The Journal of Laryngology & Otology
November 2000, Vol. **114,** pp. 861–863

Clinical Records

Bilateral defects of the tegmen tympani associated with brain and dural prolapse in a patient with pulsatile tinnitus

S. U. Kale, M.S. (Orl), DLO (Lon), F.R.C.S., A. G. Pfleiderer, F.R.C.S., J. C. Cradwick, F.R.C.R.*

Abstract
A pulsating ear drum is a rare otological finding and usually indicates a vascular pathology. We report the case of a woman who presented with pulsatile tinnitus following an upper respiratory tract infection. She was found to have bilateral pulsating tympanic membranes on otological examination, in association with bilateral dehiscence of the tegmen tympani and a spontaneous encephalocele on the right side and a meningocele on the left side, demonstrated on computerized tomography (CT) scan of her temporal bones.

Key words: Meningocele; Encephalocele; Tinnitus

Case report

A 55-year-old woman presented with a six months' history of a pulsating noise in her ears, which was worse on the left and was preceded by an upper respiratory tract infection. She also gave history of a long-standing bilateral hearing loss. There were no other otological symptoms or history of head injury or ear surgery. There were no neurological or ocular symptoms. She was on Atenolol for recently diagnosed mild hypertension.

Examination of the ears revealed bilateral atrophic tympanic membranes that moved in time with her pulse rate but not with respiration. The tinnitus was not abolished by pressure on her internal jugular veins. The ear findings were confirmed by tympanometry which showed a pulsatile trace in both ears. The trace on the right side (Figure 1) had a larger amplitude than on the left. Her audiogram revealed a bilateral low tone mixed deafness of 30 dB and 60 dB in the right and left ears respectively. The possibility of vascular aetiology was considered and a CT scan of the temporal bones was performed that revealed bilateral symmetrical bony defects in the roof of the epitympanum with dural prolapse into the middle ear (Figures 2 and 3). On the right side the dura was in contact with the ossicles and hence the tympanogram on the right was of a larger amplitude. The internal carotid artery and the jugular bulb were anatomically normal. In view of the absence of any serious underlying pathology, she was referred to the hearing therapist for tinnitus counselling, that is helping her adapt to her tinnitus.

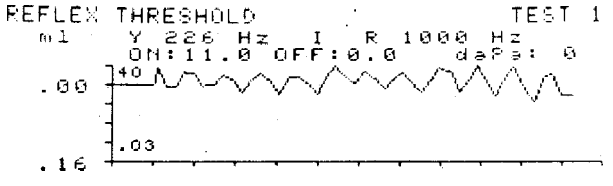


Fig. 1
Pulsatile trace on tympanometry in the right ear.

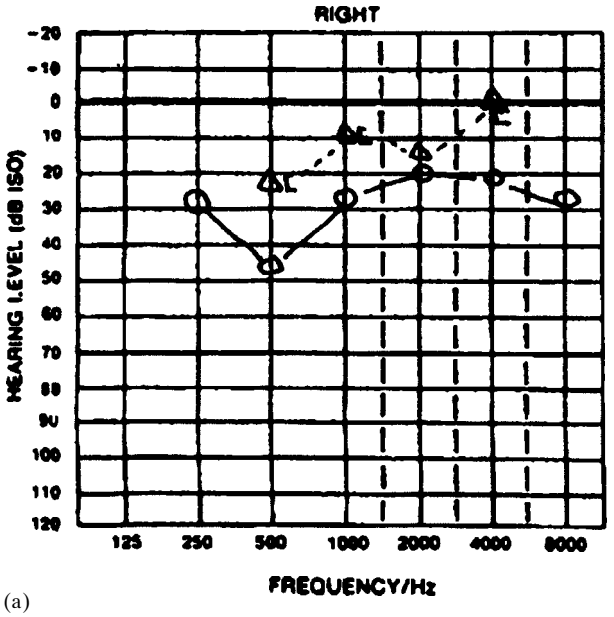
Discussion

Pulsating ear drums are a rare otological finding that in the absence of any vascular lesion appear in our patient to be due to transmitted dural pulsations arising from bilateral congenital meningoceles.

