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Case Report

Cochlear–Internal Canal and Cochlear–Facial Dehiscence: A Novel Entity

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Different types of otic capsule dehiscence restricted to the cochlea have been described. Here we describe the case of a patient with a cochlear–internal auditory canal dehiscence associated with a cochlear–facial dehiscence not reported before. A 53-year-old patient with severe to profound sensorineural hearing loss due to bilateral Meniere's disease underwent a cochlear implant surgery on the right ear. Preoperative brain magnetic resonance imaging findings were reported to be normal; during surgery, a cerebrospinal fluid gusher occurred at the time of round window opening. Postoperative computed tomography imaging showed a bony dehiscence at two levels of the otic capsule.

KEYWORDS: Dehiscence, otic capsule, CSF gusher, cochlear implantation, third window lesions

INTRODUCTION

Since the first report of otic capsule dehiscence in patients with superior semicircular canal dehiscence (SSCD) by Minor in 1998 [1], different types of otic capsule dehiscence restricted to the cochlea have been described: cochlear–carotid, cochlear–internal auditory canal, or cochlear–facial dehiscence [2]. The awareness of this phenomenon is important considering that a dehiscence in the otic capsule opens up a third window. The variety of signs and symptoms that may, thus, present can hinder the accurate diagnosis of a disease, especially if one type of dehiscence is associated with another type. To the best of our knowledge, this is the first reported case of a cochlear–internal auditory canal dehiscence associated with a cochlear–facial dehiscence.

CASE PRESENTATION

A 53-year-old patient with the diagnosis of a refractory bilateral Meniere's disease was referred to our center. He recalled that symptoms started with hearing loss fluctuation in the right ear after a head trauma 6 years ago; subsequently, the patient began experiencing vertigo spells, drop attacks, and tinnitus. During the last 12 months, fluctuating hearing loss associated with severe vertigo spells and tinnitus developed in the left ear as well. Thus, the patient was diagnosed with Meniere's disease according to the clinical American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) criteria [3].

He was previously treated with betahistine, acetazolamide, and intratympanic steroids in the left ear without any clinical improvement. Physical examination yielded the following: normal otoneurological findings and normal vestibular evoked myogenic potentials. Furthermore, video head impulse test showed normal gain for all semicircular canals except for both posterior canals. Mean tone audiometry revealed a moderate sensorineural hearing loss on the left side (55 dB) and a severe sensorineural hearing loss on the right side (80 dB). The speech discrimination score was 84% on the left side and 68% on the right side. Brain magnetic resonance imaging (MRI) performed at another institution showed normal findings (Figure 1). Hearing loss was managed with a cochlear implant (CI) on the right ear, and vertigo spells were managed with intratympanic gentamicin injections in the left ear.

Surgery was started following the standard CI procedures through posterior tympanotomy; however, due to bulging of the sigmoid sinus and a prolapsed tegmen, a canal wall-down mastoidectomy with an external auditory canal closure was performed. Surprisingly, the round window membrane was found to be distended prior to its manipulation and when punctured, a gusher of cerebrospinal fluid (CSF) was immediately noticed; it lasted over 15 min. Later, a Nucleus slim straight electrode (Cochlear, Aus-

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Figure 1. Axial heavily T2-weighted thin-slab maximum intensity projection MR image of the right inner ear showing normal anatomy without visible malformations or other anomalies.

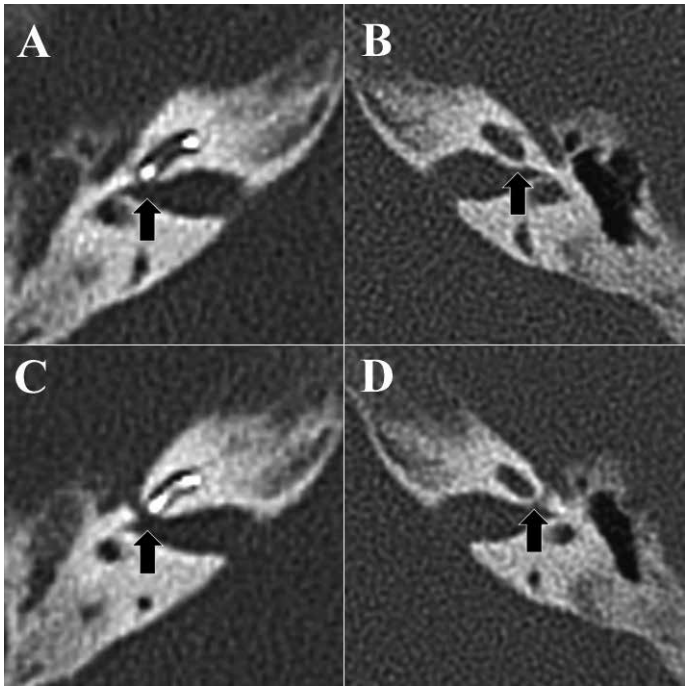

Figure 2. a-d. Axial CT zoomed-in view of in both inner ears at the origin of the facial canal (lower row: c, d) and immediately above (upper row: a, b). In the right inner ear (a, c) an osseous defect with dehiscence can be seen between the basal turn of the cochlea and the internal auditory canal (a) and between the basal turn of the cochlea and the origin of the facial canal (c). Also, note the correct location of the cochlear implant. In the left inner ear (b, d), a normal anatomy is observed, with an osseous wall between the cochlea and the internal auditory canal (b) and the facial canal (d).

