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Unusual lesions of the internal auditory canal

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Abstract
Acoustic neuromata (AN) account for nearly 90 per cent of internal auditory canal (IAC) and cerebello-pontine angle (CPA) tumours. The second most common tumour is meningioma. Rare lesions include primary cholesteatoma, facial neuroma, lipoma, angioma and various cysts.

Two cases of IAC tumour are presented, one of hamartoma in which smooth muscle was prominent and the other of lymphangioma. Of interest are the specific clinical and radiological features associated with these lesions.

Key words: Cerebello pontine angle, neoplasms; Vestibular schwannoma

Introduction

In patients with the symptom complex of dizziness, disequilibrium, tinnitus and unilateral hearing loss, the most common underlying lesion is vestibular schwannoma, acoustic neuroma (AN) in 91.3 per cent followed by meningioma in 3.1 per cent (Brackmann and Bartles, 1980). Other rare lesions are primary cholesteatoma (2.4 per cent) (Brackmann and Bartles, 1980), facial neuroma, lipoma, angioma and cysts (Kinney *et al.*, 1997; Kohan *et al.*, 1997; Babin *et al.*, 1980).

The diagnosis of tumours of the IAC and CPA has been greatly facilitated by improvement in imaging techniques such as computerized tomography (CT) and magnetic resonance imaging (MRI) (Fukui *et al.*, 1995). Rare lesions of the IAC and CPA mimic the clinical symptoms and signs of acoustic tumour. Correct diagnosis prior to surgery, probably cannot be made, even with correct imaging.

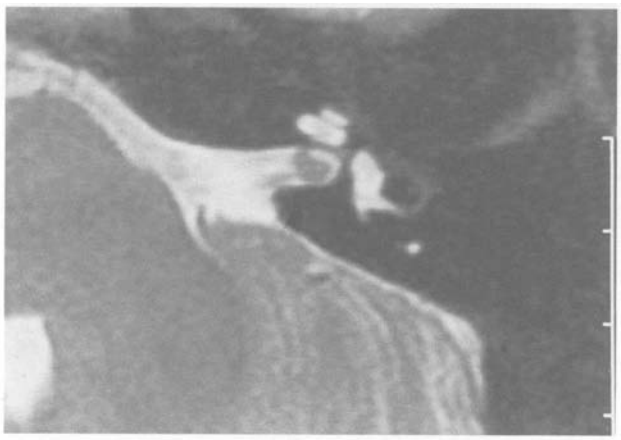

FIG. 1

Case 1 – Fast spin echo T_2 (TR 5,400/TE 90) 3 mm axial slice showing rounded 3 mm low intensity mass in the lateral end of the IAC. This is the appearance that an acoustic tumour would show.

Case reports

Case 1

A 15-year-old male presented with progressive left-sided hearing loss and loss of balance. An initial audiogram showed a flat sensorineural hearing loss of some 30–40 dB in the left ear. Evoked response was normal and electronystagmography (ENG) showed a left canal paresis. MRI scan showed a 3 mm lesion in the lateral aspect of the left IAC. The lesion was hypointense on T_2 weighted image (Figure 1), isointense with grey matter on T_1 and markedly hyperintense in T_1 with gadolinium (Figure 2). No atypical features could be seen in retrospect. These

