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Clinical Records

Cholesteatoma: an unusual presentation

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Abstract
A case is described of an extensive cholesteatoma presenting as a lump on the side of a patient's head overlying the squamous temporal bone, with erosion of the underlying bone and intracranial extension. The patient was otherwise asymptomatic. This case highlights the bone-eroding capacity of cholesteatoma and the vigilance required in assessing lumps on the head.

Key words: Cholesteatoma; Temporal Bone

Introduction

The various modes of presentation of cholesteatoma are something an otorhinolaryngologist becomes familiar with early in their career. The scenario of a patient with an offensive discharging ear, progressive conductive hearing loss and keratin accumulating within an attic or pars tensa defect is all too frequently seen in out-patients' departments across the country. Occasionally a patient may present with a complication resulting from a cholesteatoma, due to the disease extending, either intracranially or extracranially.

We describe a case of a cholesteatoma presenting as a lump on the lateral surface of the temporal bone with no aural symptoms nor signs.

Case report

A 71-year-old man, who had undergone a right modified radical mastoidectomy 50 years previously, presented to the out-patients' department with a history of a swelling above and around his right ear that was increasing in size and becoming painful. He had been aware of the swelling

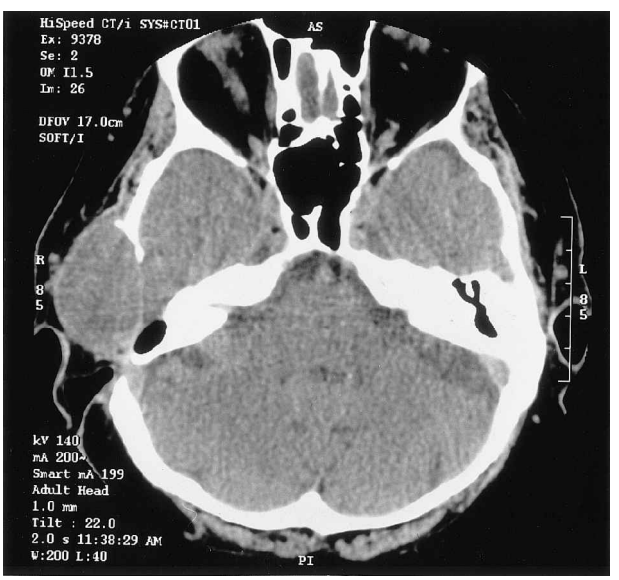

Fig. 1(a)

Axial computed tomography scan demonstrating the large soft tissue mass with the associated bony defect, the anterior margins of which are scalloped. Posteriorly, the mass has eroded the petrous temporal bone and anterior wall of the lateral sinus.

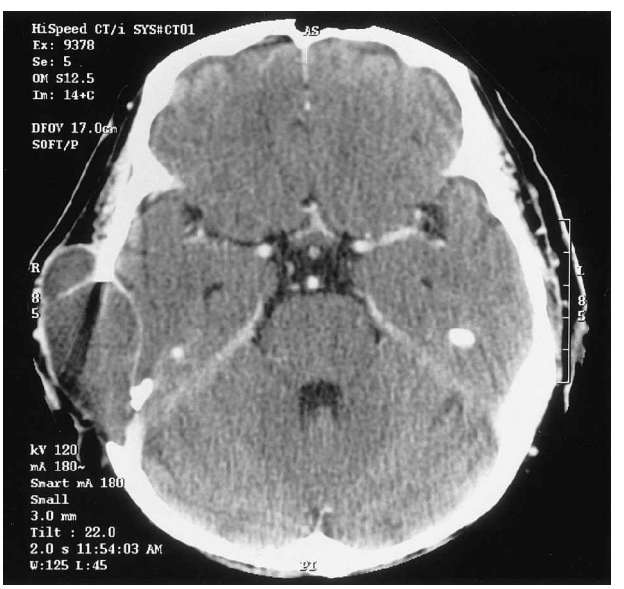

Fig. 1(b)

Post-contrast axial computed tomography scan demonstrating the multiloculated mass which has eroded the inner and outer table of the cortex and displaced the soft tissues of the scalp laterally.

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for nine months, but conceded it was likely to have been present for much longer. Apart from the recent onset of pain he was free from any recent aural or neurological symptoms. Audiology showed the expected combined conductive and sensorineural hearing loss. No notes documenting his previous admission and surgery were available.

Examination of the ear revealed a slightly wet cavity containing a little debris with a deficient bony roof with exposed pulsatile dura. The swelling itself was approximately 8 cm in diameter, subcutaneous, well-circumscribed and multilobulated. It was not tender and had no features associated with sepsis, the patient's suggestion to 'put a knife in it' was declined. The patient had no neurological abnormality, in particular no nystagmus and a negative fistula sign.

A computed tomography (CT) scan with axial slices pre- and post-contrast and direct coronal images was performed.

It was noted that this large multiloculated mass was associated with a scalloped defect in the squamous temporal bone anteriorly and of the anterior aspect of the petrous bone posteriorly. A post-surgical defect in the posterior margin of the petrous bone laterally in the region of the lateral sinus was also noted (Figure 1(a)). The lesion was noted to bulge subcutaneously and have an extra dural intracranial component (Figure 1(b)). It was felt that the clinical and radiological evidence indicated this lesion to be benign with cholesteatoma heading the list of differential diagnoses.

At operation our clinical suspicions were confirmed. A flap was raised (the incision being a variation on the middle fossa approach) exposing the 8 cm lobulated lesion in its entirety. The lesion lay beneath the temporalis muscle and extended intracranially through the bony defect previously described. The appearance was of a dumbbell-shaped lesion, with large extra- and intra-cranial deposits linked by a narrower section as it traversed the bony defect. The dura although displaced medially had not been compromised.

The cholesteatoma sac was opened revealing its contents before being removed in its entirety. The bony defect was left unrepaired before closure of the wound and drain insertion. The patient's post-operative recovery was uneventful.

Discussion

Most cholesteatomas will remain confined to the petrous bone however, spread beyond this confine, to involve the labyrinth and middle cranial fossa can occur.[1] To our knowledge no description of cholesteatoma displacing the soft tissues overlying the temporal bone has previously been recorded.

There is the question as to how it arose. It is possible it has developed as an entirely separate lesion to the original one removed 50 years previously; a second cholesteatoma that went unnoticed. It may represent a recurrence developing from a remnant left in the anterior attic region.

This case beautifully illustrates the capacity for eroding bone that cholesteatoma possesses. It is generally considered that activation of osteoclasts is the main cause for erosion. What stimulates their action is a matter for conjecture. Humoral factors, such as prostaglandins, cathepsin D and a parathyroid hormone-like protein have been suggested,[2] as have mediators of the inflammatory response, notably interleukin-1alpha from macrophages.[3]

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