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Clinical Records

Unusual cases of congenital cholesteatoma of the ear

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Abstract
Congenital cholesteatoma may originate at various sites in the temporal bone. For example, in the petrous apex, the cerebellopontine angle, the middle ear cavity, the mastoid process or the external auditory canal. The least common site being the mastoid process. We present two cases of congenital cholesteatoma of the mastoid process, each presenting with different symptoms and at different ages. Both patients underwent surgical treatment, which confirmed the diagnosis and radiological findings.

Key words: Cholesteatoma; Petrous bone; Surgery

Introduction

Howard House (1953) was the first to report a cholesteatoma behind an intact tympanic membrane (Shuknecht, 1993). Congenital cholesteatoma originates from the same ectoderm that forms the primitive notochord, and embryonic cell rests from this ectodermal structure may occur in any of the cranial bones (Proctor, 1991). The temporal bone is usually affected, and congenital cholesteatoma may occur in five sites: (1) the petrous apex (2) the cerebellopontine angle (3) the mastoid (4) the middle ear and (5) the external auditory canal (Shuknecht, 1993). According to Nager (1993) occurrence in the mastoid process is the least frequent. As initially postulated by Korner (1900) the key for diagnosing congenital cholesteatoma is the absence of previous local trauma or infection and the presence of intact drum.

Case reports

Case 1

A 55-year-old lady presented with symptoms of inability to localize sound direction properly. This had started four years ago and was followed by a left-sided diminution of hearing several months later. There was no history of ear discharge, ear infections, ear operations, dizziness or other otological complaints, nor of facial weakness, tinnitus or a decrease in speech discrimination. She used to complain of a deep-seated dull pain in the temporal region years ago, and she had a relevant family history of deafness due to otosclerosis. Examination showed normal-looking tympanic membranes on both sides, and tuning fork tests revealed a left-sided conductive hearing loss, which was confirmed by audiological assessment. This showed a conductive deafness of about 40 dB with a dip in bone conduction at 2 KHz (Carhart notch).

A computed tomography (CT) scan revealed a large mass that occupied the left mastoid cavity, the middle-ear cavity extending to the posterior cranial fossa eroding the sinus plate, the posterior wall of the mastoid cavity, the cochlea and the facial canal down to the mastoid tip (Figure 1). Magnetic resonance imaging (MRI) showed a large mass occupying the region of the mastoid process, which extended to the posterior cranial fossa. This mass

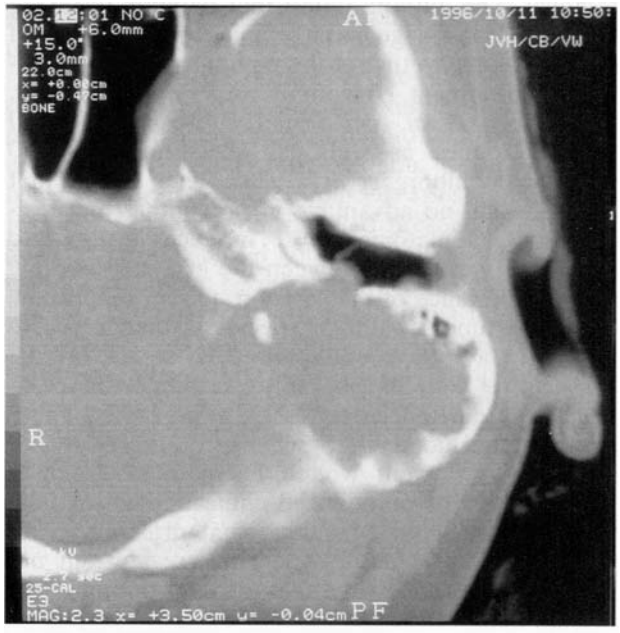

FIG. 1
CT scanning of temporal bone of the first patient showing the extent of cholesteatoma.

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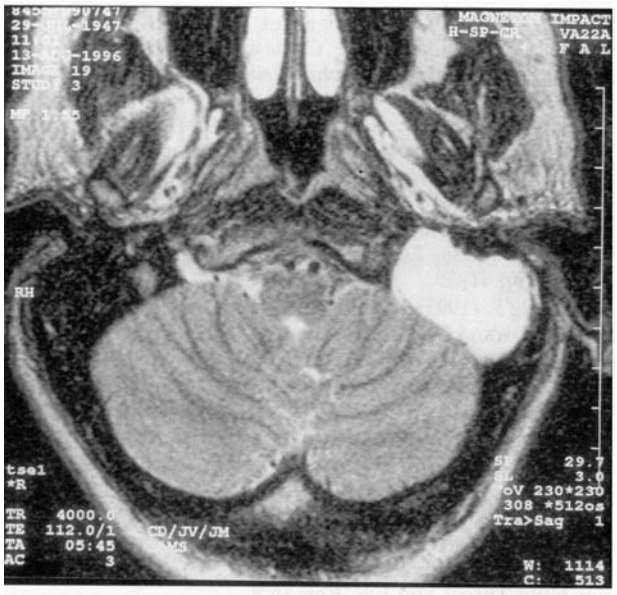

FIG. 2

T2-weighted MRI scan of the first patient showing the hyper-intense nature of the cholesteatoma.

was hypo-intense in T1-weighted and hyper-intense in T2-weighted images (Figure 2). The mass was surgically removed, and histology proved it to be a cholesteatoma confirming the radiological findings. The cholesteatoma was so extensive that it had eroded the bone on the medeal aspect of the facial nerve canal adjacent to the sinus tympani. The facial nerve had been surrounded by the cholesteatoma and was covered with granulations. In transposing the facial nerve to remove the cholesteatoma, it divided. The nerve was repaired by primary suturing. Post-operatively the patient developed a CSF leak from the wound, which was managed by a lumbar drain for five days. She also had immediate post-operative facial nerve paralysis. A follow-up after four months showed a facial weakness House Brackman grade III.

