

Pneumolabyrinth Associated with Perilymph Fistula

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Pneumolabyrinth associated with perilymph fistula, especially with the presence of air in the cochlea, has rarely been identified using imaging study. We present a 24-year-old woman who experienced right fluctuating sensorineural hearing impairment and dis-equilibrium after sustaining an open-handed slap on the right side of her face. The diagnosis of pneumolabyrinth was established through the detection of air bubbles in the right basal turn of cochlea using high-resolution computerized tomography. Right side exploratory tympanotomy confirmed the presence of a perilymph fistula. The patient was free of symptoms after surgery. Early fistula repair was beneficial in this case. (*Chang Gung Med J* 2003;26:690-4)

Key words: perilymph fistula, pneumolabyrinth.

A perilymph fistula (PLF) is defined as an abnormal opening between the inner ear and the external surface of the labyrinth capsule.⁽¹⁾ The leakage of perilymph may cause fluctuating sensorineural hearing loss and dis-equilibrium. Using high resolution computerized tomography (HRCT) to detect an air bubble in the labyrinth, Mafee et al.⁽²⁾ first described this rare image finding as pneumolabyrinth.

PLF can be a result of barotraumas, stapedectomy, head injury, or chronic ear disease.⁽³⁾ However, given a clear history, preoperative diagnosis of PLF remains a challenge in most cases using results of direct clinical examination. The widely-used fistula test and Tullio phenomenon both have low sensitivity rates⁽⁴⁾ and the presenting symptoms of a PLF are not specific from other otologic conditions. The presence of pneumolabyrinth can support the diagnosis of a PLF.

CASE REPORT

We present a 24-year-old woman who sustained an open-handed slap on the right side of her face, causing a small elliptic perforation at posterior infe-

rior quadrant of the right eardrum (Fig. 1). Her presenting symptoms of fluctuating sensorineural hearing loss on the right and dis-equilibrium, together with a positive fistula test, were consistent with clinical manifestations of PLF.⁽⁵⁾ Her pure tone audiometry (PTA) on the damaged side showed a mixed-type hearing impairment with bone conduction fluctuating



Fig. 1 A small elliptic perforation at posterior inferior quadrant of the right eardrum.

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change (Fig. 2). The diagnosis was established through the detection of air bubbles in the basal turn of the right cochlea using HRCT (Fig. 3). Right exploratory tympanotomy performed 3 days after the trauma revealed that clear fluid was emanating from

the round window niche. The niche was repaired using tragal perichondrium and bolstered with Gelfoam®. The patient was free of whirling sensation symptoms right after surgery and her PTA returned to normal (Fig. 4).

