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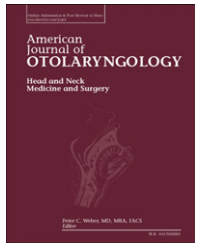

A case report of two different skull base pathologies causing hearing loss in the same ear: vestibular schwannoma and superior semicircular canal dehiscence☆,☆☆

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ARTICLE INFO

Article history:
Received 28 January 2015

ABSTRACT

Vestibular schwannoma and superior semicircular canal dehiscence are both uncommon entities, especially when present in the same ear. Here we illustrate how both of these pathologies can be repaired through the same surgical exposure, of the middle cranial fossa, with complete preservation of the cochlear nerve function and relief of symptoms caused by canal dehiscence.

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1. Introduction

Several diseases of the lateral skull base may present with hearing loss. The main pathologies include benign and malignant neoplasms, acute and chronic infections, or dehiscence of the skull base and semicircular canals. Vestibular schwannoma (VS) is the most common tumor that causes sensorineural hearing loss [1]. Superior canal dehiscence (SSCD) has also been described to cause decrease in hearing by dissemination of the acoustic energy through a mobile third window created by the dehiscence [2]. We describe a case report of a patient that presented with symptoms of hearing loss and dizziness and was found to have both of these rare pathologies in the same ear.

2. Case reports

A 40-year-old white female presented with a 12-week history of left ear tinnitus and autophony. Moreover, she complained of dizziness provoked by loud sounds. Her past medical history included asthma and Hashimoto's thyroiditis.

On physical examination she had normal bilateral cranial nerve examination and her outer ear canals and eardrums assessment revealed no pathology. When asked to perform Valsalva maneuver, patient reported a slight shift of the visual scene and dizziness. The Weber fork exam lateralized to the left ear, and the Rinne test was positive bilaterally. Audiologic testing demonstrated a mixed low frequency mild to moderate left-sided low frequency hearing loss (Fig. 1).

☆ Conflicts of interest: None.
☆☆ Disclosure of funding: No disclosures noted from authors.

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http://dx.doi.org/10.1016/j.amjoto.2015.02.001
0196-0709/Published by Elsevier Inc.

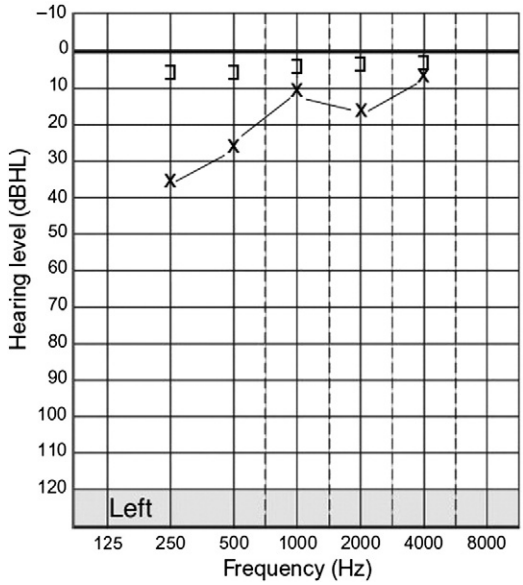

Fig. 1 – Pre-operative audiogram: Demonstrating mixed mild to moderate low frequency left-sided hearing loss with air-bone gap.

Patient was also found to have presence of stapedius reflex on the affected side.

Due to asymmetric unilateral hearing loss, a magnetic resonance imaging (MRI) with gadolinium was obtained demonstrating a 0.8 × 0.4 × 0.4 cm homogeneously enhancing mass iso- to hypointense on T2-weighted images within the left internal auditory canal fundus, most compatible with an intracanalicular VS (Fig. 2). Moreover, a concomitant left SSCD was suspected on MRI. Patient opted not to have further computer tomography imaging done. Thus, SSCD was diagnosed based on physical examination, audiometric results and MRI.

Given that this patient was recently diagnosed, and following an algorithm for incidentally diagnosed VS based on Hoa et al. [3], treatment options that were offered to the patient included observation, stereotactic radiosurgery, or surgical resection. Taking into account her refractory and sometimes incapacitating symptoms caused by SSCD, and given patient's good residual hearing, the patient opted not to wait but instead wanted to proceed with the surgery. Since both, the excision of the intracanalicular VS and a plugging procedure for SSCD, could be achieved during the same operation, the patient elected to undergo a left middle cranial fossa approach.

