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Original Research Article

External auditory canal cholesteatoma: classification and management

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ABSTRACT

Background: External auditory canal cholesteatoma (EACC) is a disease characterized by the accumulation of keratin in the ear canal and erosion of the bony wall. Etiologically, it may be primary or secondary. It presents with ear discharge and pain. Distinction from similar conditions is crucial for appropriate treatment. This study evaluates the EACC. Its purpose is to suggest categories based on disease extent. This study aimed to evaluate the presentation, diagnosis, and treatment of EACC. The goal was to classify EACC based on disease severity and propose treatment approaches.
Methods: A retrospective review of medical records for patients diagnosed with EACC over ten years. Clinical data were examined for the presentation, clinical findings, etiology, and treatment. We simplified the approach by dividing the disease into three categories: Category 1, confined to the ear canal; Category 2, eroded the canal wall; Category 3, extended to adjacent structures.
Results: 18 patients were included in the study. Ear discharge and otalgia were the most common symptoms. Etiological classification of the disease identified most as secondary EACC. Two cases were assigned to Category 1, six to Category 2, and ten to Category 3.
Conclusions: Common symptoms of EACC are ear discharge, pain, and hearing loss. Our study categorizes the condition into three categories based on severity, extension, and treatment strategies. Most cases were assigned to category 3 and extended into adjacent structures.

Keywords: Ear canal, Clinical findings, Ear discharge, Classification, Hearing loss, Extension

INTRODUCTION

External Auditory Canal Cholesteatoma (EACC) is an External Auditory Canal (EAC) disease characterized by the accumulation of keratin in the EAC and progressive erosion of the bony wall.[1-3] Primary symptoms include unilateral ear discharge and pain caused by the invasion of squamous tissue into a localized area of periostitis in the canal wall.[1] This last characteristic is essential to remember as it differentiates it from a similar condition, Keratosis Obturans (KO), in which there is no erosion of the underlying bone wall.[1,4,5] This distinction is essential as KO is treatable with topical medication, while EACC may require surgery in more extreme cases.[1] Of the various types of EAC diseases, EACC is considered uncommon, with an estimated incidence of 1:600 patients.[3] EACC is divided into primary (idiopathic) and secondary EACC.[6] Primary EACC is the rarest form, and the invasion and damage commonly occur on the floor of the auditory canal. Secondary EACC is usually categorized depending on what is brought on the condition. These categories are post-stenotic, post-surgical, post-traumatic, radiogenic, post-inflammatory, and post-obstructive.[6]

Table 1: Suggested categories of EACC.

Category	Description	Treatment Options
Category 1	Disease is confined to the EAC with localized inflammation and/or exposed bone. No erosion of the bony EAC	Serial debridement in the clinic with applications of topical otic medications.
Category 2	Erosion of The EAC wall, No extension into adjacent structure	
a	Erosion of one canal wall	Serial debridement in the clinic and application of topical otic medications.
b	Erosion of two or more canal walls	Canaloplasty
Category 3	Extension of the cholesteatoma into adjacent structure	CWU, CWD, Canaloplasty. Reconstruction of the Eroded canal wall.
a	Not invade FN canal or tegmen	-
b	Involvement of FN canal, (±FN Palsy)	FN monitoring, (±FN decompression)
c	Tegmen invasion or intracranial complication	Neurosurgery referral Treat intracranial complication if present

Holt et al described the disease in three categories: first, the appearance of a superficial defect, followed by the formation of a localized canal pocket and ending in the extension of the damage into the mastoid.[7] Later, Naim et al built on this division by adding histopathological findings, ending with four classes: first, there is hyperplasia of the canal epithelium, followed by periostitis, progressive bone canal defects and erosion of adjacent anatomic structures.[2] Treatment of EACC depends on the extension of the disease.[4,7] Mild conditions can be treated conservatively with serial debridement and application of topical antibiotics and anti-inflammatory ointments.[4,7,8] More severe cases often require surgical treatment, the precise nature of which depends on the extent to which the condition has spread.[4] These procedures range from less invasive canaloplasty to more extensive canal wall-up (CWU) and canal wall-down (CWD) surgeries.[4,7,9] This study evaluates the presentation, diagnosis, and treatment of EACC at a tertiary hospital in Saudi Arabia. The aim is to assess the classification and treatment approaches based on disease severity.

