



Case Report

Conductive Hearing Loss as the Initial Manifestation of Spontaneous Herniation of Epitympanic Meningocele

Chao-Yin Kuo, Chih-Hung Wang, Hui-Ming Feng, Hsin-Chien Chen

Department of Otolaryngology-Head and Neck Surgery, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan, ROC

Although the incidence of tegmen tympani dehiscence is relatively high, diagnosis can be difficult if there are no clinical clues or a high degree of suspicion. We describe the case of a 79-year-old male patient with progressive left-sided hearing impairment and ear ringing for 1 year who was eventually diagnosed with middle ear meningocele. Audiometry showed conductive hearing loss with an air-bone gap of 20 dB. This was caused by impaired mobility of the ossicular chain due to meningocele herniation from the dehiscence of the tegmen tympani. In the absence of typical symptoms, such as cerebrospinal fluid middle ear effusion and otorrhoea, diagnosis of tegmen tympani dehiscence may be challenging. Consequently, tegmen tympani dehiscence should be included in the differential diagnosis of conductive hearing loss, which may be the only or early clinical manifestation of tegmen tympani dehiscence. High clinical suspicion and fully assessment are necessary to prevent misdiagnosis and further serious complications.

KEY WORDS: Tegmen tympani, dehiscence, meningocele, conductive hearing loss

INTRODUCTION

The tegmen tympani is a thin bony plate that separates the cranial and tympanic cavities. Dehiscence of the tegmen tympani may be accompanied by herniation of the meninges or brain (meningocele or meningoencephalocele). The findings from an autopsy study suggest that 15% to 34% of specimens contain a single defect in the tegmen of the temporal bone ^[1].

The most common symptoms of tegmen tympani dehiscence (TTD) are cerebrospinal fluid (CSF) effusion, CSF otorrhoea, serous otitis media, CSF rhinorrhoea, conductive hearing loss (CHL), meningitis, aural pain, headache, epilepsy, and other neurological complications. However, these common symptoms may not manifest in all cases of TTD.

Herein, we report the case of a patient with atypical symptoms of TTD who had meningocele herniation causing air-bone gap hearing loss. The unique manifestations and symptoms of TTD in this case are described and discussed.

CASE REPORT

A 79-year-old man experienced progressive left-sided hearing impairment and ear ringing for 1 year. He had a history of hypertension, for which he was receiving medical treatment. He had no history of chronic otitis media, head trauma, or cranial surgery. Otoloscopic examination revealed an intact eardrum, with no evidence of inflammation or fluid accumulation in the bilateral middle ear. There was, however, a protruding lateral process of the malleus and anterolateral displacement of the chorda tympani nerve in the left ear (Figure 1). Audiography revealed mild sensorineural hearing loss with left-sided 20 dB air-bone gap hearing loss (Figure 2). Tympanography revealed bilateral normal eardrum compliance. High-resolution computed tomography of the temporal bone revealed a dehiscence of the left tegmen tympani with compression of the medial aspect of the malleus (Figure 3). Subsequent magnetic resonance imaging of the brain showed a small accumulation of CSF over the corresponding region, without brain tissue herniation (Figure 4). The imaging findings were consistent with a diagnosis of TTD with meningocele formation. The patient chose to receive conservative treatment. There was no CSF effusion in the middle ear or neurological symptoms during follow-up.

DISCUSSION

TTD can be congenital or acquired following chronic otitis media with or without cholesteatoma, iatrogenic trauma, or neoplasm. Diagnosis of TTD is crucial for preventing potential life-threatening central nervous system (CNS) infection and permanent neurological defect ^[2,3]. Associated neurological complications such as meningitis, encephalitis, or otogenic cerebral abscess may occur due to the formation of a pathological pathway between the subarachnoid space and middle ear cavity or direct exposure of dura or cerebral tissue ^[3]. Epilepsy may also occur as a result of abnormal electrical activity caused by herniated cerebral tissue or CNS infection.

