Case Report

Internal Auditory Canal Meningocele-Perilabyrinthine/Translabyrinthine Fistula: Case Report and Imaging

Running Title: Internal Auditory Canal Meningocele with Fistula

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Source of Financial Support/Funding: None

Financial Disclosures: None

Conflict of Interest: None

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Abstract:

The case of a 17 year-old patient with progressive unilateral sensorineural hearing loss and temporal bone malformations concerning for internal auditory canal meningocele with translabyrinthine/perilabyrinthine cerebrospinal fluid fistula is presented with associated computed tomography and magnetic resonance imaging. As the second reported case of an un-ruptured internal auditory canal meningocele with translabyrinthine/perilabyrinthine fistula, the case presents several clinically relevant points for otologists, neurotologists and neuroradiologists. Although rare, it is an additional entity to consider as a cause of unilateral sensorineural hearing loss and may pose risk for developing meningitis and possible "gushing" of CSF should surgical intervention be attempted.

Introduction:

Congenital temporal bone malformations are rare, but can have potential for cerebrospinal fluid (CSF) leak from perilymphatic fistula. Perilymphatic fistulas are classically divided into congenital or acquired[1]. Acquired causes are due to surgical intervention, trauma, infection, or spontaneous[1]. Congenital temporal bone CSF fistulas are rare. They are categorized as perilabyrinthine or translabyrinthine depending on the route of CSF transmission. Pre-operative diagnosis of this defect is important, as surgical intervention can result in "gushing" of CSF or perilymph. Intraoperative management of fluid "gushers" can be challenging for surgeons who attempt cochlear implantation. The increased fluid flow, the result of a bone defect in the cochlea, allows direct communication between CSF in the internal auditory canal (IAC) and perilymph.

Case Report:

Patient is a 17 year-old male who presented with progressive left sided hearing loss. His hearing loss was first detected after failing audiology screening for school. He was then followed with serial audiograms. His only symptom was progressive left sided hearing loss. He denied trauma, otorrhea, rhinorrhea, tinnitus, noise exposure, recurrent infections, meningitis, or prior otologic procedures. He reported one prior brief episode of dizziness associated with fevers and cough. There were no other vestibular symptoms. His mother reported nonspecific hearing loss in his father, but further paternal medical history was limited.

The physical examination was normal. There were no auricular malformations. Under microscopy the external canal, tympanic membrane and middle ear were normal. Audiogram at

presentation showed mild left sensorineural hearing loss, with 2K notch down to 55dB, word discrimination was 90% in left ear and 100% in the right ear. Otoacoustic emissions were absent on left, present on the right with tympanometry type A. Audiogram obtained two years later showed progressive worsening of sensorineural hearing loss in the left ear. This audiogram demonstrated worse loss at 2K down to 60 dB, new dip at 250 dB to 65 dB, word discrimination was poor at 40% in the left ear and 100% in the right ear. Otoacoustic emissions were absent on the left, present on the right with tympanometry type A.

Imaging findings were extremely complicated. Computer tomography image (CT) showed asymmetric enlargement of the left IAC with enlargement of the entire left facial nerve canal, including a bulbous geniculate segment (Fig. 1-4). No abnormal enhancement was seen along the course of the facial nerve to suggest that the widening was due to a mass lesion, such as schwannoma (Figure 4).

There was osseous thinning and probable dehiscence of the bone separating the geniculate segment of the facial nerve and the subarachnoid space. Heavily T2-weighted high-resolution axial MR imaging showed expansion of the left IAC, and dilated CSF filled meningeal spaces around the labyrinthine and geniculate segments of the facial nerve canal (Figure 4).

Discussion:

The imaging showed marked dilatation of the IAC and entire facial nerve segments. This was essentially diffuse ectasia of the meningeal sheath around the facial nerve, from the labyrinthine segment through the middle ear. There was also a perilabyrinthine/translabyrinthine fistula.

