CASE REPORT

Audiological Changes in Perilymph Fistula

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ABSTRACT

Introduction: Trauma is one of the commonest causes of perilymphatic fistulae. The classical patient presents with a sudden onset vertigo, tinnitus and hearing loss, which may be fluctuating. The audiological changes can range from a purely conductive hearing loss, to mixed hearing losses (most common) to sensorineural hearing loss. The same are dependent on a number of factors, such as extent of trauma and perilymphatic leak, the presence of superadded infection, the tardiness in obtaining appropriate treatment, etc.

Materials and methods: A case of post-traumatic perilymphatic fistula has been presented here, and discussed with regards to the presentation, clinical diagnosis, treatment and the audiological profile and eventual outcome.

Conclusion: The audiological profile of a patient should be carefully correlated with the CT scan findings, the clinical picture as well as the intraoperative findings so as to predict and understand the eventual audiological outcome after treatment.

Keywords: Perilymph fistula, Trauma, Hearing loss, Bone conduction.

Abbreviations: dB: decibels, Hz: Hertz, KHz: Kilo hertz, BPPV: benign paroxysmal positional vertigo, Deg: degrees.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Perilymphatic fistulae are essentially abnormal communications between the perilymphatic space and the middle ear.[3] These may either be congenital, or traumatic (iatrogenic, accidental), and may result in vertigo of varying degrees, hearing loss and tinnitus. Traumatic perilymph fistulae are caused commonly due to barotraumas or fractures of the temporal bone, or on rare occasions they may occue following acoustic trauma. A perilymph fistula occurs following manipulation by a medical practitioner is rarer still, and hence presented here with its management.[4]

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CASE PRESENTATION

History

A 14-year-old male student was referred with a sudden onset of tinnitus and giddiness and loss of hearing from the right ear following an attempt to remove an eraser (foreign body) from the same ear by a general practitioner. The patient had accidentally inserted the eraser into the right ear in an attempt to scratch the ear. Following his inability to remove the eraser himself, he visited the general practitioner, who tried to grasp the eraser with some sort of forceps. The eraser was successfully removed, but excessive bleeding following the procedure, associated with incapacitating vertigo and vomiting led the patient to present to the emergency department.

There was no relevant past, family and social history. The patient informed that he had suffered a bout of acute otitis media 2 years ago (details available), and had been managed conservatively for the same, with no significant perceptible audiological sequelae.

Clinical Course

On initial assessment, the patient's right external auditory canal was occluded with blood clots, with no signs of active bleeding. He complained of a noticeable hearing loss and severe giddiness and vomiting following the removal of the eraser, and tinnitus. The initial tuning fork assessment revealed a negative Rinne's test with 256 and 512 Hz on the right, and positive responses on the left. Also, the Weber's test was lateralized to the left. The absolute bone conduction was reduced in the right ear.

The pure tone audiological assessment revealed a right sided moderately severe mixed hearing loss with a dip at 4 KHz, and a normal hearing sensitivity on the left, with a similar dip at 4 KHz (Fig. 1).

A provisional diagnosis of a post-traumatic perilymph fistula with ossicular discontinuity was made, and the patient was managed conservatively with antibiotics, bed rest (with head elevation), and antivertiginous medications. Within 2 days of presentation, the giddiness had subsided for the most part, and only occurred on straining. The tinnitus had

reduced in intensity, and was more 'perceptible' in a silent ambient surrounding. The hearing loss remained the same.

A microscopic evaluation of the right ear revealed an intact but intensely congested tympanic membrane. An impedence audiogram revealed an abnormally high static compliance, suggesting an ossicular discontinuity (Fig. 2). However, the patient did not complain of a change in the existing vertiginous sensation during the rise and fall in pressure while performing the impedence audiometry (fistula test was negative).

A high resolution temporal bone CT scan showed the presence of fluid in the middle ear, with disruption of the ossicular chain. It was however difficult to appreciate the exact location and position of the stapes, which seemed to be fractured, as the individual crurae could not be identified. The malleus too was not visualized.

A week after the initial presentation (i.e. 9 days after the trauma), the patient complained of persistent hearing loss and occasional tinnitus. The giddiness was only present on straining, but was a consistent feature.

