External auditory canal cholesteatoma: Clinical and therapeutic aspects

Cholésteatome du conduit auditif externe: Aspects cliniques et thérapeutiques.

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ABSTRACT:

Objective: Describe the different clinical aspects and therapeutic modalities of the external auditory canal cholesteatoma
Methods: a retrospective study that collected four cases of spontaneous cholesteatoma diagnosed at our ENT and CCF department over a period of 10 years.
Results: The main features were: otorrhea, earache and hearing loss. Cholesteatoma extended to the middle ear in three patients, and it was limited to the external auditory canal in one patient. All of our patients underwent surgery. Good anatomical and functional results were obtained in all patients with no recurrence at 5 years of follow-up.
Conclusion: Cholesteatoma of the external auditory canal remains a rare pathology but which requires adequate surgical management based on reconstruction of the canal in order to minimize the risk of recurrence, which remains considerable.
Key-words: Cholesteatoma; Spontaneous; External auditory canal; Imaging, surgery

RÉSUMÉ

Objectif: Rapporter les différents aspects cliniques et les modalités thérapeutiques du cholestéatome du conduit auditif externe.
Methodes: Une étude rétrospective a colligé quatre cas de cholestéatome spontané diagnostiqués à notre service d'ORL et CCF sur une période de 10 ans.
Résultats: Les principaux symptômes étaient: l'otorrhée, l'otalgie et la perte auditive. Le cholestéatome était étendu à l'oreille moyenne chez trois patients et était limité au méat auditif externe chez un seul patient. Tous nos patients ont été opérés. De bons résultats anatomiques et fonctionnels ont été obtenus chez tous les malades avec absence de récidive à 5 ans de suivi.
Conclusion: Le cholestéatome du conduit auditif externe reste une pathologie rare mais qui nécessite une prise en charge chirurgicale adéquate basée sur la reconstruction du conduit afin de minimiser le risque de récidive qui reste considérable.
Mots Clés: Cholésteatome ; Spontané ; Canal auditif externe ; Imagerie, chirurgie

INTRODUCTION:

External auditory canal cholesteatoma (EACC) is relatively rare but also a miss-diagnosed disease. Although its etiology has been described as the disturbance of epithelial migration due to recurrent microtraumas and microangiopathy, it remains to be fully elucidated.[1]

A distinction from keratosis obturans has been introduced only in the last few decades. Both mainly consist of epithelial debris in the external acoustic canal. The most useful findings confirming an external auditory canal cholesteatoma are focal osteonecrosis with or without sequestration and lack of epithelial covering of the bony surface. The distinction between keratosis obliterans and cholesteatomas is not merely one of pedantry, but impacts on treatment strategy[1,2].

Diagnosis is based on clinical examination and radiological investigation: High-resolution bone window temporal bone CT is the method of choice to localize the cholesteatoma and to demonstrate bony erosions. They appear as soft tissue attenuating lesions. MRI cannot show the early involvement of compact bone. Its role in EACC is not defined [1,3].

Small lesions can be treated endoscopically under local anesthesia, whereas larger ones need surgery to remove the cholesteatoma and affected bone areas. Grafting of defects may be necessary. Prognosis depends on the extent at the time of diagnosis [1,3].

By reporting 4 cases of external auditory canal cholestreatoma, our objectives were to analyze clinical

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presentation of this rare pathology and discuss with review of literature therapeutic modalities.

METHODS:

We retrospectively reviewed the medical records of patients with previously untreated EACC in the Department of Otolaryngology, Head and Neck Surgeryof Monastir. Before therapy, all patients underwent physical examination, pure-tone audiometry, as well as computed tomography (CT) to evaluate the lesions. Histopathological examination was performed in all cases to support the diagnosis of EACC, and to exclude the possibility of EAC cancer or malignant otitis externa.

Table 1: Stages of the external auditory canal cholesteatoma [6]

STAGE	DEFINITION
I	Limited to the external auditory canal
II	Invades the tympanic membrane and middle ear in addition to the external auditory canal
III	Creates a defect of the external auditory canal and involves the air cells in the mastoid bone
IV	Involves lesions beyond the temporal bone

RESULTS:

Four patients, a man and 3 women were diagnosed in our ENT department for EACC. In all cases, cholesteatoma was spontaneous. The median quartile of age was 41 years with extremes of 20 and 82 years. We estimate that the first consultation was performed at least 17 months after the first ontological sign (3 months to 3 years).

The clinical signs were dominated by otorrhea and hearing loss found in three cases. Otalgia was reported by two patients. On examination, an erosion of the postero-superior wall of the duct was noted in one case and an excavation of the posterior wall in another one. We also found a stenosis of the external auditory canal with cholesteatoma's aspiration in three cases. The tympanic membrane was infiltrated and retracted in three cases, and normal in one case.

Examination of the controlateral ear was normal for all patients.

No facial palsy was noted, and the neurological examination was normal.