The initial interpretation, based on the bulbous morphology of the distal IAC, was of an X-linked stapes gusher. This is a classic radiologic misinterpretation based on "pattern recognition." In X-linked stapes gusher, there is enlargement of the internal auditory canals and labyrinthine/geniculate segments of the facial nerve canal, but it is *bilateral*[2].

X-linked gusher is associated with congenital absence of the lamina cribrosa, leading to profound sensorineural hearing loss with or without conductive hearing loss[2,3]. Usually the cochlear modiolus is absent and there is a type III cochlear incomplete partition ("corkscrew" cochlea)[2]. This case, therefore, is not X-linked gusher because it was unilateral, and the modiolus and cochlea were normal.

In our case, there was diffuse ectasia and dilatation of the IAC, and the dilatation extended to the entire facial nerve canal to the mastoid portion. Fortunately, there was no history of otorrhea. As there was no history of prior surgery or trauma, it was likely congenital in etiology.

Perilabyrinthine fistulas do not involve the bony labyrinth. There are 5 potential routes of these fistulas: through 1) the tegmen tympani, 2) a large apical air cell, 3) Hyrtl's fissure, 4) the petromastoid or subarcuate canal or 5) the facial nerve canal[4]. A perilabyrinthine fistula can coexist with a middle cranial fossa meningocele which was not seen in this case.

A translabyrinthine fistula occurs through a defect in the lamina cribrosa (a thin layer of bone separating the apex of the IAC and the vestibule), which could lead to perilymphatic hydrops[4]. Transmission of pressures from the CSF space can result in displacement and/or perforation of the stapes footplate resulting in a unilateral stapes gusher during surgical intervention[3].

In this case, both clinical and radiographic findings were unilateral. This case has several distinguishing features similar to both perilabyrinthine/translabyrinthine fistulas. There is bony dehiscence of both the lamina cribrosa, middle turn of the cochlea and the temporal bone overlying the geniculate segment of the facial nerve canal. The IAC and facial nerve canals demonstrate marked dilatation. The modiolus is intact and the cochlea demonstrates a normal configuration. The constellation of findings suggests an internal auditory canal meningocele with associated translabyrinthine fistula with or without a perilabyrinthine fistula. Such cases may lead to "unilateral stapes gushers" as opposed to the congenital "X-linked stapes gusher" and X-linked congenital deafness.

There are some differences in this case when compared to other perilymphatic fistulas. The patient is older at presentation (15 years old) than most patients with this abnormality who usually present in the first 5-10 years of life[4]. His presentation is also not classic; sensorineural hearing loss is his only symptom, but most patients present with: 1. CSF rhinorrhea (TM intact, with fluid being transmitted through Eustachian tube, 2. CSF otorrhea (TM is perforated either spontaneously or during myringotomy), 3. Recurrent meningitis[5,6]. The lack of CSF otorrhea or rhinorrhea in these translabyrinthine fistulas suggest the temporal bone meningocele is unruptured. There has been only one other case reported of a translabyrinthine fistula that was unruptured[6]. This previously reported patient's temporal bone meningocele was covered by the arachnoid membrane when evaluated surgically[5].

Although the case subject and his family denied any genetic testing, it is important to consider genetic contributions to the temporal bone defect development and hearing loss seen in this case. Nance *et al* first described a congenital X linked mixed hearing loss with fixed stapes footplate and perilymph gushing when approached with stapedectomy[7]. X linked mixed deafness

was localized to the X q 13q-q21.1 region by Wallis *et al*[3]. Another study by Phelps et al evaluated 7 pedigrees with known X linked mixed hearing loss; some of the affected subjects showed enlarged facial and IAM canals[8]. In this series, female obligate carries showed a lesser degree of involvement of malformation on CT imaging. Some males in the series had X linked deafness but no significant CT abnormalities[8]. All of the temporal bone abnormalities in those cases were bilateral, establishing the possibility of a new genetic variant producing the unilateral defect seen in this case. It is possible that our patient potentially has another X linked locus contributing to his defect and deafness.

