Superior Semicircular Canal Dehiscence, Not Only Vestibular Symptoms.

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Learning objectives

To describe CT and MR findings of Superior Semicircular Canal Dehiscence (SSCD) associated to Tegmen Tympani Defect (TTD) and encephalocele in patients studied for conductive hearing loss.

Background

SSCD

Superior Semicircular Canal Dehiscence (SSCD) is described as the absence of the bony covering of the superior semicircular canal. This creates a virtual third mobile window, other than the oval and round existing ones, and the sound is transmitted to the vestibular system causing vertigo by excitation of the cupula creating dizziness and nystagmus. This third window also leads the sound away from the cochlea and hence causes conductive hearing loss in some individuals. It also may remain asymptomatic.

Superior canal dehiscence probably arises from failure of postnatal bone development. The usual bilateral nature of the thinning and the mature lamellar bone on the margins of the thin areas point towards a developmental cause(4). However other factors may intervene in the pathologic process as the clinical manifestations usually appear in middle aged patients. Some authors hypothesize that patients with this disorder fail to develop a normal thickness of bone overlying the superior canal and that this abnormally thin layer of bone may become disrupted by a traumatic event or by pressure from the overlying temporal lobe or dura (6) (8).

Clinical symptoms may be unsteadiness associated with loud noises and/or pressure changes. Tullio phenomenon defined as vertigo and nystagmus precipitated by loud noises and the Hennebert sign (vestibular symptoms with pressure changes) are classically described in this entity.

Associated features are hearing loss and chronic balance problems. Some patients have exclusively vestibular symptoms and signs; some have both auditory and vestibular manifestations; and still other patients have exclusively auditory complaints.

This condition is diagnosed not only on the basis of vestibular and auditory symptoms, but also on radiological findings such as absence of bone overlying the Superior Semicircular Canal.
Very symptomatic patients with balance alterations, unsteadiness or others incapacitating vestibular or auditory symptoms may benefit from surgical repair of the dehiscent canal through middle fossa or transmastoid approach.

TEGMEN TYMPANI DEFECT

The tegmen tympani is formed by the petrous and squamous portions of the temporal bone. There is a suture between these two portions called the petrosquamous suture, which is unossified in youth and ossifies in adult age. The tegmen tympani extends posteriorly to roof the tympanic antrum and anteriorly to cover the semicircular canals. Its lateral edge corresponds with the remnants of the petro-squamous suture.

The medial part of the tegmen tympani develops form the otic capsule during chondral ossification, and the lateral portion of the tegmen tympani develops by membranous ossification. Congenital defects of the tegmen tympani develop ventrally to the geniculate ganglion and may be due to incomplete ossification of the tegmental process of the otic capsule. Inadequate closure of the petrosquamous suture may be a factor as well.

TTD can lead to meningoencephalocele and/or cerebrospinal fluid (CSF) leak and consequently recurrent meningitis, convulsions, paresis, aphasia, among other possible complications. Meningoencephaloceles are herniations of brain tissue through dehiscences of skull base.

ASSOCIATION

SSCD can be congenital or acquired, unilateral or bilateral. Its pathophysiology nowadays is not well understood but several hypotheses exist, pointing out association between SSCD and TTD, suggesting they might share a common mechanism and even be part of the same syndrome. Some recent studies have observed that there is an association between these two entities concluding that the radiological absence of Tegmen Tympani is more frequent in ears that also have radiologic SSCD (2) (6).

Clinical symptoms may be conductive hearing loss and/or vertigo associated with Tullio phenomenon or Hennebert sign. But, considering this association, there are also other symptoms and findings of extremely importance such as meningoencephaloceles or CSF leaks.

Radiologists must be aware of these possible findings when performing a conductive hearing loss study, and if SSCD is found, look further at TTD.

Findings and procedure details
We present findings on CT performed for conductive hearing loss in two patients showing associated SSCD with tegmen tympani defect. In our patients we observed absence of the bony coverage of the superior semicircular canal, tegmen tympani defect and meningoencephalocele in the middle ear in contact with the inner-ear bones. Both, encephalocele with bone fixation and SSCD can be a cause of conductive hearing loss.

