

# External Auditory Canal Cholesteatoma (EACC)

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## ABSTRACT

**Introduction:** External auditory canal cholesteatoma (EACC) is a rare form of cholesteatoma occurring commonly in the elderly patients. It can be spontaneous or secondary to several conditions. The aim of our study was to review the clinical characteristics of the EACC and to evaluate the outcome of its management approaches.

**Methods:** It's a retrospective study about seven patients managed for EACC collected between 2003 and 2017. Age, gender, previous surgical approach, time of onset of symptomatology, clinical signs, pure tone audiogram assessment, temporal bone CT findings, medical and surgical treatments and the final outcome were collected and analyzed.

**Results:** The mean age of patients was 45.6 years (range 20 –77 years), the male to female ratio was 6. Mean clinical course of cholesteatoma was  $1.4 \pm 1.4$  years [1month-5years]. Cholesteatome was bilateral in three cases. The most common symptom was purulent otorrhea. CT-Scan had shown typical radiographic evidence of EACC stage I in 1 case, stage II in 1 case, stage III in 4 cases and stage IV in 1 case. EACC was classified as primary in six cases and secondary in one case. Surgical approach was performed through modified radical canal wall down mastoidectomy in four cases, a transmeatal approach, tympanoplasty and canal wall down mastoidectomy respectively in one case each. All ears remain dry with no evidence of recurrence at last follow-up.

**Conclusion :** EACC is a rare entity with characteristic imaging and clinical features. It may exhibit features including deep extension into the middle ear, mastoid, facial nerve canal, or the tegmen tympani. These findings may be unrecognized at clinical examination and may influence the clinical and surgical management of EACC.

**Keys-words:** Cholesteatoma, External Auditory Canal, Surgery.

## INTRODUCTION

External auditory canal cholesteatoma (EACC) is a rare form of cholesteatoma first described by Toynbee in 1850, corresponding to the invasion of the squamous tissue into a localized area of bony erosion of the ear canal. It is 60 times less common than the classic middle ear cholesteatoma and occurs commonly in the elderly patients. Pediatric cases are rarely reported. The differential diagnosis of this condition includes keratosis obturans, necrotizing external otitis and malignancies such as squamous cell carcinoma. It can be spontaneous or arise secondary to several conditions. Thus the surgical treatment must be adapted in accordance with each etiology. The purpose of this study is to review the clinical characteristics of EACC and to evaluate the outcome of management approaches for the treatment.

## MATERIAL AND METHODS

We review retrospectively the medical records of all patients with EACC admitted to our department, during 15 years between 2003 and 2017. Age, gender, previous surgical approach, time of onset of symptomatology, clinical signs, pure tone audiogram assessment, temporal bone CT scan findings, medical and surgical treatments and the final outcome were collected and analyzed. The diagnosis was

based on a history of recurrent otorrhea and dull intermittent otalgia, medical examination, and radiographic results showing a focal disruption of the skin with underlying bony erosion, and was further supported by histopathologic examination. Six cases without obvious etiology were classified as primary EACC, while one case with osteoma found on examination was classified as secondary.

## RESULTS

The mean age of patients was 45.6 years (range 20 –77 years), the male to female ratio was 6. No hearing aids were revealed in all cases. Mean clinical course of cholesteatoma was 1.4 years [1month - 5years]. Four patients had a right sided cholesteatoma, and 3 patients had a left sided one.

The most common symptoms were purulent otorrhea (85.7% ears) unresponsive to topical and systemic antimicrobial therapy, followed by complaints of hearing loss (57.1% ears). Otalgia was noted in 2 cases (28.6% ears). No complaints of Tinnitus or vertigo were noted in the all cases.

Physical examination, revealed the presence of keratin debris in the EAC in 6 cases with focal bony erosion, a polyp occupied the posterior portion of the canal in one case.



Otorrhea was found in 3 cases. In one case, EAC was completely occluded by a fixed hard mass making impossible to examine the tympanic membrane.

Pure tone audiometry was normal in two cases. It concluded to a conductive hearing loss with a mean threshold of 40 dB in two cases, a deafness in one case. One patient had mixed hearing loss, the threshold was 80 dB. The last patient had a sensorineural hearing loss, the mean threshold was 30 dB.

Temporal bone CT was carried out in all cases, establishing the diagnosis, revealing circumferential involvement of the EAC in 3 cases, and the erosion involved more than one wall in the 3 others. The posterior and inferior walls were most commonly involved (85.7%)(figure1). CT-Scan had shown typical radiographic evidence of EACC stage I according to Shin classification (Table1) in one case, stage II in one case, stage III in 4 cases and stage IV in one case.