Clinically, perilabyrinthine/translabyrinthine fistulas are most often differentiated by the presence of hearing loss. Perilabyrinthine fistulas lack hearing loss whereas translabyrinthine fistulas are associated severe or complete hearing loss[4]. This patient has progressive sensorineural hearing loss not characteristically seen in translabyrinthine fistulas. There is no clear explanation for why the patient in this case presented much later and had sparing of some hearing function that only has become progressive in the last two years. The mechanism of hearing loss is thought to be due to the pressure of transmission of CSF fluid from the IAC to the cochlea or vestibule.

Regarding the management of perilymphatic fistulas, surgery is avoided in those with only hearing loss as the symptoms are unilateral. There have been cases of translabyrinthine fistulas repaired by packing the internal auditory meatus (IAM) with muscle to stop CSF gushing during stapedectomy[9]. It is also of note that patients with translabyrinthine fistulas have increased risk of CSF otorrhea with meningitis if the meningocele is ruptured. Surgical intervention for this life threatening complication can be attempted with surgical repair of leak with a subtotal petrosectomy or middle fossa craniotomy[9,10].

Conclusion: Congenital perilymphatic fistulas of the temporal bone (perilabyrinthine and translabyrinthine) are rare and identification affects clinical management. In this patient, progressive sensorineural hearing loss was the only presenting symptom, but identification of the malformation enables physicians to council their patients on possible complications of otitic meningitis as well as spontaneous CSF otorrhea, rhinorrhea, or gushing intraoperatively. This patient's imaging demonstrates temporal bone features seen in both perilabyrinthine and translabyrinthine fistulas. Recognition of these findings may alter surveillance, treatment options and counseling regarding likelihood of progressive hearing loss.

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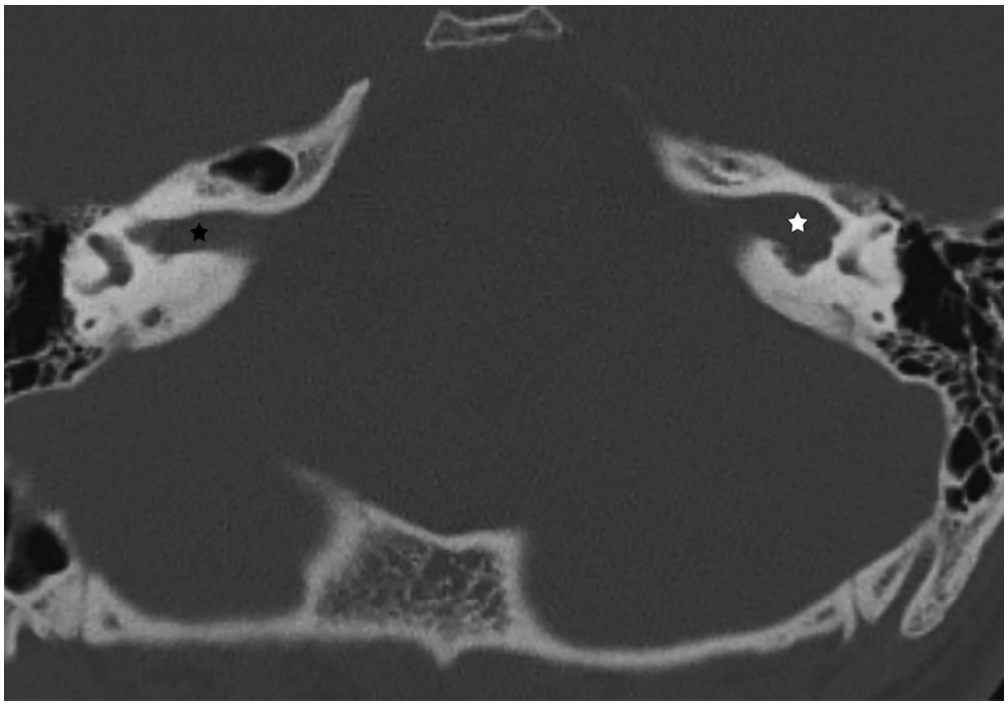
Figure 1: Axial CT image through the skull base demonstrate and enlarged and bulbous appearance to the left internal auditory canal (white star) compared to the right (black star).