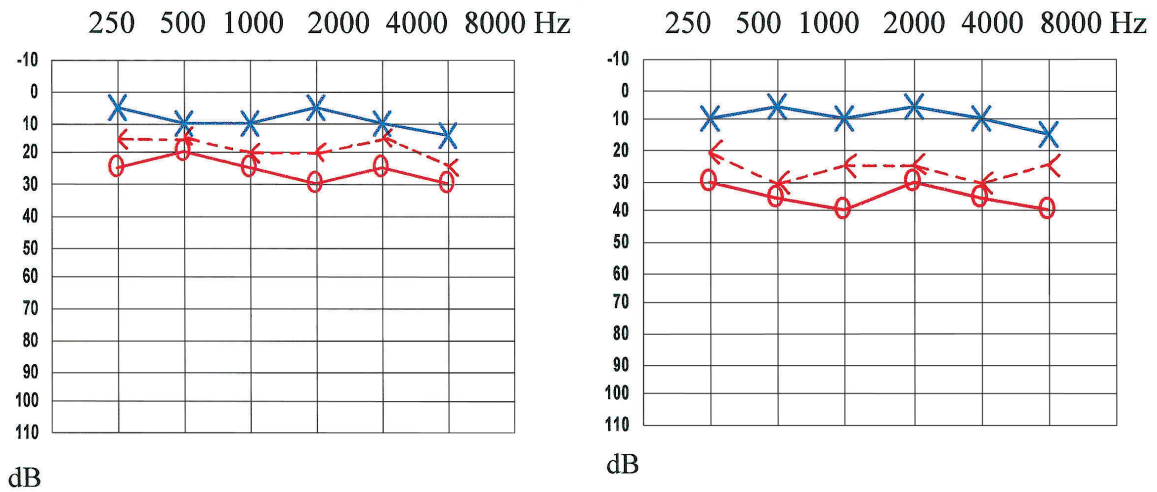


Fig. 2 Left: Pure tone audiometry, 3 days before the surgery, shows right mixed type hearing impairment with air-bone gap 7 dB. The bone conduction is at 18 dB level. Right: Pure tone audiometry, the day before the surgery, shows right mixed type hearing impairment with air-bone gap 8 dB. The bone conduction is at 27 dB level.

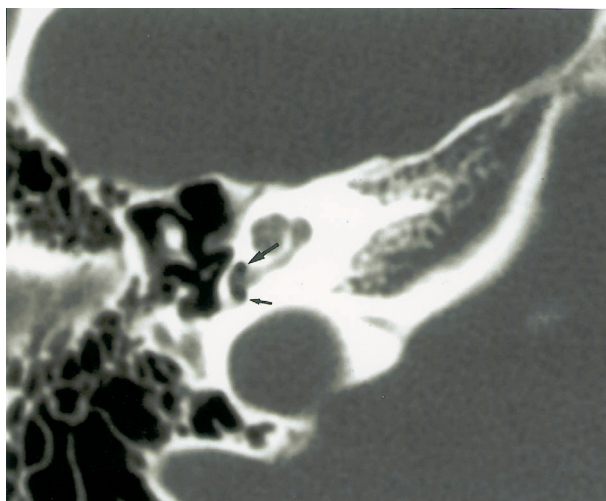


Fig. 3 Axial high resolution CT (1mm thickness, window width 4000, center 300) of the right cochlea shows a hypodense area in the non-dependent portion of the basal turn (large arrow), with the Housefield unit -630, and the Housefield unit of round window niche (small arrow) is -650.

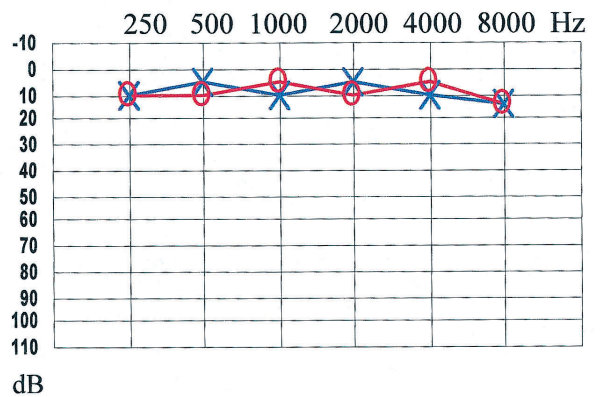


Fig. 4 Pure tone audiometry, 2 weeks after the surgery. There is no air-bone gap. The bone conduction is at 8 dB level.

DISCUSSION

Pneumolabyrinth resulting from PLF has been associated with barotraumas, stapedectomy, head trauma, and chronic ear disease.⁽³⁾ Goodhill⁽⁶⁾ proposed implosive and explosive forces as potential causes of PLF. Implosive forces resulting in PLF are those that cause pressure changes in the middle ear space and thus exert an external force on the oval and round window membranes, fissula ante fenestram, or Hyrtl's fissure.⁽⁷⁾ These forces can be triggered by barometric pressure changes, compression trauma of the ear, Valsava maneuver, or pinch-nose sneezing.⁽⁶⁾ Explosive forces are those that exert an internal pressure by increasing cerebrospinal fluid pressure. These forces can be raised by straining, lifting, coughing and sneezing.⁽⁶⁾ PLF at the round window membrane due to scuba diving-associated barotrauma was originally reported by Pullen.⁽⁸⁾ Emmet and Shea⁽⁹⁾ reported a 64% incidence of PLF associated with traumatic tympanic membrane perforation. Trauma to the cochlear could be attributed to the presence of air bubbles in the scala tympani or scala media that disturb propagation of the traveling wave of basilar membrane. Endolymphatic hydrops may develop due to low perilymph pressure and high endolymphatic pressure. Leakage of perilymph may induce relatively high endolymphatic pressure to agitate the membranous labyrinth causing dizziness or unsteadiness.⁽¹⁰⁾