METHODS

This retrospective observation study was conducted at King Abdulaziz University Hospital, Saudi Arabia. The study reviewed medical records of patients diagnosed with EACC over ten years, from October 2010 to September 2020.

Inclusion and exclusion criteria

We included patients with a confirmed diagnosis of EACC based on clinical examination (with findings of keratin in EAC) and the CT scan of the temporal bone (with findings of soft tissue density or bony erosion in EAC). Patients with no clinical findings of EACC on examination and CT scan images, history of middle ear cholesteatoma, previous history of canaloplasty or mastoidectomy, and patients who refused investigations or treatment were excluded from this study.

A detailed review of patient files was conducted, including a comprehensive analysis of medical records and CT scans. Demographic data such as age, gender, and nationality were collected, along with medical history that included presenting symptoms and duration, such as ear discharge, ear pain, hearing loss, and fullness in the ear. Etiological factors such as history of trauma, ear cleaning, ear surgery, and radiation exposure were also considered. The examination findings were also noted, including the presence and location of keratin and erosion of the EAC wall, local inflammation, extension of the disease, condition of the surrounding structures, widening or narrowing of the EAC, presence of discharge, aural polyps or granulation tissues, and assessment of facial nerve (FN). CT temporal bone findings were analyzed, including soft tissue density in EAC, canal wall erosion or destruction, and involvement of surrounding structures such as the middle ear, temporomandibular joint (TMJ), mastoid, FN, and tegmen.

The results of hearing assessments were also taken into consideration. The management of each case was recorded, which included frequent cleaning, debridement, canaloplasty, or mastoidectomy (CWU or CWD). This was evaluated further by breaking down each case by etiological classification, each with prevalence and clinical, radiological, and hearing assessment findings and treatment. With these data and literature review, this study aimed to support the development of a system for categorizing EACC that considers the kind of treatment required, how urgent it is, and possible prognosis. Based on clinical and imaging findings of bone involvement and disease extension, we categorized the disease into three main and several subcategories.

The suggested categories would be as follows: Category 1 is the category in which the cholesteatoma is confined to the EAC, with localized inflammation or exposed bone, but no significant erosion is evident by examination or imaging. Category 2 is bony erosion in the EAC but without extensions into adjacent structures. It is further subdivided depending on the level of erosion. If only one

wall is affected, the disease is classified as 2a. If more than one wall is involved, it is classified as 2b. Category 3 was defined as the presence of extensions of the cholesteatoma into adjacent structures. This was further subdivided depending on the level of extension. If the cholesteatoma does not invade the tegmen or FN canal, it is termed 3a. If the FN Canal is eroded, the disease is classified as 3b and requires immediate medical attention to prevent FN palsy. If the tegmen is invaded and extension is present into the Dura, the condition is termed 3c and requires the most urgency in treatment (Table 1).

The obtained data were analyzed using basic descriptive statistics with the assistance of Microsoft Excel software. Qualitative data were presented in the form of numbers and percentages, while quantitative data were expressed as mean and standard deviation (mean±SD).

RESULTS

Of all the records reviewed, 18 patients met the inclusion criteria (N=18). Analysis of demographic information showed the average age to be 34, and two-thirds of patients were females (N=12; 66.6%). All patients were Saudi (Table 2). Analysis of records for these patients revealed ear discharges as the most common symptom (n=16; 88.9%), followed by ear pain (N=10; 55.6%), and unilateral hearing loss (N=9; 50%). Other less common symptoms included aural fullness (N=2; 11.1%), dizziness (N=2; 11.1%) and tinnitus (N=1; 5.6%).