Corresponding Address:

Hsin-Chien Chen, Department of Otolaryngology-Head and Neck Surgery, Tri-Service General Hospital, National Defense Medical Center, 325, Sec. 2, Cheng-Kung Rd, Taipei 114, Taiwan, ROC. Phone: 886-2-87927192; Fax: 886-2-87927193; E-mail: acolufreia@yahoo.com.tw

Submitted: 27.08.2013 Revision Received: 06.01.2014 Accepted: 08.01.2014

Copyright 2014 © The Mediterranean Society of Otolaryngology and Audiology

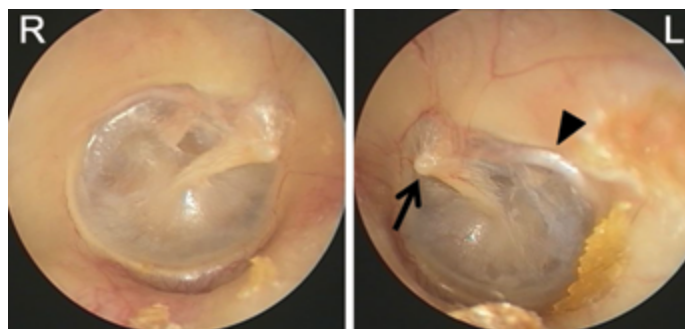


Figure 1. Otoscopic examination revealed a laterally projected process of the malleus (arrow) and anterior and lateral displacement of the chorda tympani nerve (arrow head) in the left ear (L) relative to that in the right ear (R)

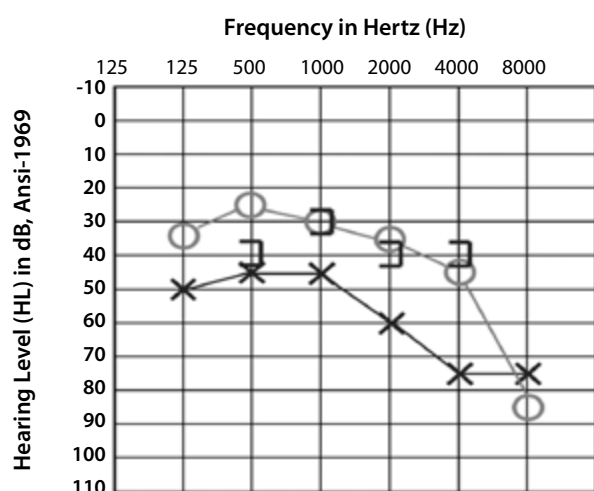


Figure 2. Audiogram demonstrating mixed type hearing loss with an air-bone gap of 20 dB in the left ear

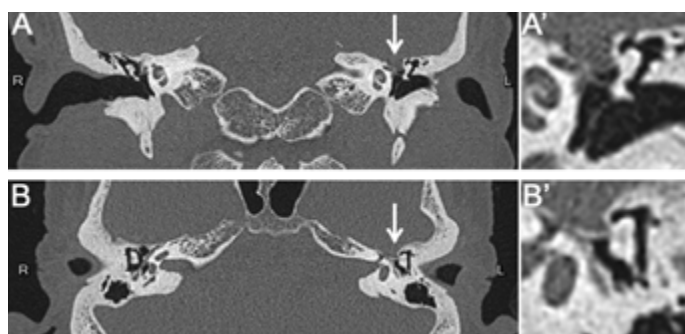


Figure 3. High-resolution computed tomography of the temporal bone revealed dehiscence in the left tegmen tympani, with soft tissue occupying the epitympanum and adjacent to the medial aspect of the malleus in the coronal (A, arrow and A', enlarged in A) and axial (B, arrow and B', enlarged in B) views

The diagnosis of TTD with spontaneous CSF middle ear effusion (MEE) or otorrhoea depends on a high degree of clinical suspicion; however, there is no consensus in the literature on the optimal approach^[1]. Nahas et al.^[4] described 15 cases of TTD, 13 of which involved middle ear CSF effusion. Most of the patients were diagnosed with TTD because of continuous clear otorrhoea after ventilation tube insertion. Some authors have suggested that persistent clear otorrhoea or serous otitis media in patients over the age of 50 may be indicative of TTD^[1].