Case 2

A 13-year-old girl presented to the ENT casualty several times with a history of a discharging left ear. Each time the ear was examined the tympanic membrane was found to be normal and she was managed with aural toilet and topical antibiotic drops. There was no history of ear operations,

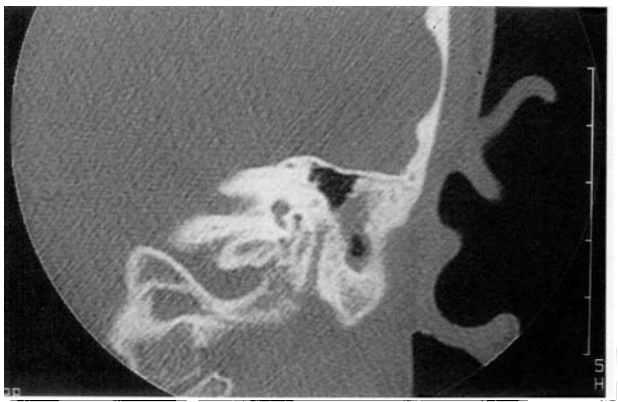

FIG. 3

CT scan of temporal bone of the second patient showing the mastoid cavity cholesteatoma with clear middle ear cavity.

ear trauma, otalgia or loss of hearing. Microscope examination revealed a small fistula in the posterior meatal wall immediately medial to the junction of the bony and cartilaginous meatus. It became apparent that the opening of the fistula had been missed because an aural speculum went beyond the fistula site when inspecting the eardrum. High resolution CT scanning showed that the whole mastoid cavity was filled by a radio-opaque mass that had eroded the external auditory canal through to the bony meatus. The extent of cholesteatoma had stopped short at the aditus leaving the middle ear and ossicles intact (Figure 3). An audiogram revealed excellent hearing with no air-bone gap.

Surgery confirmed the previous radiological extent and proved that it was a cholesteatoma. On follow-up after two months the discharge had stopped and hearing had remained the same.

Discussion

The mode of presentation of congenital cholesteatoma differs according to the site. In addition, Browning (1997) reported that there is a link between the mode of presentation and age. If it arises within the middle ear cleft, it presents with a hearing loss in childhood due to its effect on the ossicular chain. Alternatively, if the origin is within the petrous apex of the temporal bone, it presents in adulthood when it presses on the facial nerve or brainstem.

The patient in *Case 1* started to complain of hearing problems in late adulthood with no history that suggested brainstem compression or facial nerve involvement. Her hearing loss could have been attributed to either the cholesteatoma or otosclerosis, which runs in her family. Because of the typical audiogram and normal-looking tympanic membranes she could have been easily mis-diagnosed as otosclerosis. However, the presence of deep-seated pain in the temporal region suggested the value of having CT scanning, which diagnosed the condition. The absence of any previous history of ear infection, trauma or operation pointed to the congenital origin of the cholesteatoma. Luntz *et al.* (1997) presented a similar case involving a smaller cholesteatoma. The cholesteatoma described in this report had destroyed most of the middle-ear cleft, which made it similar to the case presented by Nager (1982).

This differs to the second case reported here who presented several times with an ear discharge, which, on microscopic examination was found to come from the posterior wall fistula through the external auditory canal. This was confirmed (and excised) by surgery. The presence of an intact tympanic membrane and the absence of any history of ear trauma or operation, in addition to the site of the cholesteatoma, which was confined to the mastoid cavity with no involvement of the middle ear, confirmed the diagnosis of congenital cholesteatoma. Huang and Lee (1994) reported two cases of cholesteatoma that fistulated through the external bony canal and presented with ear discharge. However, their two cases might not comply with the characterization of congenital cholesteatoma because the patients had had surgical ear operations. The second case reported here appears to be only the third recorded example of congenital cholesteatoma localized in the mastoid cavity. (Derlacki and Clemis, 1965; Luntz *et al.*, 1997). Luntz *et al.* (1997) suggested three characteristic features for congenital cholesteatoma originating in the mastoid. These are:

(1) Initial presentation as neck pain.
(2) CT findings of a cystic, expansile lesion occupying the mastoid process without involving the middle ear.

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(3) Magnetic resonance imaging findings of hyperintensity on T2-weighted images with little or no peripheral enhancement on post-contrast T1-weighted images.

We believe that these three criteria may not be present in every patient. The young girl reported here presented with otorrhoea rather than neck pain. The rarity of such cases may explain the inability to establish precise characteristic features.

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