Table 2: Distribution of studied patients according to their demographic characteristics, affected side, and duration of illness.

Variables	N (%)
Age (years) (mean±SD)	34±16.4
Gender	
Male	6 (33.3)
Female	12 (66.7)
Nationality	
Saudi	18 (100)
Other	0 (0)
Affected ear	
Right	10 (55.6)
Left	8 (44.4)
Duration (years)	4.5±2.3

On average, patients reported suffering from symptoms for approximately five years before hospital admission. Concerning the disease, the right ear was the most affected (N=10; 55.6%). Etiological classification of the disease identified most as secondary EACC (N=11; 61.1%). Of these, 36.4% (N=4) were post-traumatic, 45.5% (N=5) were post-inflammatory, one case (9.1%) was post-surgical, and one case (9.1%) was post-radiation (Table 3).

Table 3: Distribution of EACC according to the etiology.

Etiology	N	%
Primary (idiopathic)	7	38.9
Post-Inflammatory	5	27.8
Post- Traumatic	4	22.2
Post-Surgical	1	5.6
Post-Radiation	1	5.6
Total	18	100

Primary EACC

Patients in this category had no clear etiology, although risk factors were identified for three patients in the form of a prior history of ear microtrauma. Keratin was present in all cases (N=7), with 71.4% (N=5) still presenting discrete patches on the floor, anteroinferior, anterosuperior, or posterior wall of the EAC. In the remaining two cases, the EAC was completely occluded. EAC erosion was found in 85.7% (N=6) cases, with the exact locations being spread amongst the floor (N=3), anterior wall (N=3), posterior wall (N=4), and the Scutum (N=1). The remaining case had no detectable erosion during examination or CT scan, but there was localized inflammation and necrosis. Only 57.1% (N=4) had extended beyond the EAC into the mastoid (N=4), temporomandibular joint TMJ (N=2), the vertical segment of the FN (N=2), and hypotympanum (N=1). In the case where the Scutum was eroded, the tympanic membrane was intact and pushed medially by the disease. In both cases where the FN canal had been breached, no paralysis was identified. The EAC was wide in 42.9% (N=3) and edematous in 28.6% (N=2) cases. Granulation tissue was identified in 28.6% (N=2) cases; the tympanic membrane was intact in 71.4% (N=5) cases, mildly retracted in one case, and perforated in another. Mild conductive hearing loss (CHL) was reported in 71.4% (N=5) of cases. Surgery was performed on 71.4% (N=5) of patients, and the remaining two were treated conservatively.

Secondary EACC

Post-traumatic: Four cases were post-traumatic, with the mechanism of trauma being identified as a motor vehicle accident in two cases and a fall in the other two. All four cases had keratin deposits, with two localized to the site of erosion and the remaining two fully occluded. The bony EAC was eroded in all cases. Erosion was present in all cases, with locations on the anterior wall (N=2) and posterior wall (N=4), and extensions being detected in all cases into the mastoid (N=3), TMJ (N=1), Attic (N=1), and FN canal (N=1).

Table 4: Distribution of EACC cases according to the etiology and the category of the disease.

Category	Total N (%)	Primary N (%)	Post-Infectious N (%)	Post-Traumatic N (%)	Post-Surgical N (%)	Post-Radiation N (%)
1	2 (11.1)	1 (5.6)	-	-	-	1 (5.6)
2	6 (33.3)	2 (11.1)	4 (22.2)	-	-	-
2a	5 (27.8)	1 (5.5)	4 (22.2)	-	-	-
2b	1 (5.6)	1 (5.6)	-	-	-	-
3	10 (55.6)	4 (22.2)	1 (5.5)	4 (22.2)	1 (5.5)	-
3a	6 (33.3)	1 (5.6)	1 (5.6)	3 (16.7)	1 (5.6)	-
3b	4 (22.2)	3 (16.7)	-	1 (5.6)	-	-
3c	-	-	-	-	-	-

