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Co-existing cholesteatoma and vestibular schwannoma

Biodun Ogungbo, F.R.C.S., Damian Holliman, Ph.D., A. David Mendelow, F.R.C.S., John Hill, F.R.C.S.*

Abstract

A 69-year-old man presented with a cholesteatoma in the right mastoid process and a vestibular schwannoma at the left internal acoustic meatus. Cholesteatoma co-existing with a vestibular schwannoma has not been documented previously in the contemporary literature. The clinical dilemma in the management of his progressive bilateral hearing loss is discussed.

He presented with dizziness and bilateral hearing loss worse on the right side. Pressure over the mastoid process elicited vertigo and nystagmus. He had no history of previous operation or infection in the ear canal. Audiograms confirmed high-tone hearing loss. Radiological investigations revealed a symptomatic cholesteatoma on the right side and an incidental vestibular schwannoma on the left. We have elected to manage both lesions conservatively.

Bilateral cholesteatoma and bilateral vestibular schwannomas have been previously reported. Co-existing lesions, as in our patient have, however, not been reported previously. The management options of his hearing loss are discussed.

Key words: Cholesteatoma; Congenital; Hearing Loss, Sensorineural; Vestibular Schwannoma

Introduction

Cholesteatomas originate from ectodermal cell rests that arise after incomplete separation of neural ectoderm from cutaneous ectoderm during neural tube closure at embryonic weeks three to five. Usually affecting the temporal bone, the most common sites are the petrous temporal bone, the cerebellopontine angle, the mastoid and the middle ear.[2] Vestibular schwannoma is the most common tumour of the cerebellopontine angle.[3] Bilateral cholesteatoma and bilateral vestibular schwannomas have been reported previously. However, a patient with a cholesteatoma on one side and a contralateral vestibular schwannoma has to our knowledge not been previously described. Management decision in bilateral vestibular schwannomas, especially in patients with neurofibromatosis II, is difficult and we contend that our patient presents a similar problem.

Case report

We report the case of a 69-year-old retired, right-handed electrician who presented with episodic vertigo of five months duration. Pressing on the right external ear canal could precipitate this. He did not complain of hearing loss, headaches, nausea or vomiting. He had no history of ear infection or a previous otological procedure. He suffered from gout and smoked 15 cigarettes a day. He had no relevant family history apart from maternal longevity (his mother died at 98 years). Examination revealed a reddish lesion behind an intact right tympanic membrane and a positive fistula sign indicated a direct communication between the right middle-ear space and the right labyrinth. The left tympanic membrane and middle ear appeared normal. No cerebellar signs or focal neurological deficits were elicited. Audiogram confirmed a mild bilateral high tone sensorineural hearing loss (Figure 1).

Radiology

Magnetic resonance imaging (MRI) and MR cerebral angiography

Axial proton density, T1-, T2-weighted spin echo of the whole brain, axial and coronal STIR of the posterior fossa along with MRI arteriogram and venogram of the skull base vessels were performed. These revealed a large ($4.5 \times 3 \times 2.5$ cm) lobulated lesion within the petrous and occipital bones on the right side and replacing most of the right mastoid. It involved the jugular foramen, extending predominantly superiorly and posteriorly, obliterating the right internal jugular vein and displacing the right internal carotid forward. The left jugular venous sinus was patent. There was slight displacement of the right cerebellar hemisphere. No significant vascular supply to the lesion was demonstrated. It returned high signal on the STIR image with some internal septations. Post-gadolinium, there was very little enhancement of the periphery of the lesion and the septa. On the left, a second abnormality was seen in the internal acoustic meatus extending into the cerebellopontine angle. It enhanced avidly and measured $2\times 2 \times 0.8$ cm (Figures 2(a) and (b)).

