Clinical characteristics and follow-up of patients with external ear canal cholesteatoma treated conservatively

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Abstract

We conducted a retrospective study to evaluate the clinical properties and follow-up of patients with external ear canal cholesteatoma (EECC) who were treated conservatively. Our study group was made up of 15 patients—6 men and 9 women, aged 21 to 82 years (mean: 48). In addition to demographic data, we compiled information on presenting signs and symptoms, the location of the lesion, treatment, and follow-up. All EECCs occurred spontaneously, and all were limited to the temporal bone. Lesions were left-sided in 7 patients, right-sided in 7, and bilateral in 1. For purposes of defining the location of the lesions, the ear canal was divided into four quadrants: anterior, posterior, superior, and inferior. Temporal bone computed tomography (CT) and otomicroscopic evaluation revealed that 7 of the 16 lesions (44%) were located in the anterior and inferior quadrants, 6 (38%) in the inferior quadrant only, 2 (13%) in the anterior, inferior, and posterior quadrants, and 1 (6%) in the anterior quadrant only. Otorrhea was present in 7 of the 15 patients (47%), otalgia in 6 (40%), itching in 4 (27%), fullness in 2 (13%), and occlusion in 1 (7%); 3 patients (20%) were asymptomatic. All patients were treated with local debridement and aspiration under otomicroscopy, and they were followed up with repeat aspirations approximately every 10 weeks under microscopy. The duration of follow-up ranged from 6 to 75 months (mean: 41). At study's end, cholesteatoma had not progressed in any patient during follow-up, indicating that repeat aspirations and regular follow-up of limited EECC prevent recurrence of signs and symptoms and progression of the disease.

Introduction

External ear canal cholesteatoma (EECC) is a rare clinical condition first described by Toynbee in 1850.[1] Its estimated incidence is 1.2 per 1,000 new otolaryngology patients.[2]

EECC is made up of a cholesteatoma matrix and keratin debris; it closely resembles middle ear cholesteatoma. It can manifest as otorrhea, itching, hearing loss, fullness, occlusion, and mild to severe pain; on the other hand, some patients are asymptomatic. The diagnosis is based on the history and physical examination. Computed tomography (CT) of the temporal bone is useful for determining the extent of disease.

In some cases, EECC is mistaken for otitis externa, keratosis obturans, and a cerumen plug obstructing the ear canal. It might also be mistaken for an inflammatory cast of the tympanic membrane after otitis media, even if it affects the tympanic membrane, because signs and symptoms are not specific or severe enough to warrant concern after removal of the obstruction.[3,4] The most useful finding confirming an EECC is focal osteonecrosis or sequestration of bone that lacks an epithelial covering. Bony destruction can extend into the middle ear, mastoid bone, and temporomandibular joint. Involvement of the facial nerve, labyrinth, sigmoid sinus, and dura of the tegmen may also be seen.[3]

Treatment of EECC ranges from repeated cleanings with aspiration to surgical excision of the lesion with or without a meatocanaloplasty. Also, a temporalis fascia or a split-thickness skin graft can be used to cover the exposed tympanic bone. Whether or not surgical treat-

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ment is undertaken depends on the nature and severity of the signs and symptoms and the extent of the disease. In cases limited to the temporal bone, a conservative approach is usually recommended.[3,5-8]

In this article, we describe our review of the clinical properties and follow-up results of patients with EECC who were treated conservatively.

Patients and methods

We retrospectively reviewed the records of all patients who had been diagnosed with spontaneous EECC in the Department of Otorhinolaryngology at Safa Private Hospital in Istanbul. Our definition of EECC was primarily clinical. A total of 15 patients—6 men and 9 women, aged 21 to 82 years (mean: 48)—met our study qualifications. Seven of these patients had left-sided EECC, 7 right-sided, and 1 bilateral, for a total of 16 lesions (table 1). None of the patients had described ear trauma, previous ear surgery, or any disease related to the middle ear or mastoid bone.

For purposes of defining the location of the lesions, the ear canal was divided into four quadrants: *superior, anterior, inferior,* and *posterior.* The area between 10 o'clock and 1 o'clock was considered the superior quadrant, the area between 1 o'clock and 4 o'clock the anterior quadrant, the area between 4 o'clock and 7 o'clock the inferior quadrant, and the area between 7 o'clock and 10 o'clock the posterior quadrant. The location of the lesions was identified by temporal bone CT and otomicroscopy.