The presenting symptoms of PLF are not specific and the PLF is hard to prove preoperatively. By applying positive pressure to the tympanic membrane to induce nystagmus, a positive fistula test is suggestive of the presence of PLF. Fox et al.⁽⁴⁾ reported 30 out of 39 surgically confirmed PLF had positive preoperative fistula test. The Tullio phenomenon is considered positive if a patient experienced vertigo or loss of balance after exposure to a 95 dB tone burst at 500 Hz for less than 3 seconds. Fox et al.⁽⁴⁾ reported that six of seven surgically confirmed PLF had positive preoperative results to the Tullio phenomenon. Middle ear exploration may reveal fluid emanating from the oval or round window niche during operation. However, serum or anesthetic agent may be mistaken for perilymph or an intermittent leak may be missed during surgery. β 2-transferrin intra-theal injection has recently been used to indicate the leakage site intra-operatively.⁽¹⁰⁾

Electrocochleography (ECoG) may show an increased summing potential/action potential ratio in about 50% of patients with PLFs.⁽¹¹⁾ Meyerhoff and Ridenour⁽¹¹⁾ found that ECoG for PLF had a sensitivity and specificity of 60% and 52%, respectively.

Pneumolabyrinth associated with PLF, especially with the presence of air in the cochlea, has rarely been identified using imaging study. Magnetic resonance imaging is not an appropriate method for detecting pneumolabyrinth because it is difficult to distinguish air from the otic cortical bone adjacent to the air, since neither air nor cortical bone produces a signal on magnetic resonance imaging.⁽¹²⁾ The introduction of HRCT with thin slices (1.0-1.5-mm thickness) has made it possible to delineate air in the inner ear. As head injuries and stapedectomies are major causes of pneumolabyrinth, HRCT is also able to show traumatic fractures of the otic capsule. Disruption of the membranous labyrinth complicated with fractures can facilitate movement of air in the inner ear space. Air can move within the inner ear space in response to changes in head position in which HRCT is performed.⁽¹²⁾

Given a clear history of head trauma, the presenting symptoms of PLF can still be vague in most cases using results of direct clinical examination.⁽⁵⁾ Since this patient had clear antecedent history and presented with symptoms including fluctuating sensori-neural hearing impairment and dis-equilibrium, exploratory tympanotomy should be considered.⁽¹³⁾ In addition, preoperative HRCT finding of air in the basal turn of the cochlea is a reliable sign of PLF and justifies surgical exploration for the closure of fistula. Since the morbidity such as profound deafness and risk of meningitis from perilymph leakage are potentially detrimental,⁽¹⁴⁾ early repair of the fistula was beneficial in this case.

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外淋巴瘻管合併氣迷路

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因外淋巴瘻管而起的氣迷路，尤其是耳蝸內有空氣存在，在影像學上的表徵相當少見。我們報告一例24歲女性病人，於右臉遭掌摑後造成耳膜破裂，出現波動式神經性聽力障礙以及不平衡感。經安排顳骨高解析度電腦斷層掃描，於耳蝸底迴見到低密度影像的氣泡存在，因而確立外淋巴瘻管以及氣迷路的診斷，中耳探查手術於圓窗處發現外淋巴洩漏，更進一步肯定外淋巴瘻管的存在。病人經手術修補外淋巴瘻管後，症狀立即得到解除，並避免了可能引發全聾或腦膜炎的後遺症。耳蝸內有空氣存在是影像學上罕見的表現，特此提出報告。(長庚醫誌 2003;26:690-4)

關鍵字：外淋巴瘻管，氣迷路。

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