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TRAUMATIC PERILYMPHATIC FISTULA; A RARE ENTITY

ABSTRACT

Perilymph fistula is an abnormal communication between inner ear and middle ear cavity or mastoid causing perilymph leakage. The symptoms are vertigo, hearing loss, tinnitus, and aural fullness. Its management varies with variable outcome of hearing and vestibular function. A 32-year female with penetrating injury to the right ear causing dizziness and hearing loss presented with findings of bruise in external auditory canal along with dry central perforation and horizontal nystagmus in positional test. She had profound hearing loss and pneumolabyrinth. Successful surgical repair of perilymphatic fistula was performed with improved hearing and resolution of vestibular symptom. A high index of clinical suspicion is required for good outcome possible with early treatment. The surgical management with exploratory tympanotomy and fistula repair is advocated in patients with progressive hearing loss and non-resolving vertigo.

Keywords: Nystagmus, penetrating injury, perilymphatic fistula, tympanotomy, vertigo

INTRODUCTION

Perilymphatic fistula (PLF) is an abnormal communication between perilymph filled inner ear and air filled middle ear cavity causing perilymph leakage. The major causes are physical trauma (blunt and penetrating), barotrauma or iatrogenic. It is called idiopathic or spontaneous, when there are no identifiable prior events. Spontaneous ones are commonly predisposed by congenital anomalies or due to forgotten minor events like excessive nose blowing.[1] The common symptoms are vertigo, hearing loss, tinnitus, and aural fullness. The vestibular symptom is also described as of being "off balance" or disequilibrium.[2] The management of perilymphatic fistula varies with variable outcome of hearing and vestibular function. The author reports a case of traumatic perilymphatic fistula managed surgically with a good hearing outcome and resolution of vertigo.

CASE REPORT

A 32 year female was referred to ER from a rural place for suspected head injury. She gave history of an injury in right ear with a twig followed by fall to the ground causing loss of consciousness for around an hour. She had persistent dizziness along with vomiting and hearing loss in right ear. She also gave a history of intermittent right ear discharge for the last eight years. She underwent detailed ENT examination after head injury was ruled out. Otoscopy showed bruise in posterior-superior bony external auditory canal along with dry, subtotal central perforation involving both posterior and anterior quadrants. Tuning fork test showed severe to profound hearing loss in right ear and normal hearing in left ear. Spontaneous nystagmus was absent along with negative head impulse test and fistula test. Her cerebellar tests were normal, however, she swayed to right during Romberg and Fukuda tests. She was unable to do tandem gait test. She underwent positional tests which showed right beating horizontal nystagmus in right ear down position. The status of other cranial nerves along with facial nerve were normal. Pure tone audiometry showed 102 dB of profound hearing loss in right ear along with 22 dB of normal hearing in left ear according to the updated WHO 2021 classification of hearing loss.[3] Her temporal bone HRCT scan showed minimal air bubbles in the labyrinth along with soft tissue density in stapes area with doubtful stapes rotation as seen in Figure 1.

Figure 1: The temporal bone HRCT scan axial view showing

minimal pneumolabyrinth (thin black arrow) along with soft tissue density in stapes area (thick black arrow) in right side (top image). Sagittal section showing soft tissue density in right side (bottom left image) as compared to normal stapes in left side (bottom right image).

The patient was initially managed conservatively with vestibular sedatives, steroids and prophylactic antibiotics. She then underwent endoscopic exploratory tympanotomy under general anesthesia due to lack of improvement in vestibular and cochlear symptoms. The intraoperative findings were 80% central perforation along with dislocation of stapedio-vestibular joint. The stapes was tilted inferiorly and posteriorly along with disruption of incudo-stapedial joint. (Figure 2)

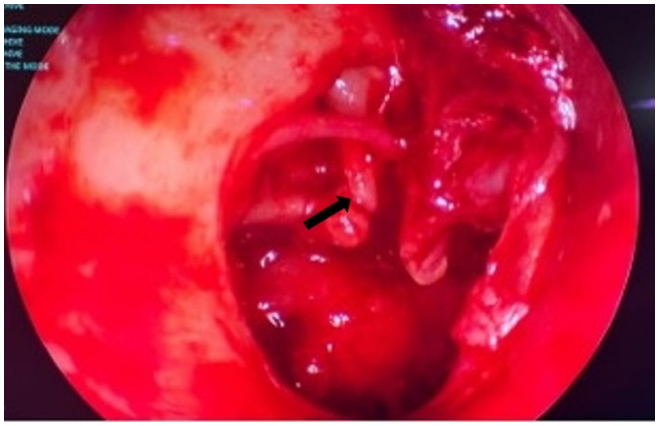

Figure 2: Endoscopic image showing postero-inferior tilting of stapes along with disruption of incudo-stapedial joint (thick black arrow).