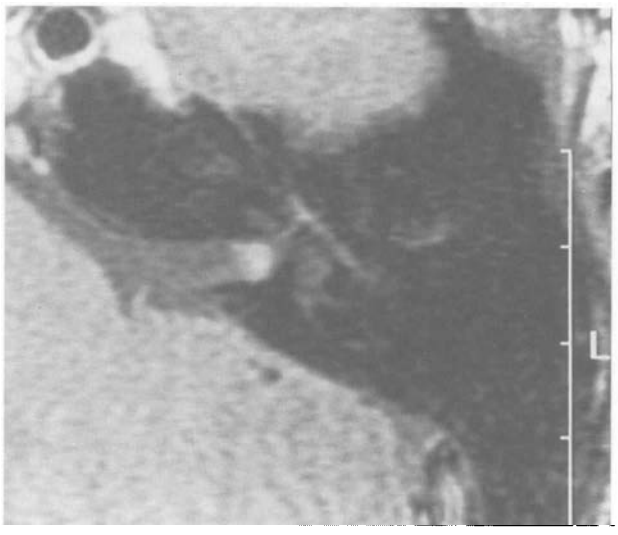

FIG. 2

Case 1 – Spin echo T_1 (TR 480/TE 20) 3 mm axial slice post i.v. Gadolinium showing enhancement at the lateral end of IAC. This slice is slightly superior to Figure 1. These appearances mimic those of an acoustic tumour.

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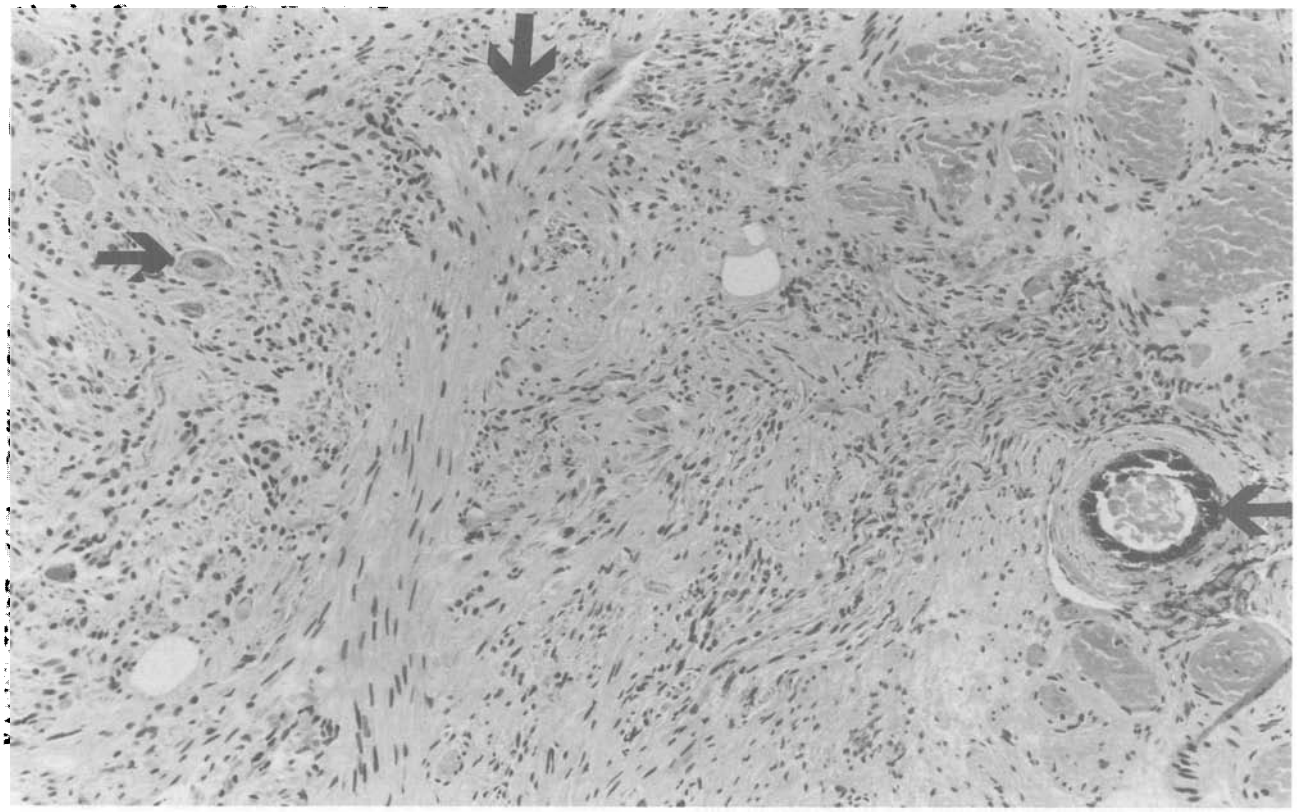

FIG. 3

Case 1 – Section of cochlear nerve hamartoma of smooth muscle, showing disorganized nerve bundle with ganglion cells (arrow to right). A cluster of vessels with thick muscular walls is present at the right, one of which has a calcified wall (arrow to left). A bundle of smooth muscle cells crosses the field (upper arrow) (H & E; × 10).