We have used a 64 multidetector scanner in bone window with axial, coronal and oblique reconstructions of the petrous bone. In one patient we have added an MR imaging study to confirm the encephalocele using a 1.5 Tesla magnetic field.

Our first patient is a 59 year old female with conductive hearing loss since 2 years ago. Physical examination revealed unsteadiness with the Valsalva maneuver (Hennebert sign).

Acumetry test: Rinne right ear negative, left ear positive (normal). Weber lateralized to the right. Impedance-audiometry was normal in both sides and Pure-tone audiometry of the right ear showed 15% conversational loss for transmissive bass sounds (conductive hearing loss).

In consequence of this findings a CT was performed. Fig. 1 on page 5 Fig. 2 on page 5 Fig. 3 on page 6 Fig. 4 on page 6

We observed absence of the bony coverage of the superior semicircular canal, tegmen tympani defect and a small image of soft tissue density located at the tegmen tympani in direct contact with the inner-ear bones suggesting encephalocele in the middle ear.

The brain MRI showed a soft tissue signal in the area of the right epitympanum on T1 and T2 sequences compatible with a small encephalocele. Even though MR imaging is not the best option when evaluating temporal bone, it is useful when there is CSF leak and/or encephalocele suspicion.

Our second patient is another 59 year old woman with chronic hearing loss. Physical examination revealed normal otoscopy; Acumetry test: Rinne negative on both sides (abnormal) Weber test not lateralized. Pure-tone audiometry results showed conductive hearing loss on both sides more severe on the left ear.

CT was performed Fig. 5 on page 7 Fig. 6 on page 8 showing bilateral tegmen tympani defect, superior semicircular canal dehiscence and middle ear encephalocele on both sides. The ossicular compromise was greater on the left side.

No surgical treatment was performed by willing of the patients as there was no severe symptomatology.
Fig. 1: Right: Absence of the bony coverage of the superior semicircular canal and absence of the tympanic roof, compatible with patient’s symptoms. Left: Superior semicircular canal’s normal morphology. The tegmen tympani is present and observed as a well defined lineal bone covering the middle ear.

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Fig. 2: Soft tissue density located in the anterior superior margin of the epitympanum in continuity with the tympanic roof, and touching the malleus head, suggestive of meningocele/ encephalocele. Left: normal anatomy.
**Fig. 3:** Right side A: Shows how the superior semicircular canal opens to the skull base with no bone covering. B: Tegmen tympani defect C: Encephalocele through the anterior superior margin of the inner ear in close contact with the malleus head. Left side. A: Semicircular superior canal with adequate overlying bone covering. B: Intact tegmen tympani at the middle ear. C: The brain and dural soft tissues are separated by the tegmen tympani.
Fig. 4: Comparison of CT and MR study over the petrous bone of the right ear. CT shows absence of the tegmen tympani and a soft tissue density against the malleus head. MR confirms small extension of the subarachnoid space to the middle ear compatible with epitympanic meningocele. Note the concave vs. convex morphology of the brain tissue above the tympanic roof due to the absence of bone covering, the intracranial structures prolapse gives such appearance.

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**Fig. 5:** A different patient, CT images showing occupation of the upper epitympanic segment by a soft tissue density component that arises from the cranium. Direct contact with the head of the malleus and the incus. Findings are more prominent on the left side. Pure-tone audiometry showed conductive hearing loss on both sides more severe on the left ear.

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**Fig. 6:** 59 year old patient studied for conductive hearing loss. There is Superior Semicircular Canal Dehiscence in right and left ear associated to bilateral Tegmen Tympani Defect. There was bilateral encephalocele aswell (not shown in this image). In the center scheme showing how the superior canal opens directly in the cranium vault and the lack of tegmen tympani.

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Conclusion

Superior semicircular canal dehiscence might coexist with tegmen tympani defect and must be suspected by the radiologist. Both can be bilateral or unilateral, asymptomatic or cause several auditory or vestibular manifestations.

It is imperative a careful evaluation of the superior semicircular canal in conductive hearing loss studies, looking for SSCD and associated TTD with small encephaloceles or CSF leak signs.

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