Spontaneous cholesteatoma of the external auditory canal: The utility of CT

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Abstract
Spontaneous external auditory canal (EAC) cholesteatoma is a rare disease. The symptoms are nonspecific. It is diagnosed by clinical examination and radiological investigation. The clinical examination alone is often insufficient for accurately assessing spread of the cholesteatoma into the temporal bone, meaning cross-sectional imaging modalities are required, and specifically computed tomography. We report three cases of spontaneous cholesteatoma of the external auditory canal. All of our patients underwent surgery. In two cases, the cholesteatoma was restricted to the external auditory canal, while in one case, it was complicated by a fistula with the lateral semicircular canal. Good anatomical and functional results were obtained in all three cases, with the external auditory canal patent and a good calibre on completion.

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Spontaneous cholesteatoma of the external auditory canal (EAC) is defined as an accumulation of keratin over an osteitic bone erosion [1–4]. It is a rare condition, or one that is not often diagnosed. It accounts for 0.1–0.5% of otologic disease [1,3].

This condition has been principally diagnosed clinically, but it has significantly benefited from progress in imaging, that means that it can be differentiated from other inflammatory pathologies or tumours of the EAC [3,4].

We report three case studies of spontaneous cholesteatoma of the EAC and use this work to describe the role of imaging in making a positive diagnosis, preoperative assessment of the lesion, and long-term follow-up of this condition.

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Clinical case studies

Clinical case study 1

S.H., a 62-year-old male with no particular medical history, was admitted to hospital for a one-year history of purulent left otorrhoea that was associated with otalgia, a humming sound in the ear, and progressively worsening partial deafness in the left ear. The clinical examination on admission found a left patent EAC of reduced calibre. The tympanum was intact and hearing was normal.

No abnormalities were noted in the contralateral side with the rhinological and remaining ENT and physical examinations being normal.

In view of these observations, the diagnosis of cholesteatoma of the left EAC was made and a CT scan of the petrous temporal bone was carried out for preoperative assessment. This demonstrated a left EAC of reduced calibre due to being partially filled with eroded bone from the posterior wall (Fig. 1a). The tympanic cavity was almost entirely blocked, as were the various recesses of the middle ear. The ossicles were normal in terms of position, density, and morphology (Fig. 1b). The lateral attic wall was intact, as were the tegmen tympani, and the cortical bone of the facial canal and lateral semicircular canal.

The patient underwent surgery, and perioperative observations confirmed a cholesteatoma of the EAC with a wide posterior cavity packed with cholesteatoma. After the tympanum was detached, the cavity was seen to be full of inflammatory tissue without cholesteatoma.

Clinical case study 2

D.H., a 76-year-old male with no medical history of interest, was admitted to hospital for a six-month history of humming in the ear, vertigo, and otalgia with no associated otorrhoea. The clinical examination revealed eczema in the left EAC with an epidermal plug and a spontaneous attic perforation. The tympanic membrane was flaccid though intact. The right ear was normal on examination. An audiometric test found that the stapes reflex was absent on both sides.

The possible diagnoses were simple chronic otitis media without cholesteatoma in view of the absence of otorrhoea, or chronic otitis media with cholesteatoma given the vertigo and spontaneous attic perforation.

Since there was no definite diagnosis, a CT scan of the petrous temporal bone was carried out. It showed increased soft tissue density of the tympanic attic and mesotympanum combined with erosion of the lateral attic wall, a fistula of the lateral semicircular canal (Fig. 2a), and partial erosion of the sheath of the second portion of the facial nerve. This was associated with erosion to the bone of the floor of the EAC and the tympanic bone (Fig. 2b).

The patient underwent surgery, with a microscopic examination showing a narrowed auditory canal with wide erosion to the posterior wall and flakes of cholesteatoma. The tympanic membrane was intact and in good condition. The cholesteatoma, which had fused at the back and ended just proximal to the tympanic sulcus, was aspirated: the incus was extracted after disarticulation, as was the malleus. The matrix of cholesteatoma and the cartilage filling the eroded lateral semicircular canal were removed.

The following perioperative observations were made: the cholesteatoma had caused erosion to the lateral semicircular canal, spreading to below the facial canal. The ossicles were intact and mobile.