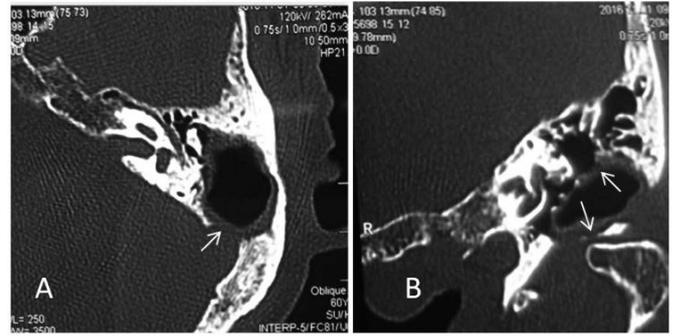
**Table I :** CT scan findings according to Shin classification

Stade	Number of patients
I	1
II	1
III	4
IV	1

Eighty five per cent of patients had evidence of extension of disease into the middle ear and 71.4 % had further erosion into the mastoid cells. Erosion exposed the temporomandibular joint in one case, one patient had exposure of the sigmoid sinus (figure 2), and one had erosion of the lateral semicircular canal. The cortical bone of the facial canal was intact in all cases. For the secondary EACC, the ear canal was filled by many irregular ovoid bony masses with soft tissue density of the middle ear with extension into the mastoid cells (figure 3). Two patients underwent MRI because they had erosion of the lateral semicircular canal and the sigmoid sinus.



**Figure 1 :** CT scan of the right temporal bone (coronal view) showing the erosion of the inferior wall of EAC.



**Figure 2 :** A: CT scan of the left temporal bone (axial view), B: (coronal view): Erosion of different walls of the sigmoid sinus



**Figure 3 :** A :CT scan of the right temporal bone (axial view), B: CT scan of the right temporal bone (coronal view) showing irregular ovoid bony masses with soft tissue density of the middle ear with extension into the mastoid cells.

For primary EACC, one case was treated through transmeatal approach : excision of the keratin debris and the eroded bone and sacrificing the pathologic skin. Five patients underwent surgical treatment through a post auricular approach: tympanoplasty in one case, modified radical canal wall down mastoidectomy in three cases and canal wall down mastoidectomy in one case. During these procedures, extensive invasion into the mastoid cavity was confirmed in 2 cases, erosion of the fallopian canal in one case reconstructed with temporalis fascia and erosion of the lateral semi circular canal in one case. The ossicular chain was noted to be normal and mobile in 3 cases and absent in 3 cases.

For the patient who had secondary EACC, excision of the osteoma and the cholesteatoma was performed through the post auricular approach. During mastoidectomy, the tumor was removed in bloc along with the overlying skin and a huge cholesteatoma found in the EAC. The stapes was absent, the cortical bone of the facial canal and lateral semicircular canal were intact. Based on examination of the resected specimen, the pathologic diagnosis was osteoma. To reconstruct the cavity, Conchal cartilage with temporalis fascia was used in one case and tragal cartilage associated with temporalis fascia for the other 5 cases.

Follow-up ranged from 6 months to 3 years (mean 1.4 year). The patient who has secondary EACC did well and there has been no evidence of recurrence or complications 6 months post operatively. However external auditory canal



stenosis was noted after 3 months of follow-up in one case of a primary EACC treated by dilatation using expandable ear sponges with good outcome.

## DISCUSSION

EACC is an uncommon condition with an annual incidence rate in the general population of 0.15 cases per 100 000 individuals and an estimated incidence rate of 0,1 à 0,5% of otologic pathology. EACC affects all ages and more common in elderly patients. The age of our patients ranged from 20 to 77 years. There isn't sex predilection reported in the literature [1,2,3].

Histo-pathologically, EACC is characterized by a dense plug of keratin debris located within the inferior portion of the bony external auditory canal associated with periosteitis in the eroded area, as well as osteitis, bone sequestra, and reactive inflammatory tissue [3,4].

The aggressivity of cholesteatoma consisted on extensive involvement of neighboring structures: facial nerve canal, semicircular canals, sigmoid sinus, temporomandibular joint and encephala. Holt divided EACC patients into 5 groups based on cause: postsurgical, posttraumatic, congenital ear canal stenosis, ear canal obstruction, and spontaneous [5,6]. In this study, the majority of cases were spontaneous and there is only one case related to ear canal obstruction by an osteoma.