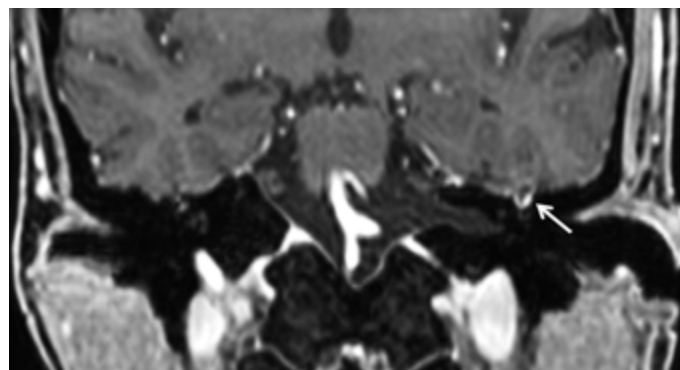


Figure 4. Post-gadolinium coronal T1-weighted magnetic resonance imaging of the brain revealed a herniation from the dehiscence of the tegmen tympani (arrow). The herniation mainly comprised dura (the contrast enhanced part) and some cerebrospinal fluid (the non-contrast enhanced part), but no brain tissue

The typical otoscopic findings in meningoencephalic herniation are a pulsatile mass lesion behind the ear drum or clear fluid effusion in the middle ear^[3]. However, the mass lesion may also present as non-pulsatile and be of variable colour, including red, pink, or white. In our case, the patient did not have MEE or clear otorrhoea, but did have tinnitus and CHL. On otoscopic examination, the lateral process of the malleus was protruding compared with that in the opposite one. This may have been due to the meningocele forcing the malleus towards the lateral side and causing anterolateral displacement of the chorda tympani nerve. Golding-Wood et al.^[2] have suggested that tegmental dehiscence may contribute to CHL through herniation of the temporal lobe, reducing ossicular mobility or disrupting the chain. CHL due to TTD and brain herniation without otitis media is uncommon; therefore, TTD should be included in the differentiated diagnosis of air-bone gap hearing loss.

Surgical repair is suggested for patients with CSF leak, CSF otorrhoea, or CSF accumulation in the middle ear to prevent repeat meningitis and further neurological complications^[3]. Numerous surgical approaches involving grafting the bony defect have been described and found to be highly successful for the management of TTD with CNS or otological problems. Gubbles et al.^[5] reported the use of a fragment of calvarial bone to reconstruct the tegmen tympani, with bone cement applied on the irregular surface and a muscle or fascia overlay to reconstruct the floor of the middle cranial fossa. Similarly, Kenning et al.^[6] reported the use of autologous bone, pericranium, and intradural collagen grafts. Semaan et al.^[7] suggested creating an epidural pocket with a cartilage graft locked in place, and a fascia graft overlying the defect. Few reports have described surgical approaches for the treatment of small meningocele without significant complications. As a consequence, it has been suggested that surgical intervention may not be required in asymptomatic patients^[8].

A number of factors have been reported to be predictive and/or predisposing for tegmen dehiscence complicated with CSF MEE or otorrhoea. Prichard et al.^[9] noted that five of seven patients with spontaneous CSF otorrhoea had an empty or partially empty sella, that is, an increased pressure in the sella turcica caused the pituitary gland to flatten out along the interior walls of the sella turcica cavity. Consequently, the authors have suggested that if spontaneous CSF otorrhoea is evident or TTD is diagnosed, it is important to rule out in-

creased intracranial pressure^[9]. In another report, Hadi et al.^[10] noted that 13 of 23 patients with TTD had semicircular canal dehiscence. Among these patients, protrusions of the superior semicircular canal in the middle cranial fossa were significant and considered to be an additional aetiological factor. Hence, semicircular canal dehiscence should be investigated when TTD is present.