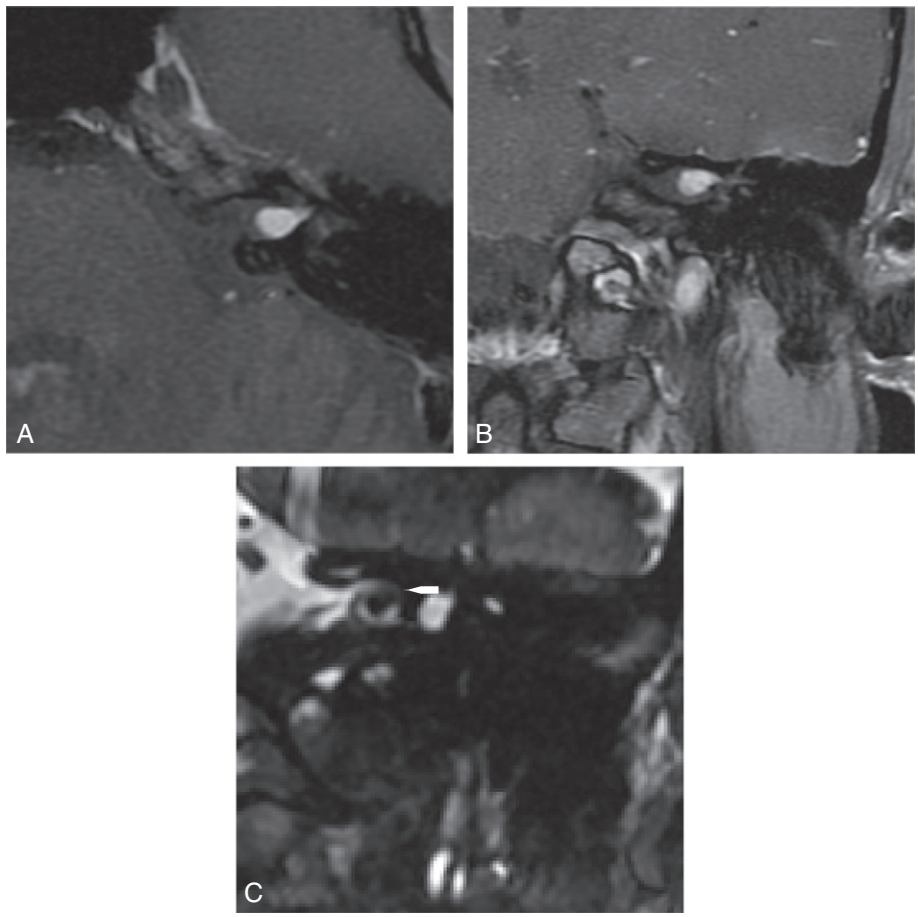

Fig. 2 – MRI imaging: (A) Post contrast T1-weighted axial image demonstrates a heterogeneously enhancing mass within the left internal auditory canal (IAC) extending to the fundus of the IAC. (B) Post contrast T1-weighted coronal image with fat suppression demonstrates that the IAC mass lies superior to the vestibular crest. (C) 3D-constructive interference in the steady state (CISS) coronal image on the left side also demonstrates marked thinning (white arrow) or absence of bone of the roof of the superior semicircular canal.

After performing temporal craniotomy, the dura was elevated off the floor of the middle fossa and canal dehiscence was identified. The dura was then further elevated anteriorly, where geniculate ganglion was also clearly dehiscent. The roof of the internal auditory canal was drilled out and dura was opened allowing tumor identification. Tumor was seen to be arising from the inferior vestibular nerve (Fig. 3). At this stage the superior canal dehiscence was packed with bone wax and tumor was then removed en-bloc using microsurgical technique. Abdominal fat graft was placed into the cranial defect. There were no complications observed during the intraoperative and postoperative period.

After 1 month, the patient reported a complete relief of dizziness, oscillopsia and autophony. There was also a partial reduction of the intensity of tinnitus in her left ear. At three month follow up, the audiogram demonstrated closure of the gap and a significant improvement of pure tone thresholds at all frequencies with word recognition score at 96% (Fig. 4).

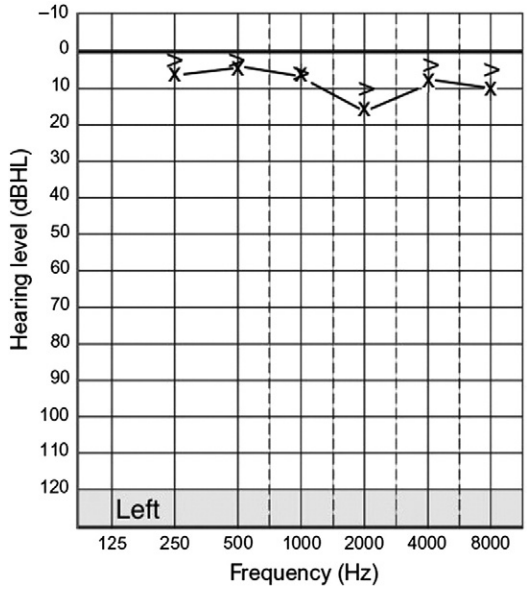

Fig. 4 – Post-operative audiogram: Three months post-operative audiogram demonstrates normalization of audiogram of the left ear with complete recovery of hearing loss and closure of air-bone gap.