Figure 2: Coronal oblique CT image through the left temporal bone demonstrates a dilated internal auditory canal (white star), widening of the labyrinthine (black arrow) and geniculate (white arrow) segments of the facial nerve canal and thinning of the lamina cribrosa (white arrowhead). The modiolus (black arrowhead) and the vestibule (not imaged) are normal.

Figure 3: Coronal CT image through the left temporal bone demonstrates an enlarged geniculate segment of the left facial nerve canal (white star). There is frank dehiscence of the temporal bone skull base overriding the dilated geniculate canal (white arrow).

Figure 4: Coronal oblique 3D T2 weighted thin section MR image (FIESTA) through the left temporal bone demonstrate cystic expansion of the left internal auditory canal at the fundus (white arrowhead) with the facial nerve (black arrow) coursing to a dilated geniculate segment of the left facial nerve canal (white arrow). Of note, there was no abnormal enhancement or abnormal signal on T1 weighted images of the left facial nerve to suggest a mass lesion such as schwannoma or cholesterol granuloma.



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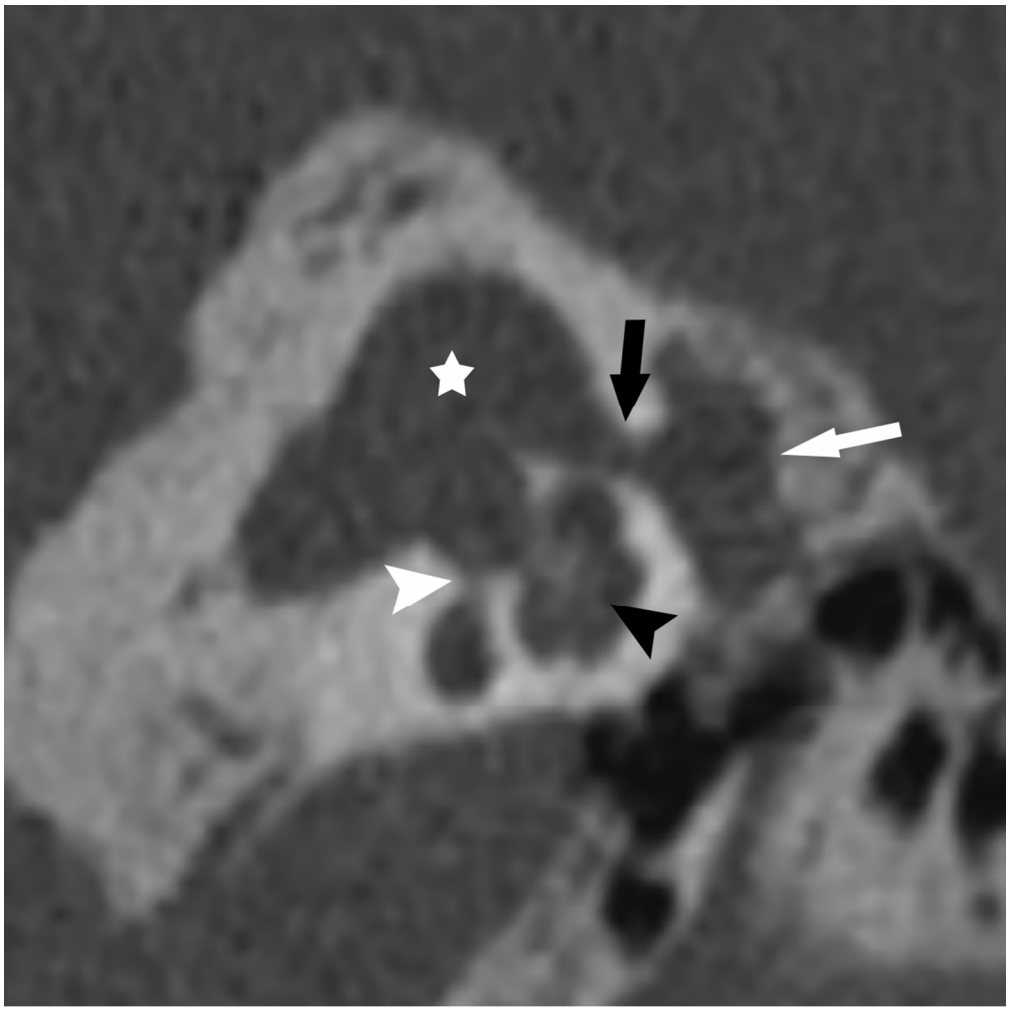