Treatment

After counseling, the patient and his relatives consented to an exploratory tympanotomy with due consent for ossicular reconstruction, under general anesthesia. On elevating the tympanomeatal flap, the malleus was found to be 'absent'. On curetting the posterosuperior bony facial shelf, the incus was found to be dislocated posteromedially, abutting the Fallopian canal (Fig. 3). The stapes was found fractured, discontinuous with the incus, and 'inverted' into the oval window (Fig. 3). On performing an 'anesthesiologist's valsalva maneuver' (forced insufflations against a closed expiratory valve by the anesthesiologist), a clear fluid was seen to emanate from the oval window region. The dislocated stapes was gently teased out of the oval window with a curved pick (Fig. 4), there was some evidence of sclerosis

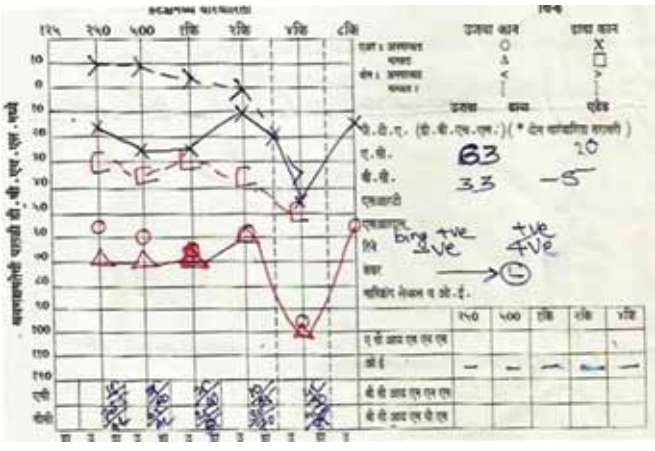

Fig. 1: Preoperative pure tone audiogram, showing the mixed hearing loss in the right (affected) ear. Also note the incidental dip at 4 KHz in the left ear as well, which otherwise has a normal hearing

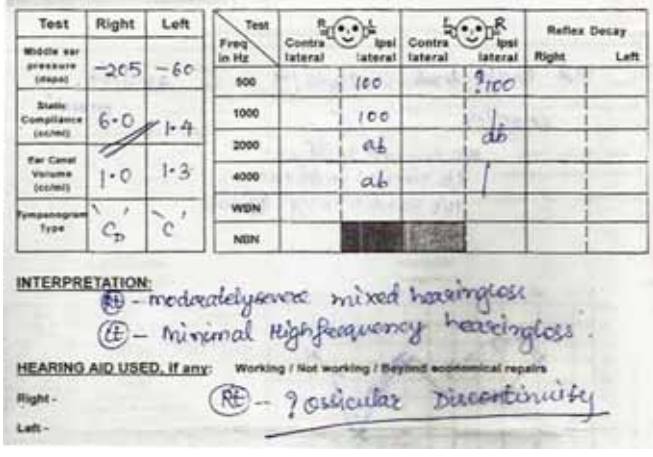

Fig. 2: Preoperative impedence audiogram showing the high static compliance in the right ear suggestive of ossicular discontinuity

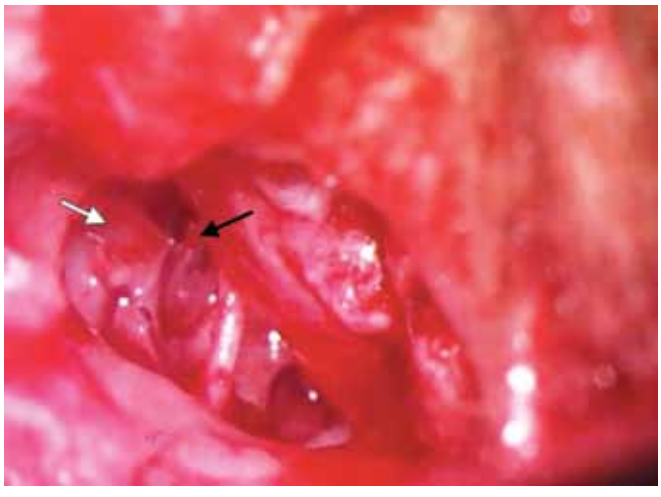

Fig. 3: On elevating the tympanomeatal flap with an endaural approach, the fractured stapes suprastructure is seen in the oval window (black arrow), and the posteromedially displaced incus is seen abutting the fallopian canal (white arrow)