Tonal audiometry was normal in one case. It founded transmission hearing loss the other three cases. The average of hearing loose was 35 dB with extremes of 20 To 55 dB.

On imaging, bone's erosion of the wall of the external auditory canal, especially the posterior and inferior one, was found in 3 cases. A balloon appearance of the duct was noted in 2 cases (Figure 1). A formation occupying the entire duct with partial erosion of the tympanal bone was found in 1 case (Figure 2).

In the two other cases, the CT scan showed a filling of the duct as well as of the middle ear (figure 3). No cases of extension to the third portion of the facial nerve or to the lateral semicircular canal were noted. Cholesteatoma was classified as stage I in one case, stage II in two cases and stage III in one case.

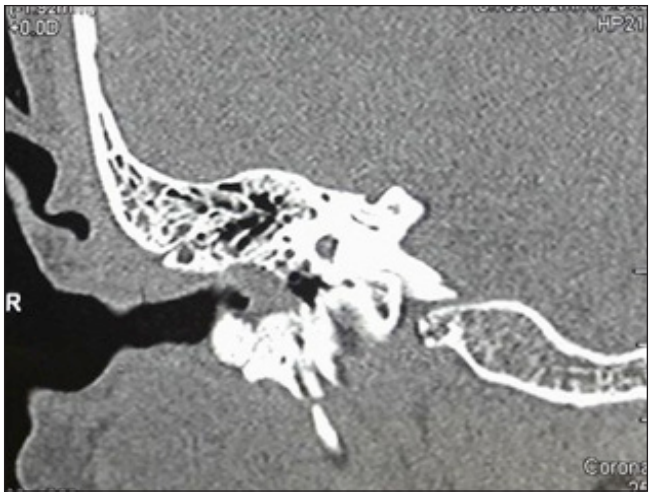

Figure 1: coronal view of high resolution CT scan of the petrous temporal bone: showing an enlarged right CAE with a soft tissue filling the entire duct with a balloon appearance

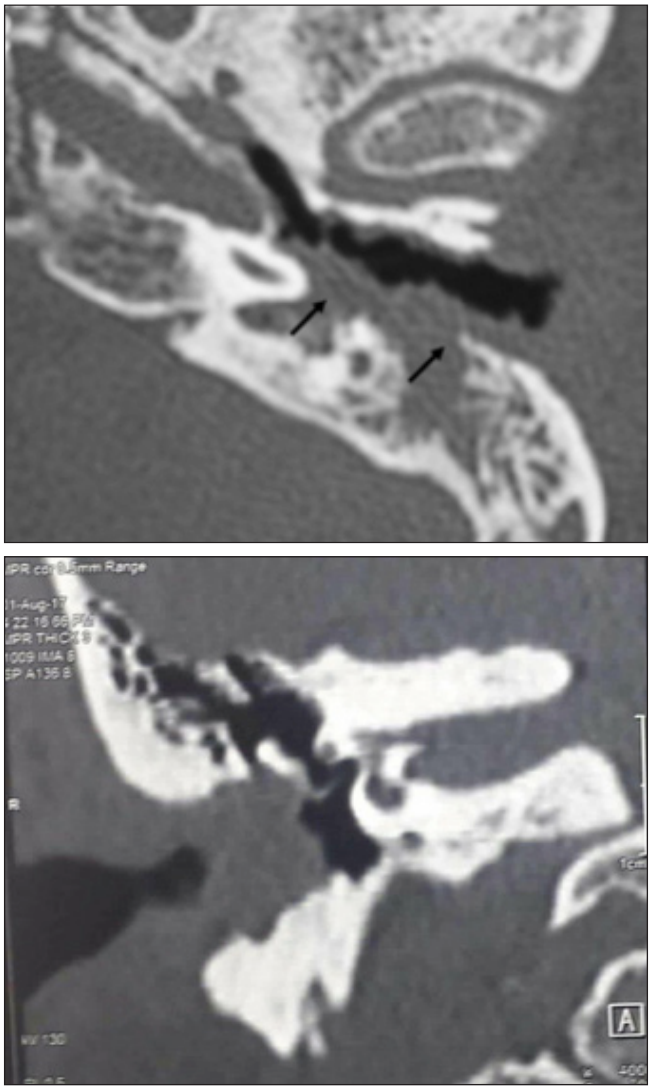

Figure 2: Axial and coronal views of high resolution CT scan of the petrous temporal bone: A soft tissue filling the entire duct associated with erosion of the posterior and inferior walls of the canal and of the lateral attic wall.