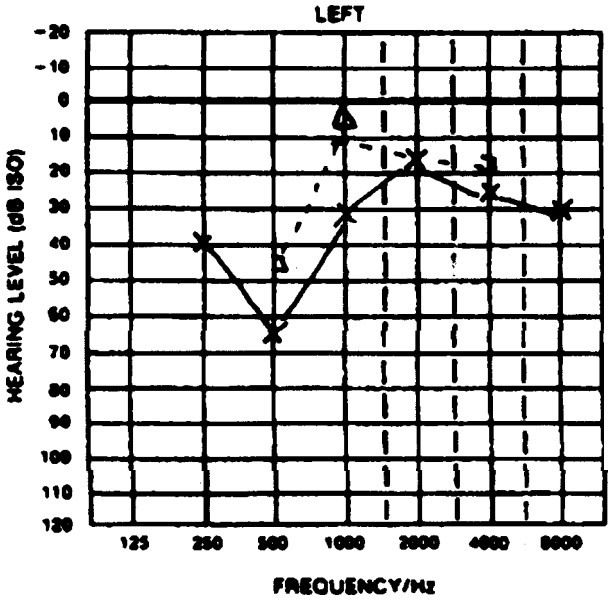
A meningocele is defined as a protrusion of meninges through a defect in the skull or the vertebral column and encephaloceles are the presence of meninges, cerebrospinal fluid and/or brain extending beyond the cranial cavity through a bony defect in the skull. Temporal bone defects resulting in a meningocele can present with a conductive hearing loss, persistent middle-ear effusion, pulsatile tinnitus and persistent otorrhoea. Pulsatile tinnitus is usually caused by direct contact of the mass with the ossicular chain,[1] as was demonstrated by CT in our patient. There may also be a dural defect in addition, that allows herniation of brain tissue and these can present with episodes of epilepsy, recurrent meningitis and cerebrospinal fluid (CSF) otorrhoea.[2] However a review of the otolaryngological literature has not revealed a case of a meningocele or an encephalocele presenting with a pulsating ear drum.

Meningoceles or encephaloceles arising from defects in the tegmen can occur as a result of previous surgery, trauma, infection, or may be spontaneous.[3] Other disease processes that can cause bone erosion such as cholesteatoma, chronic otitis media, granulomatous disease and

From the Department of Otolaryngology, Edith Cavell Hospital, Peterborough and the Department of Radiology*, Peterborough District Hospital, Peterborough, UK.
Accepted for publication: 22 June 2000.



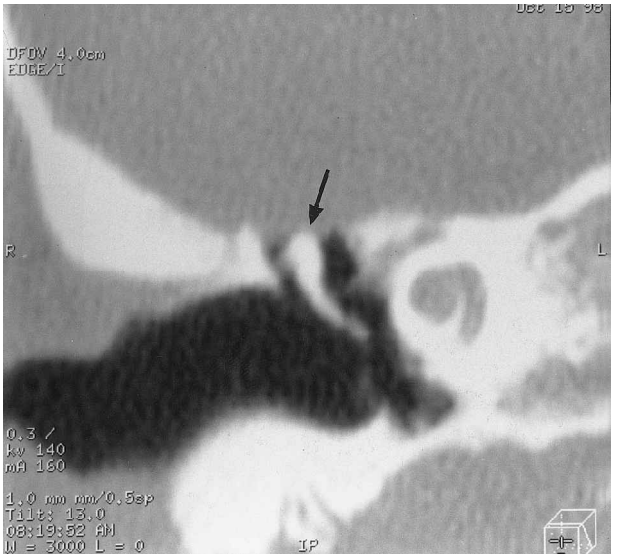
(a)



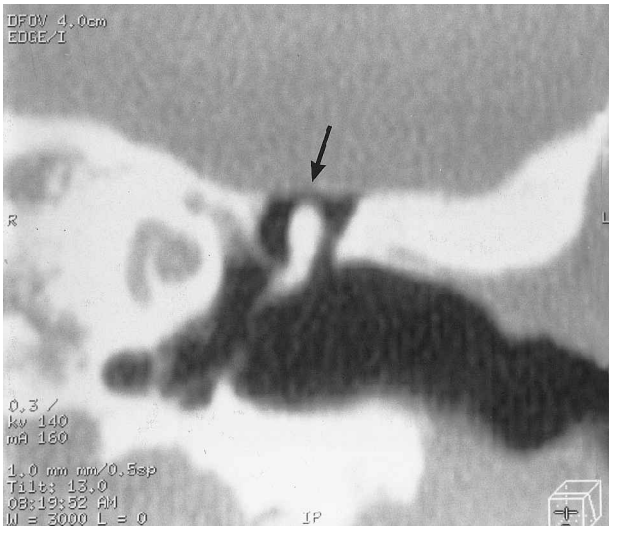
(b)

FIG. 2

Pure tone audiograms of the right (a) and left (b) ears.