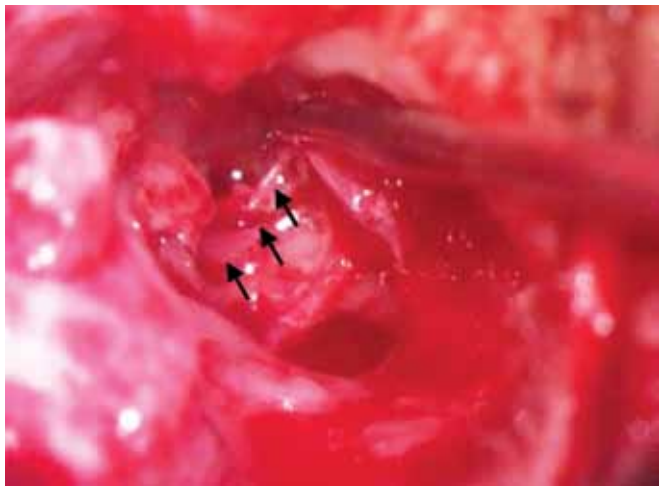

Fig. 4: The numerous fragments of the fractured stapes (black arrows) being gently teased off from the oval window with a curved pick

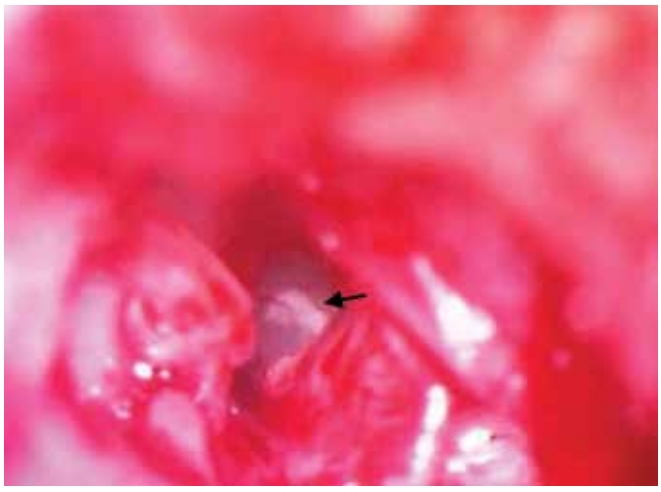

Fig. 5: The exposed saccular membrane seen in the oval window with a patch of sclerosis seen (black arrow)

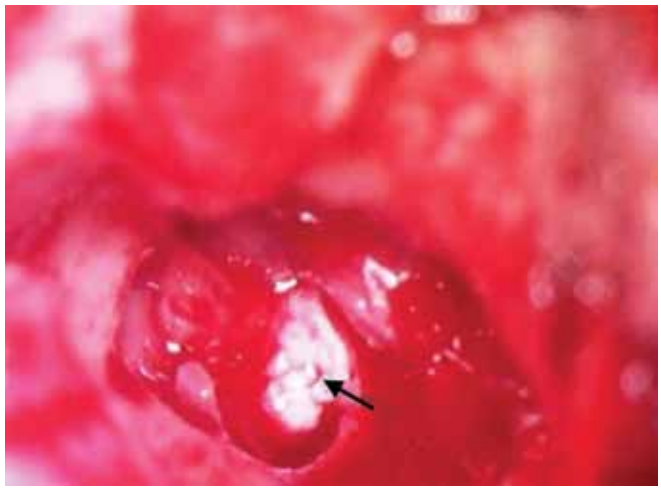

Fig. 6: Two layered seal of temporalis fascia (black arrow) placed to cover the oval window, and tucked medial to the incus for support

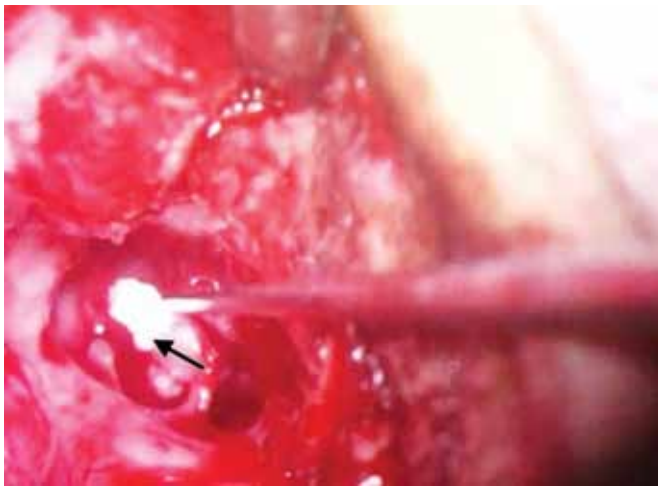

Fig. 7: Gelfoam (black arrow) being placed over the fascial seal and the incus, to reinforce the seal