The EAC was narrow in two cases (50%). Granulation tissue was identified in only one case (25%), and the tympanic membrane was intact in two cases (50%), thin in one case (25%), and perforated in the last (25%). All four cases reported mild to moderate CHL. All four cases were treated surgically. Post-Inflammatory: Most secondary EACC patients had a history of chronic (N=4) or recurrent (N=1) ear infections, with three cases reporting a concurrent inflammatory polyp in the EAC. One patient had undergone a polypectomy. Keratin was identified in all cases, occluding in two cases (40%) and localized in three cases (60%) to posterior, posteromedial, and superior walls. Erosion was present in all cases and located on the posterior wall (N=2), posterosuperior wall (N=1), floor (N=1), and Scutum (N=1). There was an extension into the mastoid in one case.

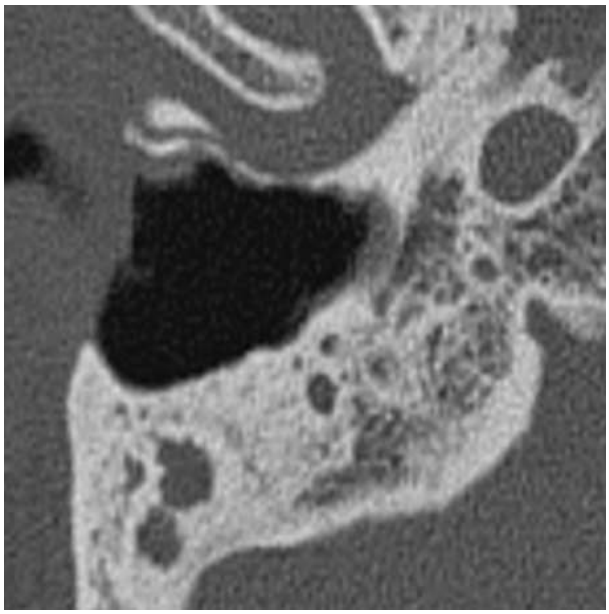

Figure 1: Temporal bone axial CT image showing EACC with erosion of the anterior and posterior walls of the EAC.

The EAC was narrow, with accompanying granulation tissue in one case (20%). The tympanic membrane was normal in four cases (80%) and thin in one (20%). Four patients (80%) reported mild to moderate CHL, and one (20%) had moderately severe CHL. Three cases (60%) were treated surgically, while the remaining two were treated conservatively. Post-Surgical: Only one case was identified as post-surgical. In this case, the patient underwent tympanoplasty twice in the same ear, the last of which had been performed nine years ago, and after which the patient began suffering from unilateral ear pain and hearing loss. The EAC was narrow, the tympanic membrane was perforated, and the patient reported moderate CHL. The CT scan showed opacity in EAC, extending into the middle ear cavity with erosion to the posterior wall of EAC. The patient underwent Canaloplasty and Meatoplasty, during which a fibrous band in the lateral EAC was identified, along with a thick keratin deposit occluding the EAC and extending into the middle ear through the perforated tympanic membrane.

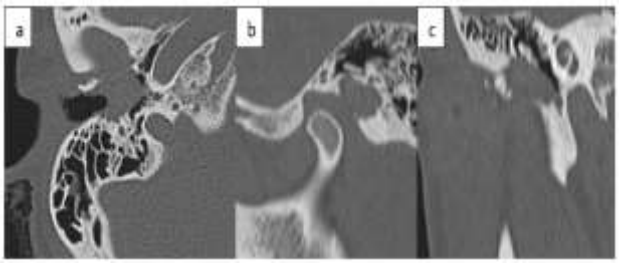

Figure 2: Temporal bone CT images showing EACC with multiple wall erosion and extension to TMJ, mastoid air cells, and FN canal, a) axial CT image shows erosion of the anterior and posterior walls of the EAC with involvement of the TMJ, mastoid air cells, and FN canal; b) sagittal CT image shows erosion of the anterior and posterior walls of the EAC with involvement of the TMJ, mastoid air cells, and FN canal; c) coronal CT image shows erosion of the EAC floor.