In conclusion, conductive hearing loss is typically the result of blockade of the external auditory canal, sclerosis or dislocation of the ossicular chain, otitis media, or eustachian tube dysfunction. TTD with spontaneous meningocele as an initial and sole cause of CHL is rare; however, we suggest that TTD should be included in the differential diagnosis of CHL. Interestingly, protrusion of the lateral process of the malleus in the eardrum and anterolateral displacement of the chorda tympani nerve may be early manifestations of TTD. Imaging investigation is necessary to rule out this clinically rare entity, facilitate the timely diagnosis of spontaneous meningocele of the temporal bone, and thus prevent potential complications.

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

Peer-review: Externally peer-reviewed.

Author Contributions: Supervision - C.H.W.; Materials - C.Y.K., H.M.F.; Data Collection and/or Processing - C.Y.K., H.C.C.; Writing - C.Y.K., H.C.C.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Brown NE, Grundfast KM, Jabre A, Megerian CA, O'Malley BW Jr, Rosenberg SI. Diagnosis and management of spontaneous cerebrospinal fluid-middle ear effusion and otorrhea. *Laryngoscope* 2004; 114: 800-5. [\[CrossRef\]](#)
2. Golding-Wood DG, Williams HO, Brookes GB. Tegmental dehiscence and brain herniation into the middle ear cleft. *J Laryngol Otol* 1991; 105: 477-80. [\[CrossRef\]](#)
3. Sanna M, Fois P, Russo A, Falcioni M. Management of meningoencephalic herniation of the temporal bone: Personal experience and literature review. *Laryngoscope* 2009; 119: 1579-85. [\[CrossRef\]](#)
4. Nahas Z, Tatlipinar A, Limb CJ, Francis HW. Spontaneous meningoencephalocele of the temporal bone: clinical spectrum and presentation. *Arch Otolaryngol Head Neck Surg* 2008; 134: 509-18. [\[CrossRef\]](#)
5. Gubbels SP, Selden NR, Delashaw JB Jr, McMenomey SO. Spontaneous middle fossa encephalocele and cerebrospinal fluid leakage: diagnosis and management. *Otol Neurotol* 2007; 28: 1131-9. [\[CrossRef\]](#)
6. Kenning TJ, Willcox TO, Artz GJ, Schiffmacher P, Farrell CJ, Evans JJ. Surgical management of temporal meningoencephaloceles, cerebrospinal fluid leaks, and intracranial hypertension: treatment paradigm and outcomes. *Neurosurg Focus* 2012; 32: E6. [\[CrossRef\]](#)
7. Semaan MT, Gilpin DA, Hsu DP, Wasman JK, Megerian CA. Transmastoid extradural-intracranial approach for repair of transtemporal meningoencephalocele: a review of 31 consecutive cases. *Laryngoscope* 2011; 121: 1765-72. [\[CrossRef\]](#)
8. Tsutsumi K, Asano T, Shigeno T, Matsui T, Ito S, Kaizu H. Transcranial approach for transsphenoidal encephalocele: report of two cases. *Surg Neurol* 1999; 51: 252-7. [\[CrossRef\]](#)
9. Prichard CN, Isaacson B, Oghalai JS, Coker NJ, Vrabec JT. Adult spontaneous CSF otorrhea: correlation with radiographic empty sella. *Otol Head Neck Surg* 2006; 134: 767-71. [\[CrossRef\]](#)
10. El Hadi T, Sorrentino T, Calmels MN, Fraysse B, Deguine O, Marx M. Spontaneous tegmen defect and semicircular canal dehiscence: same etiopathogenic entity? *Otol Neurotol* 2012; 33: 591-5. [\[CrossRef\]](#)