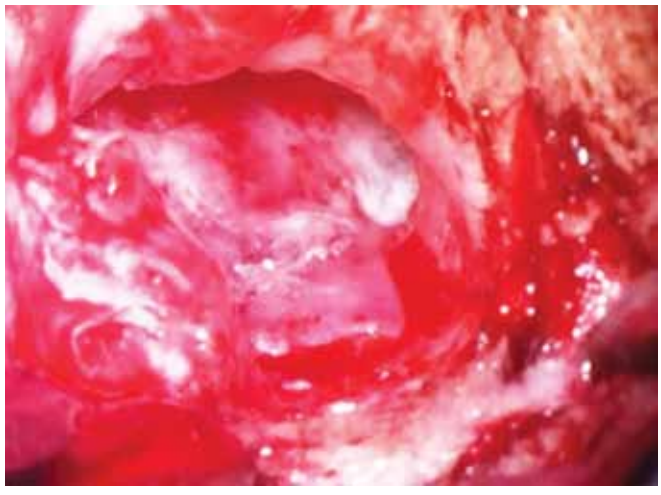

Fig. 8: The reposited tympanomeatal flap

on the saccular membrane (Fig. 5) (probably post-traumatic scarring). The incus was extremely unstable (probably from lack of support from the 'missing' malleus, and failed to retain position on attempting to reposit it anterolaterally (away from the Fallopian canal). It was thus decided not to perform any sort of ossicular reconstruction at a primary stage. The perilymph fistula at the exposed oval window was sealed with two layers of temporalis fascia (Fig. 6), after making the promontory surface around the oval window bare, by gently denuding the mucosa with a straight pick. Gelfoam was used to stabilize the oval window graft (Fig. 7), which was also tucked medial to the incus for support. The tympanomeatal flap was reposited (Fig. 8), and the endaural incision closed in layers. A mastoid bandage was given. The postoperative course was uneventful, and the bed rest with an elevated head position was continued for a week after surgery. The patient was then gradually mobilized into normal activity, with strict avoidance from any strenuous

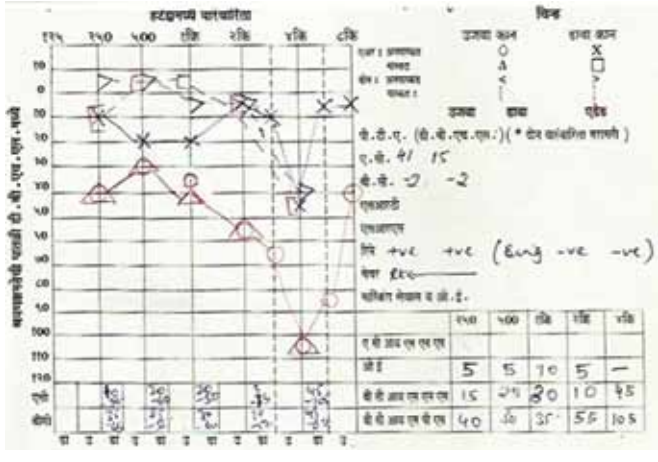

Fig. 9: The pure tone audiogram after 1 month of surgery, showing a normal bone conduction on the right side, with an air bone gap of about 40 decibels on an average

activities for a month after. He was also counseled about the findings during surgery, and the likeliness of a persistent hearing loss after surgery.

Outcome

Interestingly, on the follow-up visit after a month, the first thing that the patient mentioned is that the hearing in the right ear had improved, and there was no giddiness. The tinnitus was however present, but drastically reduced, and perceptible only when due efforts were made to notice it. The neotympanum appeared dull, but otherwise unremarkable. Surprisingly, the audiogram showed a marked improvement at 250 Hz, 500 Hz, 1 KHz and even 2 KHz, with a persistent dip at 4 KHz. The air-bone gap however remained consistent at 40 to 45 dB in most frequencies (Fig. 9). The patient was counseled to maintain the same postoperative lifestyle for a total period of 3 months, and asked to follow-up. A year later, on a routine follow-up visit, an audiogram revealed a 20 dB dip in the bone conduction at 1 KHz and 2 KHz, with the rest of the audiogram remaining the same as at 1 month postsurgery (Fig. 10). There was no perceptible change however in the hearing, and the patient was audiologically asymptomatic.

DISCUSSION

History

The case described above had a classical history at presentation, with all the symptoms of a perilymph fistula following direct trauma to the affected ear.[2]

Hearing Loss

The initial mixed hearing loss on the preoperative audiogram could be explained by the 'absent' malleus, and ossicular discontinuity contributing to the 'conductive' component and the fistula itself explaining the 'sensorineural' component. It must also be noted that the inability of the perilymph to be contained within the perilymphatic space also contributes to the 'conductive' component of the hearing loss.

