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Minor's Syndrome or Dehiscence of the Superior Semicircular Canal: A Case Report

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Abstract **Case Report**

Semicircular canal dehiscence is a congenital syndrome that mainly affects the superior and, less commonly, the posterior semicircular canal in the temporal bone. This syndrome was first reported by Minor et al. in 1998. The prevalence is about 0.5% in the general population. The most characteristic symptom is the Tullio phenomenon, predominantly vertigo and nystagmus. Hearing loss (in particular conductive loss) often completes overall clinical picture. The dehiscence may be completely asymptomatic, in which case it is seen as an incidental finding on radiological investigations. Audiological evaluation includes audiometry and vestibular evoked myogenic potentials (VEMPs). Clinical symptoms are based on the pathophysiological concept of a third mobile window within the inner ear. Diagnosis is confirmed by high resolution petrous temporal bone CT with multiplanar reconstructions (MPRs) in the plane of the canal (Pöschls' plane for the superior semicircular canal) and 3-D surface reconstructions. MR findings are less sensitive for this entity. The most common differential diagnosis is otosclerosis. Clinical–radiological correlations have therapeutic applications even though the indication for surgery depends on the severity of the vestibular symptoms.

Keywords: Semicircular canal; dehiscence; congenital syndrome; CT..

INTRODUCTION

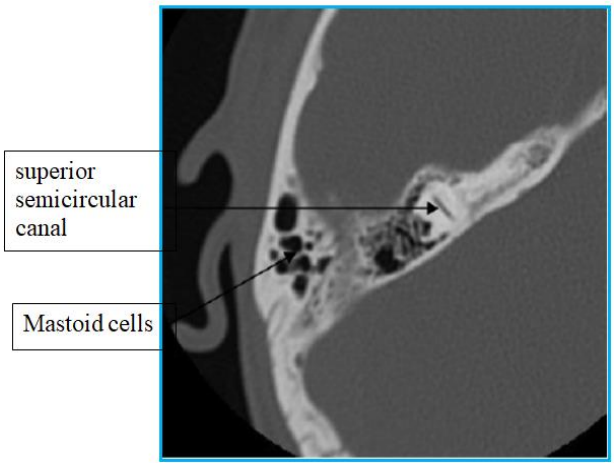
Dehiscence of the semicircular canals (CSC) is a rare entity. It is a malformation consisting of an absence of the bony cortex of the upper CSC, also known as Minor's syndrome.

Clinically, it presents as mixed or conductive hearing loss with normal eardrum, usually accompanied by vertiginous phenomena of varying severity. Tullio phenomenon may also be present.

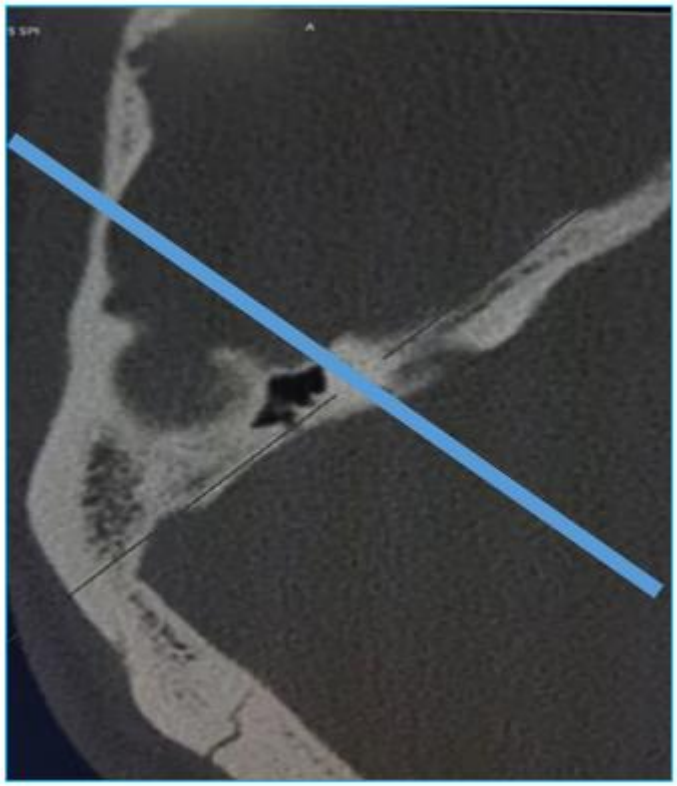
OBSERVATION

This is a 46-year-old man with right conductive hearing loss and left sensorineural hearing loss, with normal eardrum on both sides.

