SUPERIOR SEMICIRCULAR CANAL DEHISCENCE WITH CHRONIC OTITIS MEDIA

KRONİK OTİTİS MEDİA İLE BİRLİKTE SÜPERİOR SEMİSİRKÜLER KANAL DEHİSANSI

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ABSTRACT

Superior semicircular canal dehiscence is rare syndrome combined of both auditory and vestibular symptoms. Superior semicircular canal dehiscence may co-exist with other diseases as chronic otitis media, which may complicate the symptoms. In this case a 58 year old female patient who applied to our clinic with severe vertigo, nausea and vomiting for 15 days, diagnosed with chronic otitis media and concomitant superior semicircular canal dehiscence on the same side will be discussed in terms of symptomatology and differential diagnosis.

Key words: *Hearing loss, nystagmus, superior semicircular canal, vertigo*

ÖZET

Süperior semisirküler kanal dehisansı işitsel ve vestibüler semptomların bir arada belirdiği nadir görülen bir sendromdur. Kronik otit gibi semptomların karışabileceği ek hastalıklarla birlikte gözlenebilir. Bu yazıda 15 gündür şiddetli baş dönmesi, bulantı, kusma şikayetleri ile kliniğimize başvuran kronik otitis medialı 58 yaşındaki bayan hastada aynı kulakta eşlik eden süperior semisirküler kanal dehisansı semptomatoloji ve ayırıcı tanı açısından tartışılmıştır.

Anahtar kelimeler: *İşitme kaybı, nistagmus, süperior semisirküler kanal, vertigo*

INTRODUCTION

Superior semicircular canal dehiscence (SSCD) was first described by Minor et al in 1998. It is known as a pathological 'third window' that leads to an altered hydro-acoustic transition through the cochlea and labyrinth. It is characterized with the absence of the bony fragment of the otic capsule that lays on the superior semicircular canal (1). SSCD is a syndrome that vertigo and oscillopsia occur in patients by loud sounds or pressure alterations in the external ear canal or middle ear (2). Symptoms as autophonia, hyperacusis, pulsatile tinnitus, conductive hearing loss and aural fullness may accompany as well (3).

The etiology of the disease still remains unknown. In one third of the affected patients SSCD may be bilateral(2). The incidence rate is reported as 0.7-9% in the current literature(4). In this case, a patient with concomitant chronic otitis and SSCD on the same side is discussed in terms of the symptoms, differential diagnosis and treatment modalities in the light of the current literature.

CASE

A 58 year old female patient applied to our clinic with severe vertigo, nausea and vomiting over the past 15 days. The patient also had mild hearing loss on the right ear for the past 2 years, however did not complain of otorrhea, tinnitus or head ache headache. The patient's history did not reveal any otological surgery or acoustic/baro trauma. She did not have any chronic diseases but migraine and hypertension which were well controlled with medication.

On physical examination nystagmus was not detected, facial nerve functions were evaluated as normal. Otoscopic and microscopic examinations revealed a 3 mm dry perforation localized on the postero-inferior portion of the right tympanic membrane and a normal intact left tympanic membrane. The fistula test was negative for both ears. Other otolaryngological examinations were normal.

The patient's hemogram and biochemical analysis were found normal. The audiometry revealed conductive hearing loss on the right, as the 4 frequency (500-1000-2000-4000 Hz) average pure tone airway threshold was 29 dB on the right and 10dB on the left. The bone conduction threshold was 13dB on the right and 8 dB on the left **(Figure 1).** Tympanometric analysis and acoustic reflexes were normal. There were no pathological findings on the cranial diffusion weight magnetic resonance imaging (MRI). High resolution computed tomography (HRCT) of the temporal bone revealed a dehiscence over the right

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superior semicircular canal wall. Additionally, it was observed that the mastoid aeration on the right was lost **(Figure 2).** The carotid artery doppler ultrasonography (USG) revealed a fibro fatty plaque over the anterior wall of the left internal carotid artery and at this level stenosis up to 50% was detected. Vertebral artery doppler USG was reported as normal.

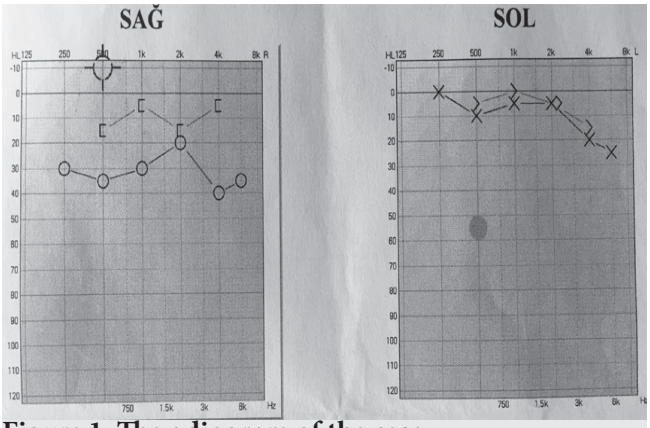

Figure 1: The odiogram of the case.