In all cases, there was bony erosion filled with keratin debris and surrounded by granulation tissue at its edge, which was limited to the temporal bone. The diagnosis was further supported by histology, which revealed keratinized debris containing various amounts of sequestered bone.

In addition to demographic data, we compiled information on presenting signs and symptoms, the location of the lesion, treatment, and follow-up.

Results

At presentation, otorrhea was seen in 7 of the 15 patients (47%), otalgia in 6 (40%), itching in 4 (27%), fullness in 2 (13%), and occlusion in 1 (7%); 3 patients (20%) were asymptomatic (table 2).

Bony erosions ranging from 5 to 15 mm were seen just lateral to the tympanic sulcus in 8 of the 16 ears (50%). Seven of the lesions (44%) were located in the anterior and inferior quadrants, 6 (38%) in the inferior quadrant only, 2 (13%) in the anterior, inferior, and posterior quadrants, and 1 (6%) in the anterior quadrant only (table 1).

Table 1. Summary of clinical data in the study group

Pt.	Age/sex	Side	Location	Follow-up (mo)
1	66/F	Left	Anterior, inferior, posterior	75
2	62/M	Right	Anterior, inferior	72
3	24/F	Right	Anterior, inferior, posterior	63
4	57/F	Left	Inferior	55
5	21/F	Left	Anterior, inferior	52
6	23/F	Bilateral	Inferior	51
7	34/F	Right	Anterior, inferior	44
8	70/F	Right	Inferior	40
9	82/M	Right	Anterior, inferior	34
10	30/M	Left	Inferior	34
11	51/M	Right	Anterior	31
12	35/F	Right	Anterior, inferior	28
13	44/M	Left	Anterior, inferior	19
14	75/F	Left	Anterior, inferior	11
15	46/M	Left	Inferior	6

In all patients, granulation with crusts in the external ear canal was removed with debridement via otomicroscopy, and the eroded bony cavity was repeatedly cleansed by aspiration to remove keratinized debris. The granulation tissue was sent to pathology to confirm the histopathologic diagnosis. A topical antibiotic/steroid agent (ciprofloxacin/dexamethasone) was used to treat infection when needed. This treatment was continued until the external ear canal was dry. Cleansing with aspiration under otomicroscopy was performed in all patients approximately every 10 weeks.

During a follow-up that ranged from 6 to 75 months (mean: 41), no patient experienced a worsening of his or her condition, indicating that repeat aspirations and regular follow-up of limited EECC prevents recurrence of signs and symptoms and progression of the disease.

Discussion

EECC is a very uncommon entity.[2] It is characterized by the invasion of squamous cell epithelium into localized areas of bone in the external ear canal.[5] Patients present with nonspecific external ear canal complaints, including otorrhea, itching, hearing loss, fullness, occlusion, and mild to severe pain; on the other hand, some patients are asymptomatic. Because of the nonspecificity of the signs and symptoms, EECC is often not diagnosed until the lesion extends into the surrounding anatomic structures outside the temporal bone and becomes symptomatic.

EECC has been subclassified into six types: *congenital, post-traumatic, iatrogenic, postobstructive, postinflammatory,* and *spontaneous,* depending on the etiology. The spontaneous type is defined as an EECC of an unknown cause.[5] In previous studies, spontaneous EECC has been attributed to the use of ear-cleaning sticks, hearing devices, and smoking.[3] In addition, many authors have proposed that a decrease in the migration capacity of the epithelium in the inferior external ear canal leads to in situ keratinization, which in turn leads to the development of EECC.[9,10]

Makino and Amatsu demonstrated slower migration rates in the inferior wall in patients with EECC and suggested that this could be due to hypoxia arising from a poor blood supply.[11] Because no specific etiologic factor was identified in our patients, they were all considered to have the spontaneous type.

Staging systems. Naim et al staged EECC according to histologic and clinical findings.[10] They identified four stages:

Table 2. Summary of signs and symptoms in the study group

Pt.	Sign or symptom
1	Otorrhea, itching
2	None
3	Otalgia, fullness
4	Otorrhea
5	Otalgia, occlusion
6	Itching (bilateral)
7	Otorrhea, otalgia
8	Otalgia, fullness
9	None
10	Otorrhea
11	None
12	Otorrhea, itching
13	Otorrhea, itching
14	Otorrhea, otalgia
15	Otalgia

- stage I: the presence of hyperplasia of the canal epithelium;
- stage II: the presence of periosteitis;
- stage III: the presence of a defective bony canal; and
- stage IV: the presence of erosion of adjacent structures.