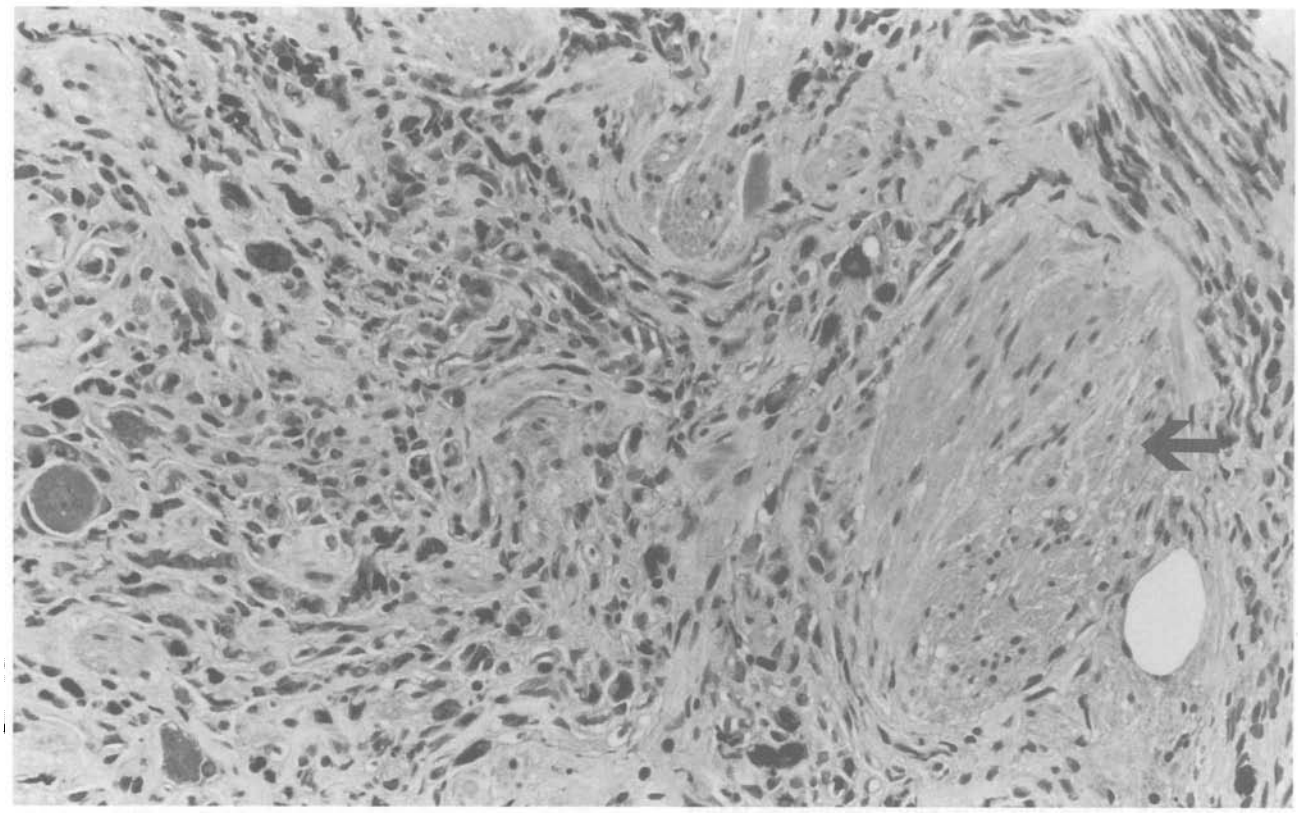

FIG. 4

Case 1 – Section stained for neurofilaments showing dark staining of ganglion cells and axones. Bundles of smooth muscle cells (arrow), were unstained with this antibody, but were positive with an antibody against desmin. (Immunoperoxidase, × 20).