Computed tomography

Computed tomography (CT) scans demonstrated erosion of the right temporal bone, the posterior wall of which was completely destroyed. The mastoid air cells were well

From the Department of Neurosurgery, Regional Neurosciences Directorate, Newcastle General Hospital, and the Department of Otolaryngology*, Freeman Hospital, Newcastle-upon-Tyne, UK.
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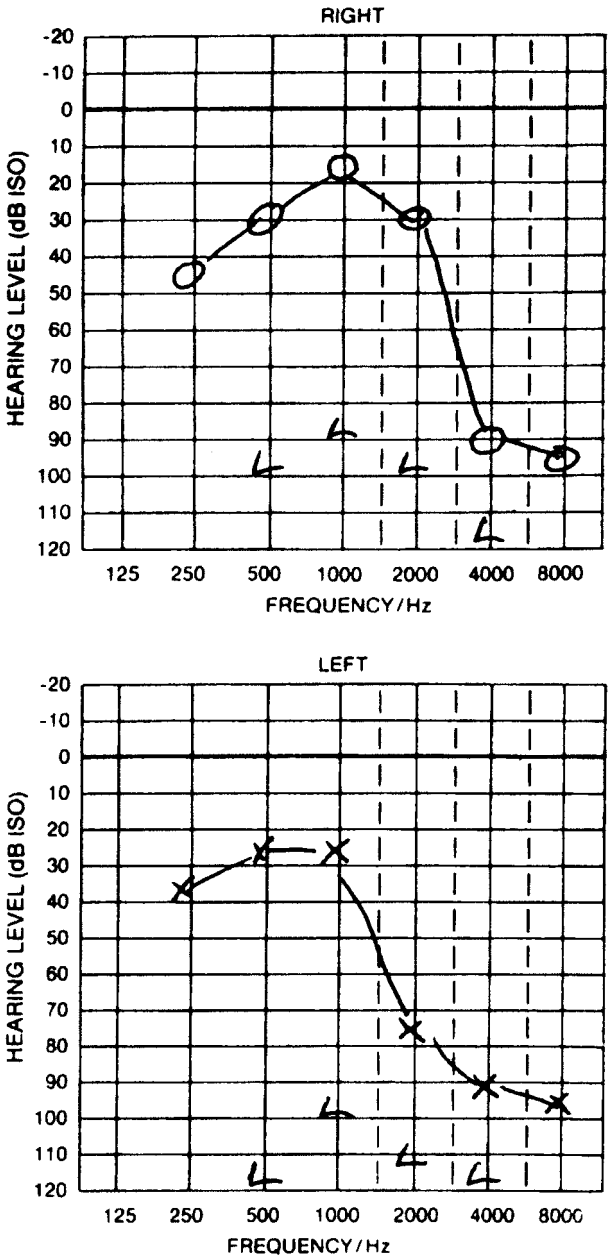
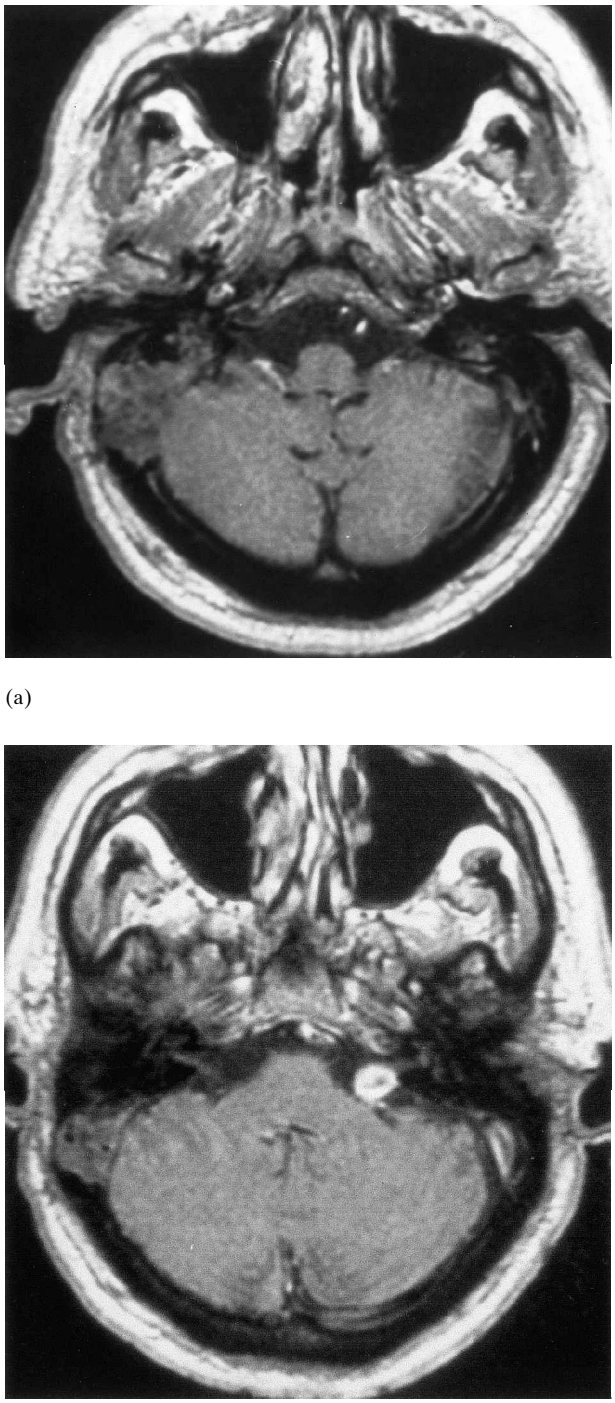