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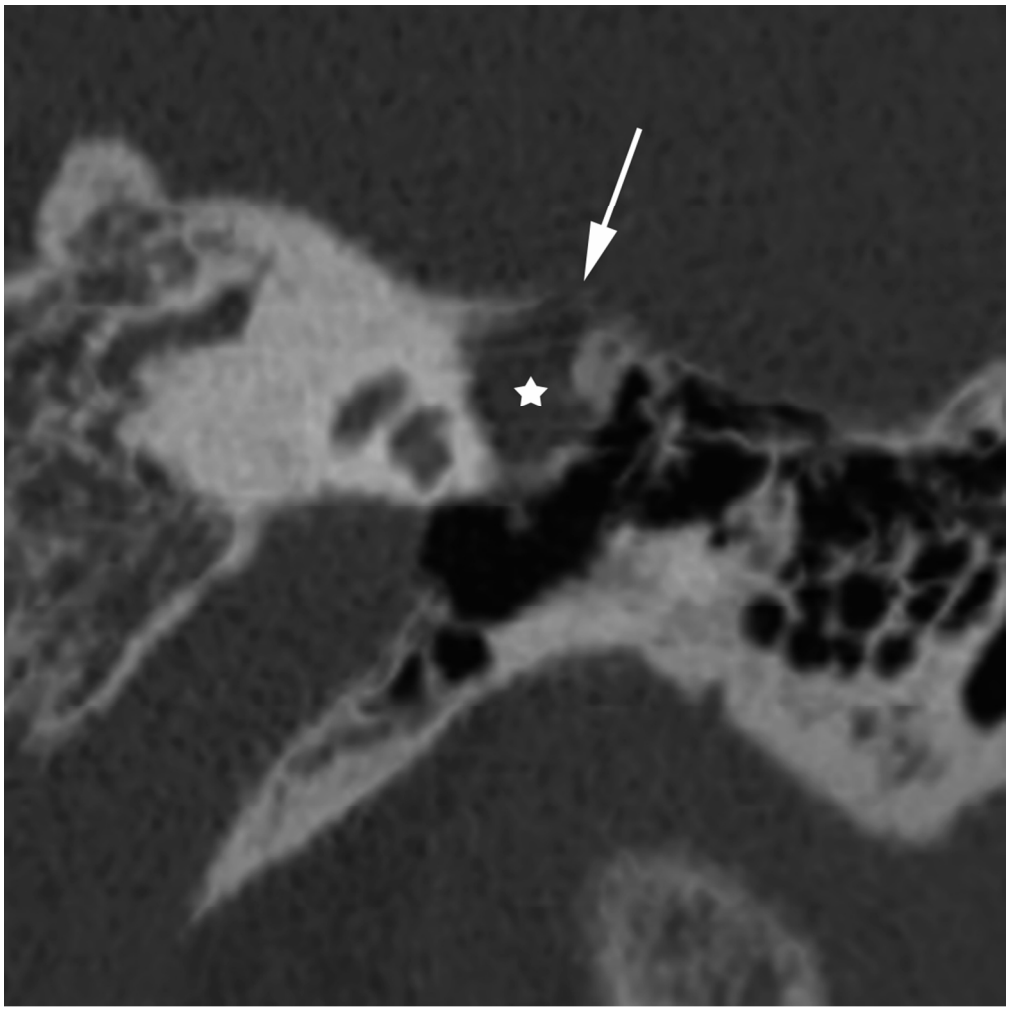