tralia) surrounded by the fascia in the base was inserted. There was no suction or entry of blood directly into the inner ear, and no resistance was perceived during the insertion. Thus, the presence of the gusher when opening the round window before the electrode array insertion excludes the possibility of a fistula created during the procedure. The electrophysiological tests (impedances and NRT threshold) showed values in normal ranges, and the intraoperative radiological control were adequate. The round window was then sealed with fascia; the leakage of CSF was controlled, and finally, the cavity was filled with surrounding muscle tissue and fibrin glue. In order to determine the cause of the gusher, computed tomography (CT) was performed the following day (Siemens Somatom Definition 1 mm slice thickness/0.7 slice increment Axial). A dehiscence affecting the cochlear–internal auditory canal and facial canal on the right side was found; there were no signs of temporal bone fracture, and the left side showed no abnormalities (Figure 2). Regarding the cochlear aqueduct, only the initial portion was visible, without dilatation or patency of the distal part (not shown on images). The disyllabic word test recognition for speech discrimination performed 5 months postoperatively was 72%. Vertigo spells stopped after the intratympanic gentamicin injections in the left ear. No facial stimulation or spasms were observed after implant activation. Informed consent was obtained from the patient.

DISCUSSION

The presence of dehiscence in the otic capsule can be an acquired syndrome when associated with pathological conditions (tumors or cholesteatomas), elevated intracranial pressure (as described in tegmen thinning), or an age-related mechanism causing otic capsule bone reabsorption [4]. On the other hand, this entity may be congenital, caused by an alteration in the embryologic development of the otic capsule such as cases of cochlear–internal auditory canal dehiscence in Mondini-like dysplasia or cochlear–facial dehiscence associated with Fallopian canal dehiscence. The fact that the cochlea and vestibule show no malformations suggests that the defect develops at a stage of development that is more advanced than the otocyst stage [5]. The labyrinthine capsule remains cartilaginous until the inner ear reaches its final size at about 25 weeks of gestation. The interscalar septum and the bone between the cochlea and the vestibule develop from the ossification centers. Partly missing this bone suggests over-resorption, resulting in a defect of the cartilaginous capsule and a failure of ossification [6]. In our patient, presence of an otic capsule dehiscence at two levels, internal canal and facial nerve, suggests that it may be a congenital disorder rather than an acquired one related to the history of head trauma. The fact that CT was performed postoperatively may bring up the possibility of such dehiscence being an artifact. Yet, as it is clearly shown in the images (Figure 2 a, c), the study was clear of metal-related artifacts, and as can be noted on the opposite side of the cochlea, the implant itself does not impede the evaluation of the adjacent bone. Also, we have not found any similar image on CT scans of other patients after cochlear implantation (not shown).

An otic capsule dehiscence opens up a third window into the inner ear, and clinical findings such as a low-frequency bone conduction hearing loss, dizziness, vertigo, or Tullio phenomenon have been reported [2,7]. When presuming an otic capsule dehiscence, it is important to consider other conditions that might resemble or mimic a third window syndrome, as is the case of vestibular aqueduct enlargement or perilymphatic fistula. The first one is often associated with a fluctuating and progressive neurosensorial hearing loss, and in some cases, an air-bone gap in low-frequencies similar to a third window may appear; however, its underlying mechanism is still controversial [8]. A perilymphatic fistula usually occurs after an identifiable traumatic event, and vestibular symptoms, mainly dizziness and disequilibrium, are more common than hearing loss [9].

In our patient, the clinical symptoms started in the right ear as a low-frequency mixed hearing loss that fluctuated and that was associated with vertigo spells. Clinical symptoms suggested Meniere's disease. However, symptoms and the disease persisted despite medical and intratympanic treatments. As in other reported cases of otic capsule dehiscence, VEMPs were preserved [7]. Although VEMPs are highly sensitive and specific for SSCD [10], the negative finding in this scenario may be attributed to the small size of the dehiscence [2]. Unfortunately, we do not know for certain the exact role that this entity plays in the present case; could this dehiscence be the cause of the refractive treatment in this particular case of bilateral Meniere's disease? Was the right ear more affected because of it? To be able to answer the above questions, we need to become aware of and better understand this entity.

An abnormal communication between the subarachnoid space and the perilymphatic space is the underlying cause of a CSF gusher. The inner ear abnormality that is most often responsible of this phenomenon is a bony defect in the fundus of the internal acoustic meatus [11]. There are no previous reports of CSF gusher in the context of an otic capsule dehiscence, and this clinical condition may be a sign of dehiscence, as it was in the present case. In our patient, a total electrode insertion was possible and all electrodes were activated.

We present a case of a previously undescribed entity, an otic capsule dehiscence at two levels (cochlear–internal canal and cochlear–facial dehiscence) in the context of bilateral Meniere's disease, which was responsible for a CSF gusher during a CI surgery. An accurate diagnosis of otic capsule dehiscence can only be made using a high-resolution temporal bone CT (2, 10). We did not perform preoperative CT in this patient because at our center, MRI is the imaging study that is routinely performed first in patients with the indication of a CI; CT is performed only in those cases where a malformation or other anatomical anomalies are detected. Thus, as a preoperative study, we recommend both MRI and CT scan.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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