3. Discussion

VS is a nerve sheath tumor that arises from the superior or inferior vestibular nerves. The most common clinical manifestation of VS is unilateral sensorineural hearing loss (present in 95% of patients), often in association with tinnitus [1]. As the tumor grows larger patients can present with symptoms of facial numbness or weakness from compression on the trigeminal and facial nerve respectively [4]. MRI with gadolinium contrast is the gold standard for the diagnosis or exclusion of VS [5]. Surgical removal and stereotactic radiosurgery are the primary management modalities for VS. The middle cranial fossa approach is well suited for patients with good residual hearing (pure-tone average ≤50 dB) and tumor size <1.5 cm (at our institution <2 cm when appropriate) in the cerebellopontine angle [6]. The middle fossa approach is unique, compared with the posterior fossa craniotomies, because the entire internal auditory canal (IAC) is accessible without violating the inner ear. This exposure allows for the removal of intracanalicular tumors while preserving hearing [6].

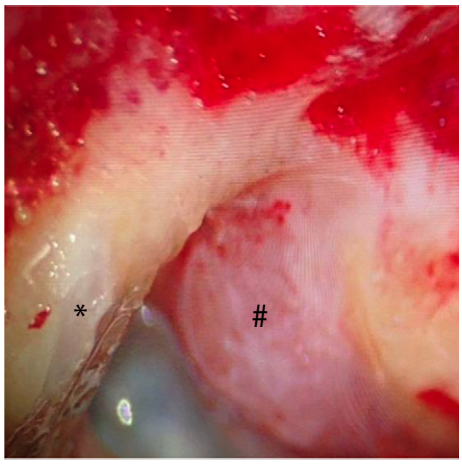

Fig. 3 – Surgical exposure: Middle cranial fossa exposure of left vestibular schwannoma (#) with SCCD (*), demonstrating easy accessibility to both pathologies from one surgical approach.

Semicircular canal dehiscence causes an abnormal communication between the inner ear and the surrounding structures. The most common is the SSCD, which may lead to a syndrome of vertigo and oscillopsia, being referred to as SSCD syndrome [2]. Patients complain of vertigo when exposed to loud noises (Tullio phenomenon), Valsalva maneuvers, pressure changes in the ear (Hennebert sign), or with factors that raise intracranial pressures. Auditory symptoms include sensitivity to bone-conducted sounds and autophony. The dehiscent portion of the superior canal acts as a third mobile window allowing acoustic energy to be dissipated there. The presence of stapedius reflex with low-frequency conductive hearing loss should prompt radiological imaging of the inner ear to exclude the possibility of dehiscence of the inner ear [7]. Patients with debilitating symptoms may require surgical repair via an open or endoscopic middle cranial fossa approach during which the dehiscent area of the canal may be repaired with canal plugging, capping, or resurfacing procedures [2]. Round window tissue reinforcement and plugging of the superior semicircular canal via a transmastoid approach are alternate approaches for SSCD repair [8,9].

The overall incidence of VS is about one per 100,000 persons/year [2]. However, the incidence appears to be increasing, due at least in part to the incidental diagnosis of asymptomatic lesions with the widespread use of MRI and computed tomography [1,10]. As an example, a retrospective analysis of 46,000 MRI scans, done for other reasons, identified eight unsuspected vestibular schwannomas (0.02%), and autopsy studies further suggest that the prevalence may even be higher. Carey et al. [11] studied 1000 temporal bones (596 individuals) obtained from autopsies,

with the purpose to determine the prevalence of SSCD in the general population. They found a 0.7% of the total individuals with dehiscence of the superior semicircular canal.

VS and SSCD are considered uncommon entities in the general population. Moreover, both are frequently asymptomatic and may present incidentally on imaging. Our case presents a significant challenge as she had two infrequent diagnoses (VS and SSCD) each of which could be contributing to her clinical picture. The patient likely presented due to symptoms of SSCD, but given her imaging and audiogram findings early hearing loss caused by her intracanalicular VS could not be ruled out. Both clinical algorithm [3] and patient's preference were used when choosing the type of treatment for her disease process. Given the patient's young age, small tumor size, the desire to preserve good residual hearing, and patient's preference against long-term follow-up with annual MRIs, operative resection was her treatment of choice for the VS independent of her SSCD. Furthermore, her SSCD symptoms guided the patient in choosing microsurgery over Gamma knife for her VS. Based on the location and the size of her tumor, a middle fossa approach was deemed most appropriate. This had the putative effect of allowing for simultaneously repairing the SSCD via the traditional middle fossa craniotomy.

After a review of literature, we report a first case of VS and SSCD identified in the same ear, and especially, both generating symptoms. This report shines light to pay attention that SSCD could be followed by other neurotologic entities, such as VS and vice versa, and that a full otologic and audiologic exam should always be done to not miss either of these pathologies. The investigation of these pathologies with appropriate imaging preoperatively was important, as both can be approached through the same surgical exposure of middle cranial fossa. This allowed removal of the cerebellopontine angle tumor and plugging of the superior canal dehiscent with complete preservation of the cochlear nerve function and relief of symptoms caused by the SSCD syndrome while preserving and restoring hearing.

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