Clinical case study 3

H.K., a 32-year-old male patient with no particular medical history attended a consultation due to an eighteen-month history of mild, intermittent left otalgia without associated otorrhoea or reduced hearing. No rhinological signs.

The clinical examination showed a widened EAC with eroded walls that were covered with epidermal squames. The tympanum was intact and hearing was normal. No abnormalities were found on examination of the contralateral ear. A diagnosis of cholesteatoma of the EAC was made. A CT scan of the petrous temporal bone confirmed that the EAC was enlarged and showed increased soft tissue density, which was associated with erosion to the posterior and inferior walls of the canal (Fig. 3). The tympanic cavity was patent.

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Figure 1. High resolution CT scan of the petrous temporal bone, axial views: a: left EAC of reduced calibre partially filled with erosion from the posterior wall (arrows); b: almost completely filled middle ear with ossicles appearing normal (arrowhead).
and the ossicles were intact, with normal pneumatization of the mastoid bone and a second segment of the facial nerve in its sheath.

The patient underwent surgery via the posterosuperior approach. There was a cholesteatoma in the left EAC with significantly eroded posterior and inferior walls extending as far as the tympanic sulcus. The cavity was clean and the ossicles were intact.

The lesions were debrided and cartilage was used to reconstruct the EAC. Good progression was made and the anatomical result was good.

**Discussion**

Spontaneous cholesteatoma of the EAC is defined as an accumulation of keratin over an osteitic bone erosion [1–5].

Cholesteatoma usually affects the middle ear and it is much less common for it to involve the EAC. Its incidence is estimated at 1/1000 new patients in otology [2,5]. It is problematic to make a positive diagnosis, as well as to exclude differential diagnoses and to choose a treatment due to lesion spread [2]. Our second case study illustrates this and raises the difficulties in diagnosing this condition.

The aetiology and pathogenesis remain poorly elucidated [3–6]. Several theories have been put forward: localised periostitis, chronic inflammation of the EAC, a failure to spontaneously eliminate desquamated epithelial cells, and dehiscence of the petrotympanic fissure. This is how the idea came about that this was primarily bone involvement that could be caused by infection or ischaemia leading to necrosis and reactive periostitis or the reverse. The excessively desquamated appearance of the epithelium due to blocked lateral epithelial migration, a phenomenon specific to the epidermis of the bony portion of the EAC, was in this case thought to be triggered by an inflammatory process, whether this originated locally to the canal or in an adjacent structure [1,3,6–8].

**Figure 2.** High resolution CT scan of the petrous temporal bone: axial (a) and coronal views (b). Cholesteatoma with fistula of the lateral semicircular canal: tissue blocking the tympanic cavity associated with osteolysis of the bony sheath of the lateral semicircular canal (white arrow); this is associated with erosion to the bone of floor of the EAC (black arrows).

**Figure 3.** High resolution CT scan of the petrous temporal bone: axial (a) and coronal views (b). Enlarged left CAE with partially increased soft tissue density (white arrows), associated with erosion of the posterior and inferior walls of the canal (black arrows).
In terms of anatomical pathology investigations, a study of cholesteatoma of the EAC confirms the presence of periostitis restricted to the eroded area, as well as osteitis, bone sequestra, and reactive inflammatory tissue. Epidermal cells are found in the excavated portion surrounding bone sequestra. A build-up of keratin can from there form a sac in a bone recess or around a sequestrum, and progressively spread to the mastoid bone [1,9].

Clinically, spontaneous cholesteatoma of the EAC is normally considered to affect elderly patients [1,3]. The ages of our patients ranged from 32 to 76 years.

Usually only one side is involved, but bilateral presentations do exist. There is no predestination for either sex [4,5,7].

On interview, these patients do not report any specific ENT history of interest. There is huge variation in the abundance of otorrhea, while otalgia is fairly mild and facial paralysis is rarely reported. Sometimes the condition is an incidental finding [1,6,10,11]. For our patients, the main reason for consultation were severe otalgia and humming in the ear.