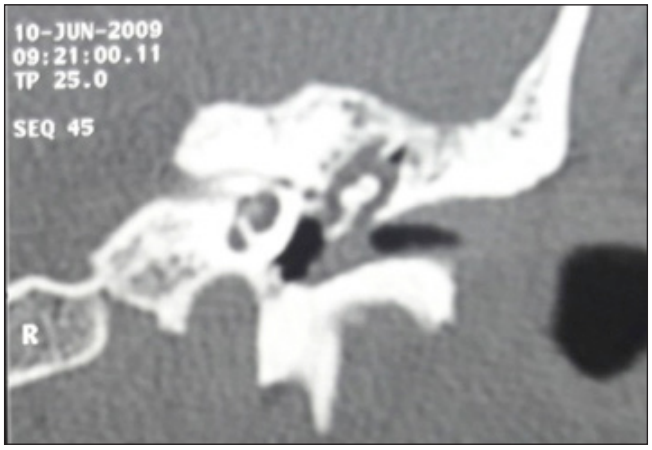

Figure 3: coronal CT scan view showing a filling of the duct and of the middle ear with a soft tissue

All our patients underwent surgery. In one case cholesteatoma was removed by way of the canal. In another case, bony meatoplasty was performed through a postauricular approach, with an ear canal skin incision just lateral to the cholesteatoma pocket. The cholesteatoma pocket was elevated, and the bony irregularities and ridge were drilled to smooth the cavity. The third patient had a canal wall up tympanoplasty and the last one a canal wall down tympanoplasty.

Patients showed good outcomes without recurrences, in a postoperative period extended from one to nine years.

DISCUSSION:

Cholesteatoma is most commonly found in the middle ear cavity. Rarely, cholesteatoma occurs in the external auditory canal, with an estimated incidence rate of 1.2 to 3.7 per 1000 new otology patients [1]. The annual incidence rate of external auditory canal cholesteatoma in the general population is 0.15 cases per 100 000 individuals, compared with 9.2 cases per 100 000 individuals per year for middle-ear cholésteatome [2].

The etiology of the external auditory canal cholesteatoma remains unclear. Many authors referred to the explanation of an absent or reduced migratory capacity of the canal epithelium leading to keratinization in situ and thus EACC formation [3].

Epidemiologically-linked risk factors for the development of external auditory canal cholesteatoma included smoking, diabetes mellitus and repeated micro trauma [4]. Smoking and diabetes are thought to result in microangiopathic changes in the ear canal that potentially impair epithelial migration [5]. The majority of idiopathic EACC occurred in the inferior canal wall. This is thought to be due to the relatively poor blood supply to the skin along the inferior wall of the canal [6]. 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 [6,7]. The eroded area is filled with bone sequestra. This aspect is one of the positive diagnosis criteria. There is no argument in favour of an associated middle ear pathology. Pure tone audiometry testing may show mild conductive hearing loss or normal hearing [1,7].

None of the cases presented here had an identifiable etiological factor. They can be regarded as idiopathic external auditory canal cholesteatoma. One of the patients was smoker and another was diabetic.

A recent meta-analysis of published case series concluded that the most common presenting symptoms of idiopathic external auditory canal cholesteatoma are unilateral otorrhea with mild to moderate earache [7].A minority of patients complained of unilateral hearing loss; or were asymptomatic. These findings were consistent with our experience. Tow patients presented with unilateral earache, three had otorrhea. One patient had painless otorrhea and three complained of progressive unilateral hearing loss.

Computed tomography of the temporal bone is the gold standard for staging and pre-operative planning in external auditory canal cholesteatoma [8,9]. It allows accurate evaluation of the extent of local bone erosion and the involvement of adjacent structures. Naim et al developed a (I–IV) staging system for external auditory canal cholesteatoma based on the extent of erosion into nearby structures (table1). The majority of cases reported in the literature are of at least stage II [10]. Reports of stage I external auditory canal cholesteatoma are very rare [11]. For our patients, cholesteatoma was classified as stage I in one case, stage II in two cases and stage III in one case.

Treatment depends on the stage of the condition. When lesions are localized, the treatment consists of frequent debridement of necrotic tissue. The deepest pockets will require surgical reconstruction of the auditory canal usually with the aid of cartilage grafts to protect denuded structures, and fill any canal wall defect. Mastoidectomy is indicated when the mastoid bone is involved [1,2,3,7].

The criteria for surgical treatment include chronic pain, failed medical treatment, recurrent infections that encourage resistant strains, cholesteatoma complicated by facial palsy or vertigo [9,10,11,12,13]

It is difficult to find a timescale for recurrence in the literature. There is general agreement that long-term monitoring is needed [13]. 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 [12,13].

CONCLUSION:

External auditory canal cholesteatoma is a rare disease entity, which presents diagnostic and therapeutic challenges. Computed tomography is the most useful investigation for disease staging and pre-operative planning. The treatment is based on surgery. Its aim is to remove the cholesteatoma and to restore a smooth, self-cleaning canal wall epithelium.

The real challenge is to maintain and reconstruct the external auditory canal if significant lesions occurred, while trying to minimize as much as possible factors that may increase the risk of recurrence of these lesions.

Compliance with ethical standards

Conflict of interest: The authors stated that there is no conflict of interest.

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