Post-radiation: Only one record was identified as post-radiation. The patient underwent radiotherapy to treat a nasopharyngeal carcinoma when he presented with unilateral purulent, foul-smelling discharges with associated ear pain and hearing loss. There were keratin flakes over the denuded bone on the floor of EAC. There was no evidence of erosion or extensions beyond the EAC. The patient was treated by conservative means.

Extension of EACC

We suggested categorizing the cases based on bony involvement and disease extension. We divided the disease into three categories to simplify the treatment decision and urgency of the treatment (Table 4). Only two cases (11.1%) were included in category 1. All cases were treated by conservative means (debridement, etc.) on regular visits to the clinic.

Six cases were assigned to Category 2. These were further divided into Five in 2a and one in 2b. Two category 2 cases required surgical intervention (polypectomy and canaloplasty) due to associated polyps in the EAC. The other three cases were treated conservatively by frequent cleaning and the application of antibiotic drops. The category 2b case was primary EACC, with erosion on the anterior and posterior walls (Figure 1). Surgery was required in the form of a Canaloplasty and cholesteatoma removal. The remaining ten cases (55.6%) belonged to category 3, subdivided into six in 3a and four in 3b (Figure 2, 3). There was no evidence of cases fitting the 3c category.

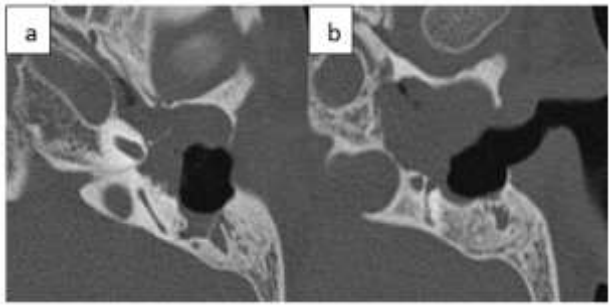

Figure 3: Temporal bone CT images showing EACC with posterior wall erosion and extension to mastoid air cells and FN canal, a) axial CT image showing erosion of the posterior wall of the EAC with involvement of the mastoid air cells; b) Axial CT image showing the extension to the FN canal.

All six 3a cases required surgical intervention, the exact procedure depending on the location and degree of the extensions; four cases were treated with mastoidectomy; three with CWD mastoidectomy; one with CWU Mastoidectomy, with concurrent reconstruction of the eroded posterior canal wall; one with canaloplasty; and the final case with an atticotomy. The four cases assigned to category 3b were treated by CWD mastoidectomy and canaloplasty. Monitoring of the FN was required in two cases, one of which developed post-operative FN palsy. This palsy was delayed and partial and was treated with systemic steroids; the condition was cured within a week of treatment.

DISCUSSION

External auditory canal cholesteatoma is a disease characterized by the accumulation of keratin in the EAC and progressive erosion of the bony wall.[1-3] Of the various types of EAC diseases, EACC is considered uncommon, with an estimated incidence of 1:600 patients.[3] Here, we attempted to evaluate the presentation, diagnosis, and treatment of EACC.

Presenting symptoms

According to published literature, the main symptoms of EACC are longstanding ear discharge and pain.[1,4,7] Our study agrees with these facts, with the highest reported symptom being ear discharge, followed by pain. We also had evidence of unilateral hearing loss, aural fullness, dizziness, and tinnitus. Although the symptoms are not primary, they are expected in a disease that erodes and occludes the ear canal and causes significant damage to other structures and tissues.