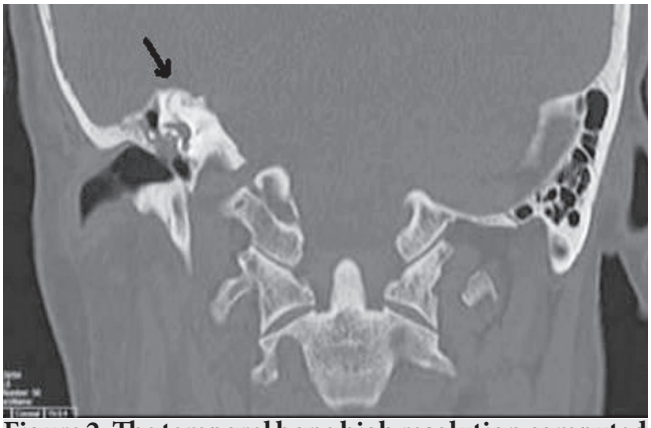

Figure 2: The temporal bone high resolution computed tomography section of the case. Black arrow: Right superior semicircular canal dehiscence.

The patient whose neurological examination was evaluated normal was hospitalized afterwards. Acetylsalicylic acid 1x300 mg, ceftriaxone 2x1g, dimenhyrdrinate 3x50mg, piracetam 4x3g, dexketoprofen 1x500mg and olmesartan medoxomil 1x20mg were administered daily. After a week of treatment, the patient was discharged as her complaints improved. She did not have any complaints at her first and third month follow-up and an elective tympanoplasty was planned. The patient gave consent for this paper.

DISCUSSION

Superior semicircular canal dehiscence is a dehiscence of the otic capsule localized over the apical part of the superior semicircular canal near the middle cranial fossa. It may be congenital however martial arts, repetitive diving, cranial traumas with low intensity may lead to acquired SSCD as well(2,5).

In the cochleovestibular system, there are two windows, one is the oval window that enables the sound waves transition to scala vestibuli through the base of stapes, the other is the round window which provides the transition of sound and mechanical energy out side the inner ear through scala tympani. This is a closed hydraulic system. In case of a dehiscence, the hydroacoustic waves passing through the cochlea are mistakenly transmitted through the labyrinth system. This leads to activation in the vestibular system and as a result vertigo is perceived. Additionally, the fluctuations of the intracranial pressure may also be transmitted through the dehiscent superior canal and consequently stimulate the vestibular end organs until the round window releases the pressure. Briefly, SSCD alters the systems hydrodynamic stability and leads to an exaggerated activation of the endolymph. The sound and pressure related vertigo in SSCD is explained by this mechanism(2,6,7).

The increased compliance of the inner ear plays a role in the conductive hearing loss and pulsatile tinnitus which may be present in these patients. The pathological gap due to SSCD may diminish the energy transmission derived from the movement of the base of stapes thus the sound transmission to the cochlea may decrease and eventually lead to hearing loss. On the other hand the normal difference between the oval and round window impedance may be altered because of the third window which results in bone hyper conductivity that is perceived as autophonia or hyperacusis by patients. This mechanism explains why the bone threshold values are obtained less than zero in the audiometry(2,8).

In SSCD syndrome the symptoms belong to auditory and vestibular systems. Auditory symptoms are autophonia, pulsatile tinnitus, bone hyper conductivity and aural fullness. The vestibular symptoms, which are triggered by sound (Tullio Phenomenon) and pressure (Hennebert Phenomenon), are; the characteristic torsional nystagmus, which is upwards and opposite to the affected ear, oscillopsia and vertigo(5,9).

The clinical symptoms are not enough for the diagnosis of SSCD sydrome(5). Apart from the clinical presentation, a HRCT of the temporal bone is necessary to confirm the diagnosis(4). In the cervical vestibular evoked miyogenic potential (cVEMP) test, although thresholds can be obtained, frequently lowered responses are received(10).

In this case, the absence of the characteristic symptoms of SSCD and the conductive hearing loss due to the concomitant chronic otitis media, did not suggest SSCD in the first place.

Having no discharge from the ear and because of the lack of supportive findings in otoscopic and microscopic ear examination and temporal bone computed tomography; the possible diagnosis of complicated chronic otitis

media was ruled out. The temporal bone HRCT revealed a dehiscence over the bony canal of the superior semicircular canal near the middle cranial fossa.

Most of the patients diagnosed with SSCD syndrome do not require surgical intervention. During attacks, avoiding provocative factors and inhibition of the vestibular system with medication are usually efficent(5). Operative intervention is indicated for persistent or debilitating symptoms despite conservative treatment and vestibular sedation(9).

In patients with chronic otitis media; symptoms as vertigo, nausea, vomiting and hearing loss are primarily thought to be findings of the present disease or its complications. In chronic otitis media, it is quite rare that these findings occur due to the underlying SSCD. When the symptoms and the examination findings are not correlated, the temporal bone HRCT must be carefully evaluated as other problems may be present concurrently. In this case the temporal bone HRCT revealed a dehiscence over the right superior semicircular canal and the symptom-disease correlation was established correctly. The complaints of the patient improved with medical treatment hence surgical intervention was not considered.

CONCLUSION

SSCD is a rare syndrome that both auditory and vestibular symptoms are present. SSCD may co exist with other diseases that can complicate the clinical picture as chronic otitis media. Differential diagnosis is crucial in determining the underlying disease of the related symptoms and treatment modality. A good analysis of the clinical symptoms and detailed examination combined with a temporal bone HRCT is important in making the differential diagnosis. Informed consent was obtained from the presented case.

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