing and occlusion techniques. Resurfacing technique is to give flap over the dehiscence of the canal with a special material to prevent the transmission of intracranial pressure into the inner ear. The materials used can be bone, cartilage, fascia or ceramic implants. This technique can be effective in reducing the symptoms of vertigo and autophonia. The occlusion technique is clogging the canal thereby eliminating the function of the semicircular canal[12].

The operation can be performed by trans cranial medial fossa, transmastoid, and transcanal approaches. The cranial media fossa approach is a traditional technique with first craniotomy to find arcuate eminence and areas of dehiscence. The transmastoid approach is less invasive than craniotomy. This approach also allows for plugging and resurfacing through a mastoidectomy. The success rate for this approach is also quite high at 94%. The transcanal approach is the newest technique by strengthening (reinforcement) the foramen ovale or rotundum. This technique can reduce the risk of complications of facial nerve paralysis, cerebrospinal fluid leakage, and intracranial hemorrhage[13].

Conclusion

A 65 year old woman diagnosed with SSCD showed symptoms of the Tullio phenomenon, namely vertigo when hearing loud sounds, especially low frequencies. On audiogram examination, bilateral sensorineural deafness and air-bone gap of the right ear were found at low frequency (250 Hz). Temporal CT scan showed dehiscence of the right superior semicircular canal. Romberg's test results, Gans test and Fukuda's step test gave positive results to the right. Therapy in patients is carried out conservatively.

Conflict of Interest: The authors declare that they have no conflict of interest.

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Ethical Approval: All procedures performed in studies involving human participants were in accordance declaration of helsinki the Ethics Committee in Dr. Soetomo General Academic Hospital, Surabaya, Indonesia.

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