The improvement following surgery was surprisingly better than expected. Given the fact that the malleus was missing, and the incus too was displaced medially, with no stapes (as it had to be removed), the postoperative residual conductive hearing loss only had an air-bone gap of about 40 dB on an average, which is far less than that seen in ossicular discontinuity. This is due to the fact that the oval window had sealed well with the fascial graft, which isolated it from the round window membrane, thus creating the phase difference/time lag for the sound presenting to the two windows, and compensating partially for the air conduction.

What was even more surprising was the dramatic improvement in bone conduction a month after surgery, to a normal level in the lower and mid frequencies, and a dip at 4 KHz (which was actually present in both ears, and may represent an earlier insult). Since, the only intervention was removal of the fractured stapes fragments from the oval window and placing a connective tissue seal over the latter, the improvement in bone conduction may be explained by the fact that the perilymphatic compartment was now a contained fluid filled space, which was earlier being kept 'stented' open by the stapes fragments; and this space was now conducting sound normally, with no leakage of fluid, which was affecting the 'middle ear mechanisms' of bone conduction (inertial and osseotympanic).

It is a well known fact that bone conduction can occur by three mechanisms, namely osseotympanic, inertial and compression mechanisms.[1] Of these, the first two can be affected by any pathology affecting the middle ear and tympanic membrane. The same may be considered to be contributory to the bone conduction, and thus even in middle ear pathologies affecting the inner ear and perilymphatic space, there is always the chance of some improvement in the bone conduction thresholds as can be seen from this example.

The sustained bone conduction in lower frequencies (seen in the 1 year postsurgery audiogram), with a slight dip in the mid-frequencies (although still within the normal range) may represent a stabilization of the sound conduction mechanism, or a recurrence of a perilymph fistula (at the same or another site). However, since there were no other symptoms at 1 year after surgery, it would be reasonable to assume the former explanation.

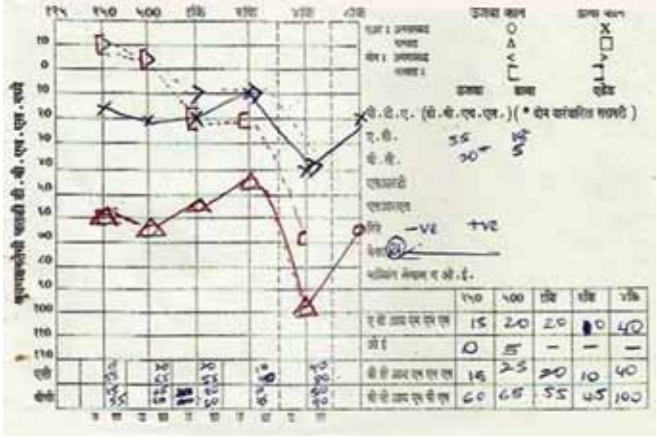

Fig. 10: The pure tone audiogram after 1 year of surgery, showing a slight decrease in the bone conduction in the mid frequency range

Surgical Findings

The 'missing malleus' cannot be explained. It was neither seen on CT scan, nor during surgery, and hence cannot be assumed to be dislocated. It may thus be hypothesized to

have got resorbed following trauma (which seems unlikely in the short time period between trauma and surgery), or may have been removed during the trauma itself (although there was no tympanic membrane perforation seen, there were numerous blood clots in the external auditory canal, which may have resulted from an initial perforation of the tympanic membrane with avulsion of the malleus out to the exterior). Although both the hypothesis seem unlikely, the latter would seem relatively more probable to have occurred.

Since, the malleus was missing, and the incus was highly unstable and dislocated, any primary ossicular reconstructive procedure would have not only been unsupported, but there was also an added risk of it subluxating into the 'empty' oval window. Thus, it seemed reasonable to defer the ossicular reconstruction during the primary surgery. Also, since after 1 year, the hearing loss remained for the most part essentially conductive, and the patient seemed satisfied with his hearing, no further intervention was done. It is true, however, that the patient would have to live with a lifelong potential weakness at the oval window.

CONCLUSION

Perilymph fistulae are to be considered when dealing with patients giving a history of trauma or increased central venous pressures, and presenting with a fluctuating hearing loss, which may be conductive, mixed or purely sensorineural in nature. A detailed history assumes prime importance, since the definitive confirmation of diagnosis is usually during exploratory surgery to diagnose and seal the fistula. Astute postoperative care and prevention of risk factors for an increased central venous pressure are vital in assuring a sustained successful outcome after surgery. Understanding the audiological changes that can possible occur in such circumstances, can help in predicting the eventual hearing outcome with a greater degree of certainty.

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