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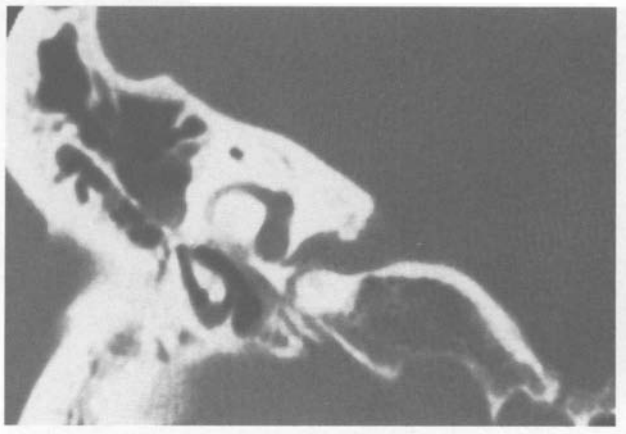

FIG. 5

Case 2 – Axial CT without contrast showing bony irregularity (not expansion) of the posterior wall of IAC. The bone changes are those of bone projection in the canal and adjacent bony erosion.

radiological findings are consistent with acoustic neuroma. Over the ensuing 28 days all hearing in the affected ear was lost. At operation, carried out by the translabyrinthine approach, the eighth nerve was found to be infiltrated with tumour in fusiform fashion. Tumour removal was judged to be complete and facial function was preserved in toto.

Histological examination revealed a lesion which consisted of smooth muscle, vessels and disordered nerve fibres (Figures 3 and 4) with only a very small adipose tissue component. No skeletal muscle was present. The lesion was judged to be a hamartoma.

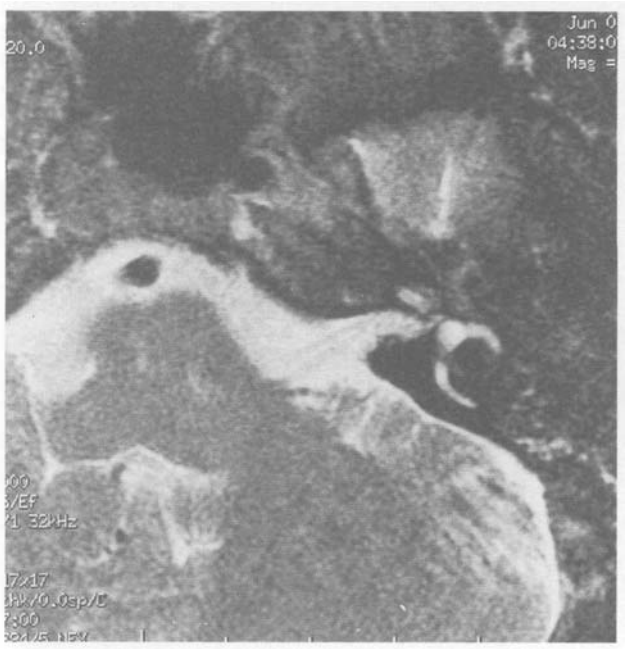

FIG. 6

Case 2 – Fast spin echo axial T_2 (TR 5,400/TE 90) showing displacement of nerves anteriorly with hyperintense poorly defined mass in lateral half of IAC. This, particularly when combined with the bony changes seen on CT, makes it very unlikely that the lesion will prove to be an acoustic. T_1 images after Gadolinium showed marked enhancement.