FIG. 1

Audiogram of the patient showing bilateral high tone sensorineural deafness.



(a)

(b)

FIG. 2

(a) (b) MRI scan of the patient revealed the right-sided cystic cholesteatoma and the enhancing vestibular schwannoma in the left cerebellopontine angle.

pneumatized and openly eroded, with extension into the occipital bone (Figure 3).

Radiological diagnosis

The appearances were suggestive of a left vestibular schwannoma and a right cystic lesion. The differential diagnosis for this lesion was cystic schwannoma, a chordoma, glomus tumour or metastasis. Meningioma and paraganglioma were less likely possibilities.

Management

An open biopsy of the right-sided mastoid lesion was undertaken for histological diagnosis. The operation was performed through a small retro-auricular incision over the soft area of bony destruction as shown in the CT scan (Figure 3). It revealed acellular keratin deposits with no inflammatory cell reaction or lining epithelium suggestive of cholesteatoma. This is possibly congenital. Since initial presentation we have managed him conservatively with no change in his hearing subjectively on either side. At routine follow-up in the clinic 18 months later he remained well but his hearing had deteriorated slightly bilaterally. Follow-up MR scan appearances of both lesions were unchanged. We intend to continue with regular interval MRI scans and audiogram assessment.

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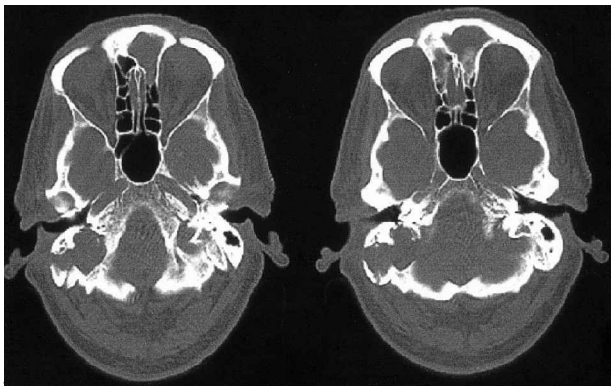

FIG. 3

Un-enhanced CT scan demonstrating significant bony erosion and defects in the right mastoid bone.