The pathogenesis of EACC remains not well understood. Many risk factors are put forward for the secondary cholesteatomas, but in primary lesions or spontaneous lesions which are more common, no previous auditory canal insult is found. In this form 'keratinisation in situ' was reported in many studies, due to reduced migratory capacity of the canal epithelium, occurring with age [3,7], others theories were advanced, localized periostitis, chronic inflammation of the EAC, dehiscence of the petrotympanic fissure [4].

Disruption of the local microcirculation by micro trauma from use of cotton swabs, smoking, or diabetes mellitus are also suggested [7,8,9].

The differential diagnosis of EACC includes keratosis obturans (KO), malignant otitis externa (MOE), malignant tumors of EAC and post-inflammatory medial canal fibrosis [3], the KO is the most similar entity sharing with EACC many characteristics: both are rare conditions, characterized by accumulation of keratin debris, their etiology and pathogenesis remain unclear [10]. The KO is usually bilateral and more common in less elderly patients whose main complaints were severe otalgia and acute conductive hearing loss, rarely otorrhea. Patients with KO typically show pathologic tympanic membrane and widened ear canal from circumferential involvement of bone, In contrast, EACC is characterized by erosion commonly of the posterior and inferior walls.

Symptoms at presentation are not specific and the diagnostic latency can be long. Asymptomatic forms are noted in literature with a percentage between 25 and 31% [1]. Patients present most commonly one sided otorrhea with often otalgia described as dull pain or discomfort related

probably to the extent of disease [1]. Other symptoms are described as hearing loss, tinnitus, facial paralysis appear rarely [4,7,9].

The appearance on CT results in an oval soft tissue mass in the EAC, eroding the underlying bony walls. Typical location is at the bone-cartilaginous junction of the canal. The most important feature is presence of tiny hyperdensities within the soft tissue, probably representing bony fragments or sequestra [3,11,4]. The CT scan with axial and sagittal reconstructions is also the investigation of choice for assessment of extension of the disease, and for pre-operative planning. In fact, the treatment is determined by the degree of bone necrosis and bony erosion [9]. There is a classification based on CT data extension with a simple system that comprises 4 stages: stage I for cholesteatoma of the external canal only, stage II for extension to tympanic membrane or middle ear, stage III for invasion of mastoid air cells and stage IV for lesions beyond the temporal bone [3,11]. MRI is more specific than CT by demonstrating increased signal intensity in diffusion-weighted imaging (DWI) for cholesteatomas.

Therapeutic approaches used depend on the extent of the disease and whether it is considered as a primary or secondary cholesteatoma [12,13]. Two strategies are suggested: conservative procedure or surgical approach. Conservative treatment consists on topical antibiotic preparations associated with repeated aspiration of the epidermic debris and sequestered bone in the cholesteatoma cavity under local anesthesia [7, 11, 13]. It's preferred for very limited lesion, when the entire extension can be visualized on examination and when the patient has no pain; as an expectative procedure, to assess the evolutive potential of the cholesteatoma; and in patients with a general contraindication for surgery or who refuse surgery [13].

Surgery is usually used for patients with secondary EACC. The aim of surgery is to excise the cholesteatoma and to restore a smooth, self-cleaning canal wall epithelium to prevent progression and continued erosion [14], usually with the aid of cartilage and fascial grafts to protect denuded structures, and fill any canal wall defect. The deepest pockets require a canaloplasty by removing diseased skin and bone and exteriorizing the recess [14]. However a modified radical mastoidectomy may be indicated if the mastoid air cells are invaded [4,7,14].

For patients with EACC and osteoma, regular follow-up may be sufficient for the small osteoma. However surgery should be performed in the case of a growing osteoma threatening the epithelial migration [15]. Surgery excision is also recommended for patients complaining of a recurrent infection and conductive hearing impairment [15,16].

With surgical treatment, good outcomes can be achieved with good hearing [9]. In the absence of surgical intervention, clinical monitoring with regular follow up and CT scan is necessary to monitor patients for potentially serious complications [7].



## CONCLUSION

EACC is a rare entity with characteristic imaging and clinical features. A soft-tissue attenuating mass in the EAC with erosion of adjacent bone defines the CT presentation of an EACC. The EACC may exhibit features including deep extension into the middle ear, mastoid, facial nerve canal, or

the tegmen tympani. These findings may be unrecognized at clinical examination. These features may influence the clinical and surgical management of EACC.

**Conflicts of interest:** Authors declared no conflicts of interest.

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