The stapes was repositioned and covered with temporalis fascia along with realignment of incudostapedial joint. (Fig. 3) The tympanic membrane perforation was repaired with temporalis fascia. The patient was put on strict bed rest for 24 hours under stapedotomy protocol.

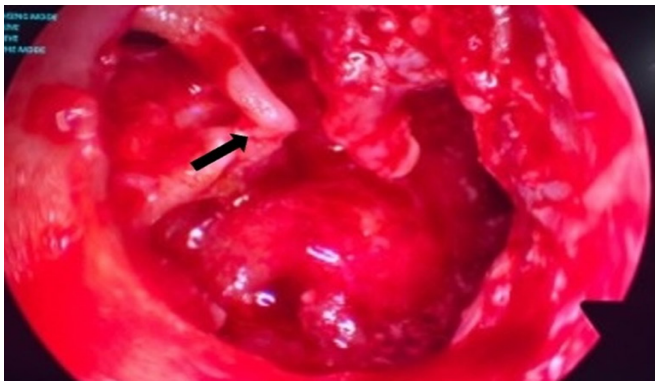

Figure 3: Endoscopic image showing repositioning of stapes along with realignment of incudo-stapedial joint (thick black arrow).

The post-operative period was uneventful with immediate improvement in vestibular symptoms. On third month of surgery, she had intact tympanic membrane graft along with negative positional tests. Her pure tone audiogram showed 55 dB of moderate mixed hearing loss in right ear along with 7 dB of normal hearing in left ear.

DISCUSSION

The perilymphatic fistula is defined as a leak of perilymph at round or oval window and excludes other conditions of bony canal dehiscence causing third window syndrome.[1,4] The mechanism of PLF was classically described as to be either due to implosive or explosive forces by Goodhill et al.[5] It was first recognized as a complication of stapes mobilization and stapedectomy surgery.[1] Spontaneous perilymphatic fistula, as described in literature, accounts for around 24 to 51% of cases.[6]

Traumatic perilymphatic fistula due to penetrating trauma of inner ear should be identified early to resolve cochleo-vestibular symptoms and to prevent permanent loss of labyrinthine functions. It is critical for primary care and ENT clinicians to have high index of suspicion for early diagnosis and timely treatment. The presence of vestibular symptoms and disproportionate cochlear symptoms and mode of injury were highly suggestive of penetrating inner ear trauma in discussed case even though patient had preexisting central perforation of tympanic membrane due to chronic otitis media.

The presence of air bubbles in labyrinth known as pneumolabyrinth was suspected in this case along with other positive findings of inner ear trauma. Pneumolabyrinth accounts for the unusual clinical features of the paroxysmal positional vertigo. The mechanism is explained by a pneumopositional theory by Edries et al, in which position induced vertigo and nystagmus is attributed to be caused by the movement of air bubbles within the semicircular canal causing cupular deflection and ampullary stimulation.[7] Positional nystagmus present in this case is also explained by this theory. Positional nystagmus is described to be of multidirectional, longer duration and of smaller amplitude in the literature.[8] Pneumolabyrinth are almost always associated with perilymphatic fistula. However, contrary is not true especially in cases of delayed imaging.[4] It is observed in 83.3% of cases of PLF.[9] Temporal bone HRCT is sensitive in detecting pneumolabyrinth when present. However HRCT is not pre-requisite for diagnosis of PLF. High index of suspicion based on vestibular and hearing symptoms along with history is important for diagnosis.[9,10] There are no identifiable predictors for PLF described in literatures.[10] Portman grading system was described in 2016 indicating high probability of fistula with score of more than 7.[11] Similarly, diagnostic criteria for PLF is also described in literature.[12]

There are variation in choice of treatment of perilymphatic fistula among clinicians. Some literature suggest no difference between conservative and surgical management

in terms of improvement in cochleo-vestibular symptoms.[13] However, surgical management with sealing the abnormal communication between inner ear and middle ear is generally considered effective to improve symptoms.[1,6,14] Surgical treatment is described to be more effective in reducing vestibular symptoms more than the cochlear symptoms.[6] Similar outcome of complete resolution of vestibular symptoms with moderate improvement in hearing was observed in this patient as well. Surgical treatment is beneficial to avoid further deterioration of hearing loss and to prevent permanent hearing loss in cases of PLF due to identifiable cause.[6] Early surgical treatments is also advocated in case of PLF due to barotrauma.[15] On the contrary, in some cases of PLF with stable hearing with resolving vestibular symptoms, management with conservative treatment can be adopted.[9]

CONCLUSION

The early diagnosis of PLF requires high index of suspicion based on history and clinical features. Pneumolabyrinth is a diagnostic sign of inner ear trauma. The surgical management with exploratory tympanotomy and fistula repair is advocated in patients with progressive hearing loss and non-resolving vertigo.

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