Location and extension

According to most published studies, primary EACC is mainly located on the floor of the EAC, while secondary EACC has a more randomized distribution and is often multifocal.[4] In this study, both primary and secondary EACC favored a multifocal presentation. EACC can extend into one or more adjacent structures, including the mastoid, TMJ, middle ear, FN canal, and Tegmen.[2,4,10] Most of our studied cases had one or more extensions into adjacent structures for both primary and secondary EACC. This is in full accordance with published literature (Table 5).

Table 5: Distribution of cases according to the extensions of the disease.

Category	Extension	N
3a	Mastoid	2
	TMJ	1
	ME	1
	Mastoid + ME	2
3b	Mastoid + FN	2
	Mastoid + FN + TMJ	1
	Mastoid + TMJ + ME + FN	1

Describing EACC in a series of predictable categories was first done by Holt in which he named three discrete classes: formation of a superficial defect, followed by a localized canal pocket, and lastly, an extension into the mastoid.[7] The flaw in this suggested system was the omission of extensions into other adjacent structures. This led Naim to suggest a different system, using histopathological differences as well as physical changes in which the presentation was divided into four classes: first hyperplasia of the canal epithelium, then periostitis, followed by defects in the bony canal; and finally, erosion of adjacent anatomic structure. [2,4] Upon inspections, this system had a few flaws, namely that the first two classes could not be easily differentiated without a microscopic examination and that it didn't differentiate between minimal extension into, say, the mastoid and extension to

the intracranial cavity through the Tegmen, which may result in long-standing neurological sequelae and necessitate an urgent intervention. Here, we attempted to establish categories that consider the severity and type of damage to determine which treatments to administer, how urgent they are, and what the expected outcome might be (Table 1).

Category 1 was defined as a category in which the cholesteatoma is confined to the EAC, with localized inflammation or exposed bone but no significant erosion evident by examination or imaging. Options for treatment in this category are serial debridement with applications of topical antibiotics and anti-inflammatory medications.[1,4,7,8] Category 2 was bony erosion in the EAC but without extensions into adjacent structures. It is further subdivided depending on the level of erosion. If it is confined to one wall, the disease is classed as 2a. If more than one wall was affected, it was classed as 2b. Category 2a treatment involves serial debridement, topical antibiotics, and anti-inflammatory meds. Canaloplasty may be necessary if conservative treatment fails or follow-up visits are uncertain. For category 2b, canaloplasty is recommended.[4,9] Category 3 refers to the presence of extensions of the cholesteatoma into adjacent structures. This category is further divided into two subcategories based on the extent of the extension. If the cholesteatoma does not invade the tegmen or the FN canal, it is classified as 3a. If the FN canal is eroded, the disease is classified as 3b, which requires more urgent medical attention to prevent FN palsy. If the tegmen is invaded and an extension is present into the Dura, the condition is termed 3b and requires the most urgent treatment.

Treatment options for 3a are always surgical, but the exact technique (CWD or CWU mastoidectomy or Canaloplasty) depends on the location and degree of the extension, associated diseases, and personal preference.[4,7,11] Treatment options for 3b depend on whether FN palsy is present and, if so, to what degree. If no palsy is present, treatment options are identical to class 3a. If palsy is present, additional FN decompression is required.[12,13] It is essential to highlight the importance of Intra-operative FN monitoring for patients in this category as there is a high likelihood of FN injury.[14,15] We did not encounter 3c category patients in our study, which is not entirely unexpected as this degree of EACC is rare. However, treatment of such a patient would be urgent and complicated, most likely requiring the aid of a neurosurgeon.[16,17]

Limitations

The study has some limitations, as it is retrospective and relies on the analysis of medical records, which may contain incomplete or missing information. Also, because EACC is a rare disease, the study only includes a small sample size.

CONCLUSION

The most common presenting symptoms are ear discharge, pain, and hearing loss. Our study attempted to reclassify the condition into three categories that would consider the urgency and lines of treatment. We found our suggested categories to be adequate. Most of our studied cases assigned to category 3 as had one or more extensions into adjacent structures.

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