On examination, the bony portion of the canal shows erosion to the floor with posterior and sometimes anterior extension. This erosion is usually located close to the sulcus but it can also be seen further out, or indeed at the junction between the bony and cartilaginous portions of the EAC [1,3]. The eroded area is filled with bone sequestra, and these signs form the criteria for a making a positive diagnosis. The tympanum is normal or mildly irritated if there is significant inflammation. There is no argument in favour of an associated middle ear pathology. Pure tone audiology testing may show mild conductive hearing loss or normal hearing [3,7].

Cross-sectional imaging modalities and especially computed tomography are very useful for positive diagnosis, since this approach can pinpoint the localisation of the cholesteatoma and accurately assess the lesion, in particular bone involvement and depth of spread [2–4,8,10,11].

Cholesteatoma typically appears in the form of an oval soft tissue opacity associated with bone lysis with distinct and regular margins. These bony erosions can reach as far forward as the temporomandibular bone, below to the hypotympan and the jugular bulb, and back to the mastoid bone and the wall of the facial nerve canal. The tympanic membrane usually remains intact [1,3,7].

The first classification of cholesteatoma of the EAC into four stages using clinical and histological information was established by Naim et al.: stage I: hyperplasia of epithelial cells in the canal; stage II: presence of periostitis; stage III: erosion of the bony portion of the canal; stage IV: erosion of adjacent structures [7,9,12].

More recently, Shin et al. have put forward a CT classification of EAC cholesteatoma that provides an improved guide in terms of therapeutic management. This classification consists of four stages: stage I: cholesteatoma of the EAC only; stage II: invasion of the tympanic membrane and the middle ear as well as EAC involvement; stage 3: EAC defect and involvement of the air cells of the mastoid bone; stage 4: lesions beyond the temporal bone [12].

EAC cholesteatoma must be distinguished from other pathologies of the canal that can have a similar clinical presentation. Firstly, the entity defined as spontaneous cholesteatoma must be dissociated from any secondary cholesteatomatous lesions, such as: accumulations of keratin in the canal resulting from congenital or acquired stenosis (due to trauma or surgery), epidermal cysts beneath the skin of the canal found some time after surgery to the middle ear, epidermal invasions secondary to a fracture of the tympanic bone, and the very rare congenital epidermal cysts to the posterior canal wall [3,10]. A localised form of osteoradionecrosis of the tympanal bone can also produce a very similar clinical picture of bone erosion, sequestra, granulation tissue, and keratin accumulation, so is essential to look for a history of head and neck irradiation in this situation [7,8,10]. Necrotising otitis externa can be identified by its specific predisposing factors (it mainly develops in elderly and diabetic patients) and on examination of the EAC, which shows the canal to be inflamed and with a least some degree of stenosis as well as granulation tissue being present at the junction of the bony and cartilaginous portions of the EAC. Imaging is essential in order to confirm the diagnosis at an early stage and to pinpoint the localisation and spread of the infection [1]. Finally, the possibility of an ulcerated neoplastic lesion must be considered in principle, and this will be confirmed by biopsy [1,3].

Treatment depends on the stage of the condition: when lesions are localised, the treatment consists of frequent cleansing and debridement of necrotic tissue as well as evacuation of thick cerumen. The deepest pockets will require surgical reconstruction of the canal and a mastoidectomy is indicated when the mastoid bone is involved [1–3,7].

The criteria for surgical treatment include chronic pain, failed medical treatment, recurrent infections that encourage resistant strains, cholesteatoma complicated by facial paralysis or vertigo, and cholesteatoma spreading in spite of medical treatment that is well adhered to [3,5,8,11,13].

It is difficult to find a timescale for recurrence in the literature. There is general agreement that long-term monitoring is needed [13]. When an open surgical technique is used, a follow-up clinical examination under the microscope is sufficient. When conservative techniques are used, a post-operative CT scan will be required, and this will provide a comparator for future CT scans to be carried out annually, or if the patient develops symptoms such as otorrhea or pain [13].

Conclusion

Spontaneous cholesteatoma of the EAC is a rare and little-known condition, when compared to acquired cholesteatoma. There are no specific clinical symptoms, and this can lead to confusion with other pathologies of the EAC. It is diagnosed clinically and the assessment of disease spread requires very close radiological analysis. The chosen treatment will depend on the extent of the lesions and the clinical symptoms. In progressive forms, reconstructive surgery of the EAC is required.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.
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