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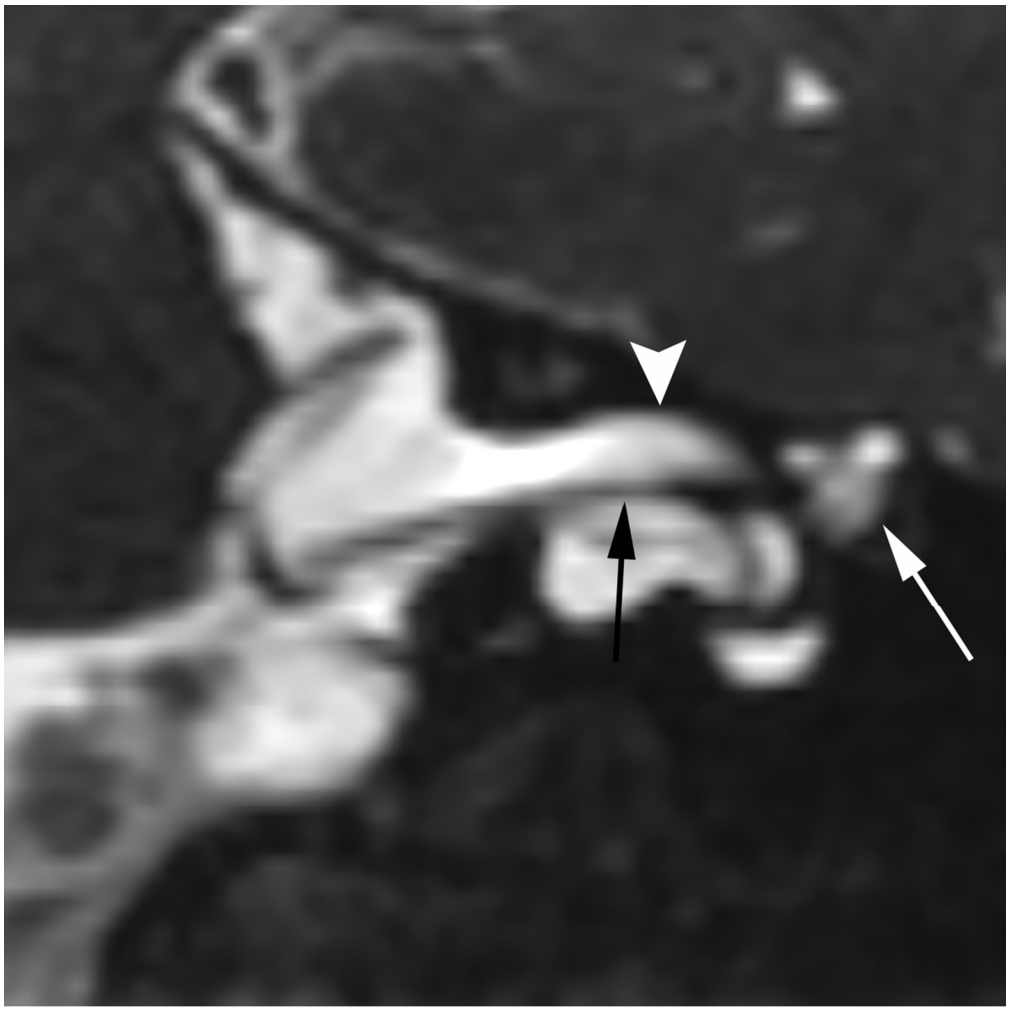

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