Discussion

Cholesteatomas can be congenital or acquired. Congenital cholesteatoma is the third commonest lesion found in the cerebellopontine angle. They are thought to be the cause of two to 24 per cent of all cholesteatomas, that have an annual incidence of approximately 12.6 per 100 000.[1] It originates at various sites in the petrous bone, most commonly in the petrous apex, and less often in the mastoid process.[4,5] It must be differentiated from vestibular schwannomas, meningiomas, metastatic tumours, arachnoid cysts and lipomas. These tumours can be associated with congenital malformations sometimes involving the auditory apparatus.[6] MRI and CT scans are complementary in the diagnosis. CT is useful in the visualization of the bony architecture and the relationships with the labyrinthine structures. MRI allows evaluation of intra-cranial extension of the tumours, which are of low or intermediate signal intensity on T1-weighted images and high intensity on T2-weighted images.[7]

Bilateral congenital cholesteatomas have been reported previously.[8] Surgical excision has been the management of choice in most cases. However, congenital cholesteatoma co-existing with a vestibular schwannoma is distinctly rare and we have not found a similar case report in the contemporary literature.

Management of cholesteatomas

Congenital cholesteatomas have the propensity for slow asymptomatic growth. The mean age of presentation is approximately five years of age.[1] Early discovery in childhood is essential for complete surgical removal with little morbidity,[9] resulting in 91 per cent retention of functional hearing. The case is perhaps less well-defined in the adult where the incidence of ossicular destruction and deafness ranges from 40–50 per cent following surgery.[1,10] Complete removal of congenital cholesteatomas is advocated as the treatment of choice.[1,11] Satisfactory hearing preservation has been reported in canal wall up and canal wall down procedures for cholesteatomas.[12] Canal wall down procedures had the advantage of lower residual disease. Residual disease has however, been reported in 30–57 per cent of cases,[9,10] suggesting that these may require re-operation and, consequently additional morbidity. Our patient is a fit 68-year-old man with no significant past medical history. His mother lived until the age of 98 years. It was felt that as the radiological evidence suggested a large fistula into his inner ear on the right side the risks of sensorineural hearing loss following surgical intervention were high. Others may wish to operate on this lesion and arguments advanced relate to the potentially progressive damage to his inner ear function by the cholesteatoma. However, not operating presents the best chance of preserving his hearing for as long as possible.

Management of vestibular schwannoma

The success rate for hearing preservation surgery in unilateral and bilateral vestibular schwannoma is variable and considerably less than 50 per cent in many series (with a sizeable patient population and satisfactory follow-up periods). This deteriorated to 24 per cent in 136 attempts at long-term follow-up review.[13] The exhaustive review demonstrated a wide array of criteria and results in hearing preservation surgery.[13] There is debate as to what constitutes 'useful' and 'serviceable' hearing.

Our proposed plan

Our patient has functional hearing at the present time in both ears, slightly worse on the left. Hearing preservation has a less than 30 per cent chance of success in his case. Tumour growth is slow in the elderly, averaging 0.2 cm/year or less in 80 per cent of patients.[3,14] No growth or worsening of auditory acuity has been seen in up to 38 per cent of patients.[15] This has led to the advocating of conservative management in elderly patients with small tumours.[3] Both traditional microsurgical techniques and stereotactic radio surgery have rates of successful preservation of cochlear function of around 50 per cent.[13,16] Therefore, it was felt that the risks of causing hearing loss in the left ear using either technique were considered too great and of no value in the management of this case. A future consideration may be the use of a cochlear implant after surgery on either lesion.

Conclusion

We have reported the case of a 69-year-old man with a possible congenital cholesteatoma co-existing with a vestibular schwannoma. Our management strategy is outlined.

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Address for correspondence:
A. David Mendelow, F.R.C.S.,
Department of Neurosurgery,
Regional Neurosciences Directorate,
Newcastle General Hospital,
Newcastle-upon-Tyne NE4 6BE, UK.

Fax: 0191 2563267

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