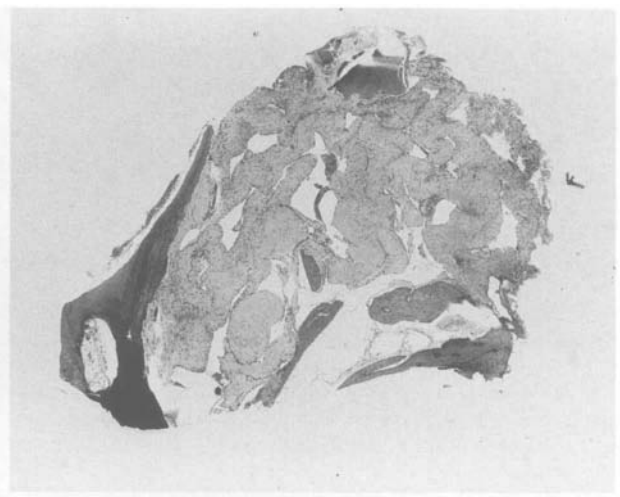

FIG. 7

Case 2 – The angioma in this case is composed of a collection of large vascular spaces with thick fibrous walls. Bony trabeculae are seen within the mass. No blood is seen, suggesting that this is a lymphangioma rather than haemangioma (H & E; × 2).

Case 2

A 44-year-old lady presented two years after a successful tympanoplasty. For some six months there had been a steady loss of hearing in the operated ear and unsteadiness, which began one year after surgery, was a major feature. The grafted tympanic membrane was found to be intact. A high resolution 3D CT study showed bone projections in the posterior wall of the IAC and adjacent bony erosion. No IAC expansion was noticed as usually seen in IAC tumours (Figure 5). MR showed a poorly defined mass in the lateral IAC that was not well seen on a limited T_1 series, was hyperintense on 3-mm FSL T_2 and 0.7 3D FSE T_2 sequences (Figure 6). This mass displaced the nerves anteriorly and enhanced strongly following i.v. gadolinium. These CT and MR studies showed an IAC mass that was highly atypical of an acoustic tumour.

At surgery, carried out by the translabyrinthine approach, a lesion was encountered in the IAC, arising from the dura of the posterior wall of the canal but this was separate from the cranial nerves. The vestibular and cochlear nerve was divided and resected along with the tumour and facial function remained normal.

Histopathology revealed a lesion consisting of large vascular spaces lined by endothelium. Most of the vascular spaces were empty, leading to a diagnosis of cavernous lymphangioma rather than haemangioma. Some ossification was noted within the lumina of the vessels (Figure 7).

Discussion

Most lesions of the IAC and CPA are acoustic tumour and meningioma. Others are rare (Brackmann, 1980; Lalwani, 1992; Hung *et al.*, 1997).

There are few reports in the literature of smooth muscle hamartoma and indeed none could be found in the IAC. Fatty hamartoma can be diagnosed prior to surgery by MR and CT (Reid *et al.*, 1991) but given the similarities in the imaging studies between those found in acoustic neuroma and the present study, it is unlikely that the correct diagnosis could have been made prior to surgery.

Similarly, lymphangioma of the IAC or CPA is fairly rare and has not been reported in the English literature, although haemangioma is more common (Kinney *et al.*, 1997). The latter are often found associated with the facial

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nerve. The changes noted on MR were hypointense on T_1 and hyperintense on T_2. However, these lesions were not characteristic of any common tumour in this area.

Little is known about the behaviour of these benign tumours. Each had shown a steady progression of symptoms and complete destruction of hearing. Given that the likely course of such lesions is steady growth, early excision is recommended. In these cases, even though each lesion was very small, there was no hearing to be saved and the translabyrinthine approach was the method of choice.

Conclusion

Although correct imaging can differentiate fatty hamartomas of the IAC from acoustic tumours, a smooth muscle hamartoma cannot be diagnosed before surgery. However, in cases of angiomatous lesions of the IAC, it is possible to state that the lesion is not an acoustic tumour but it is difficult to be more dogmatic.

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