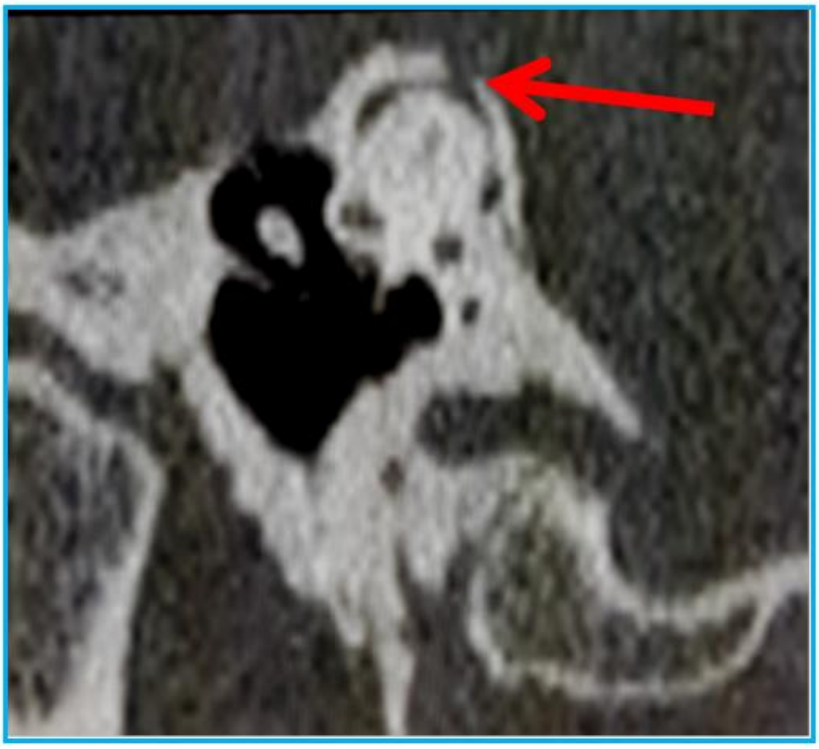
A CT scan was performed in millimetric slices with multiplanar reconstruction, including the Poschl plane (plane of the CSS), which revealed a dehiscence of the right CSS measuring 2.8mm. The tympanic fundus and inner ear elements were without abnormality. The left rock was normal.



CT scan: Axial sections through the superior semicircular canal



Poschl plan = CSCS plan (blue line)



CT scan: Poschl section showing dehiscence of the CSCS. (Red arrow)

DISCUSSION

Semicircular canal (SCC) dehiscence is a congenital syndrome primarily affecting the superior SCC and, less frequently, the posterior SCC within the temporal bone.

It is a recently discovered pathology (1998), described by Minor *et al*., Its prevalence is estimated at 0.5% in the general population. The average age is 50, with no gender predominance.

Clinical features:

The most characteristic symptom is the Tullio phenomenon: the patient presents with repeated vertigo to pressure variations (coughing, sneezing, flying, diving. . .) and/or nystagmus induced by loud sounds. Conductive deafness often completes the picture.

ENT evaluation includes audiometry, most often conductive deafness with a Weber test lateralized on the affected side, and a negative Rinne test.

Analysis of myogenic vestibular evoked potentials (MVEP) reveals a marked decrease in threshold compared with the healthy side, as well as an increase in amplitude.

This is an essential paraclinical criterion for diagnosis, making all the difference between Minor's syndrome and radiological dehiscence.

Differential diagnosis:

At the clinical stage, it's otosclerosis. However, stapedial reflexes are preserved in superior CSC dehiscence, which is not the case in otosclerosis.

Imaging:

Multi-bar helical CT is of great interest in diagnosing this syndrome, as it enables us to analyze the lack of bony coverage of the superior CSC in all spatial planes, but especially in the sagittal plane, obliquely in the plane of the superior CSC or Pöschl's plane. This dehiscence is most often wide, with slightly blurred contours, corresponding to an isolated bone coverage defect.

The dehiscence may be uni- or bilateral. The average bilateral rate in the literature is 25%.

Beware of very thin bone coverings (less than 0.5 mm), which are not visible even on high-resolution CT and thus mistaken for dehiscence.

Treatment:

Essentially medical and rehabilitative. Due to the complexity of the surgical procedure, it is only proposed when vestibular symptoms remain extremely incapacitating.

CONCLUSION

When faced with a symptomatology involving conductive deafness with normal eardrum or mixed deafness associated with vertigo, it is important to consider a CT search for dehiscence of the superior CSC, in particular by performing MPR reconstructions in the axis of the superior CSC (Pöschl plane).

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