(a)



(b)

FIG. 3

(a) and (b) CT scan showing bony defects in the epitympanum with an encephalocele in the right ear and dural prolapse in the left ear.

cholesterol granuloma[4] should also be considered but were excluded in the case presented in the absence of any clinical signs, and as the bony defects on CT were smooth and symmetrical in appearance they are more likely to represent congenital defects in the temporal bone.[5] This is demonstrated in an autopsy study of 50 temporal bones from 25 cadavers, of which nine bones had congenital type defects in the tegmen tympani and one had a defect in the tegmen antri. Three subjects had bilateral defects of the tegmen. None of them had any dural defects.[6]

Pulsatile tinnitus is an uncommon symptom, but may indicate a serious pathology. It is almost always the result of the sound of non-laminar blood flow and can occur in local disorders which are anatomically close to, or within the petrous bone.[7] The causes of pulsatile tinnitus can be vascular or nonvascular. Vascular causes include anomalies of the jugular bulb and intrapetrous carotid artery, arteriovenous fistula, atherosclerosis, aneurysms and fibromuscular dysplasia of the internal carotid artery and vascular loops and paragangliomas of the temporal bone. Meningoceles of the skull base[1] are amongst the non-vascular causes that also include benign intracranial hypertension.[8] Hypertensive patients on enalapril and verapamil can also present with pulsatile tinnitus, that disappears after cessation of the medication.[8] The most frequent cause of pulsatile tinnitus varies according to the series reported. In a series of 145 patients reported by Sismanis,[9] benign intracranial hypertension was the most common diagnosis, while in another study of 84 patients, the most common causes were dural ateriovenous fistula, and glomus tumours.[7] Nevertheless, since pulsatile tinnitus can be a symptom of serious pathology, the authors of the above studies recommend non-invasive investigations such as a CT scan, which led to the discovery of the underlying meningocele and encephalocele in our case, followed if necessary by magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA).

https://doi.org/10.1258/0022215001904176 Published online by Cambridge University Press

The occurrence of the pulsatile tinnitus, in the presence of an abnormality which appears to be long-standing, is explained by the term 'conductive tinnitus'. This type of tinnitus is associated with, exacerbated by, or just possibly created by, a middle-ear disorder and/or a conductive hearing loss. The conductive hearing loss causes a reduction of the ambient noise which causes enhancement or revealing of the tinnitus,[10] Hallam *et al.*,[11] introduced the idea that a process of habituation occurs, which involves not only the auditory system but also the attention and arousal systems. Habituation to tinnitus is normal and tinnitus distress occurs when this process is interrupted. Hence we postulate that our patient probably had pulsatile tinnitus to which she was habituated, but was brought to her awareness after the upper respiratory tract infection.

Conclusion

In summary, meningoceles and encephaloceles should be considered in the differential diagnosis of a pulsating ear drum, in addition to other vascular causes. We present a case of bilateral defects of the tegmen tympani of the middle ear that were diagnosed because the patient presented with pulsatile tinnitus, the investigation of which led to the discovery of the underlying encephalocele on the right and meningocele on the left.

Acknowledgements

I would like to thank the medical photography department at Peterborough District Hospital for supplying photographs for the case report.

References

1 Mulchay MM, McMenomey SO, Talbot JM, Delashaw JB. Congenital encephalocele of the medial skull base. *Laryngoscope* 1997;**107**:910–4
2 Williams DC. Encephalocele of the middle ear. *J Laryngol Otol* 1986;**100**:471–3
3 Gray BG, Willinsky RA, Rutka JA, Tator CH. Spontaneous meningocele, a rare middle ear mass. *Am J Neuroradiol* 1995;**16**:203–7
4 Gottlieb MBC, Blaugrund JE, Niparko JK. Imaging Quiz Case 1, Tegmental encephalocoele. *Arch Otolaryngol Head Neck Surg* 1998;**124**:1274–7
5 Ferguson BJ, Wilkins RH, Hudson W, Farmer J. Spontaneous CSF otorrhoea from tegmen and posterior fossa defects. *Laryngoscope* 1986;**96**:635–44
6 Kapur TR, Bangash W. Tegmental and petromastoid defects in the temporal bone. *J Laryngol Otol* 1986;**100**:1129–32
7 Waldvogel D, Mattle HP, Sturzenegger M, Schroth G. Pulsatile tinnitus – a review of 84 patients. *J Neurol* 1998;**245**:137–42
8 Sismanis A, Smoker WRK. Pulsatile tinnitus: Recent advances in diagnosis. *Laryngoscope* 1994;**104**:681–8
9 Sismanis A. Pulsatile tinnitus, a 15 year experience. *Am J Otol* 1998;**19**:472–7
10 Coles RRA. Tinnitus. In: Kerr AG, ed. *Scott Brown's Otolaryngology*. 6th edn. London, Oxford: Butterworth-Heinemann, 1997;**2**:2/18/5
11 Hallam RS, Rachman S, Hinchcliffe R. Psychological aspects of tinnitus. *Contributions to Medical Psychology* 1984;**3**:31–53

Address for correspondence:
Mr A. G. Pfleiderer, F.R.C.S.,
Department of Otolaryngology,
Edith Cavell Hospital,
Bretton Gate,
Bretton,
Peterborough, PE3 9GZ,
UK.

Mrs S Kale takes responsibility for the integrity of the content of the paper.
Competing interests: None declared