Some previous reports have shown that EECC is often more extensive than what may be suggested by the clinical findings.[12] Naim et al recommended high-resolution CT of the temporal bone, since the defective bony canal contains a pocket that cannot be assessed on clinical examination.[10]

After we diagnosed our patients clinically and histopathologically, we ordered temporal bone CT in all cases to evaluate the surrounding anatomic structures and to determine the depth of bone erosion; these factors affected our treatment approach.

Another four-stage classification system was developed by Shin et al.[13] They based their staging on the results of temporal bone CT and clinical findings:

- stage I: the EECC is limited to the external ear canal;
- stage II: the EECC invades the tympanic membrane and middle ear;
- stage III: the EECC creates a defect of the external ear canal and involves the air cells in the mastoid bone; and

• stage IV: the EECC extends beyond the temporal bone.

In our patients, CT showed that the lesions in all cases were limited to the temporal bone, and thus they were classified as stage I according to the Shin et al system.

Age and sex. The age range and mean age of our patients—21 to 82 years (mean: 48)—is consistent with those reported by others. Naim et al[10] reported a range of 23 to 74 years (mean: 50), and a group studied by Darr and Linstrom[14] had an age range of 20 to 75 years (mean: 50).

The female-to-male ratio in our study was 9:6. Anthony and Anthony[2] reported a female-to-male ratio of 7:5, and Sismanis et al[15] reported a ratio of 4:6. These numbers, while small, indicate that EECC apparently has no predilection for either sex.

Laterality. In our study, the lesion was left-sided in 7 patients, right-sided in 7, and bilateral in 1. A left-sided predominance was reported by Anthony and Anthony,[2] and a right-sided predilection by Sismanis et al.[15] In the study by Darr and Linstrom, the lesion was left-sided in 5 patients, right-sided in 3, and bilateral in 1.[14]

Bilateral involvement was seen in 3 of 48 patients studied by Owen et al[9] and in 2 of 12 studied by Anthony and Anthony.[2] Thus, the overall incidence of bilateral involvement, including our finding, was 8% (7/84).

Location. In our study, inferior, anterior, and posterior wall involvement was seen in 94% (n = 15/16), 63% (n = 10), and 13% (n = 2) of ears, respectively. Involvement of the superior ear canal was not found in any patient. According to most studies, spontaneous EECC is typically located in the inferior quadrant.[2,10,14] Darr and Linstrom found that the inferior wall was involved in 80% of their cases.[14]

Signs and symptoms. The most common signs and symptoms of EECC reported in previous studies were otalgia and otorrhea, with the incidence ranging from 25 to 100%.[3,9,14] In our study, otorrhea (47%; n = 7/15) and otalgia (40%; n = 6) were the most common symptoms, as well.

Previous studies have shown that the incidence of itching, occlusion, hearing loss, and fullness ranged between 0 and 25%.[5,9,15,16] In our study, itching, fullness, and occlusion occurred in 27% (n = 4/15), 13% (n = 2), and 7% (n = 1) of patients, respectively. Because our patients all had been diagnosed in the early stage of disease, none of them described hearing loss at presentation.

It is interesting that 3 of our patients (20%) were asymptomatic at presentation. This rate has varied between 25 and 31% in other studies.[5,16] In asymptomatic patients, the diagnosis was made incidentally during an otolarygologic examination for other reasons.

Treatment. Treatment options include surgery and conservative management, the choice depending on the severity of signs and symptoms and the degree of the lesion's extension.

In cases in which the lesion has extended beyond the temporal bone, surgical treatment is almost inevitable. Surgery is also required if the disease leads to complications, which can include facial nerve paralysis, erosion of the middle ear bones, and a labyrinth fistula. Still, some authors recommend surgery as the first-line option, even when the lesion is limited to the external ear canal.[2,12] However, most authors have found that conservative treatment is sufficient for EECCs that are limited to the temporal bone.[3,5-8]

Conservative therapy entails debridement of all necrotic tissue and aspiration of all keratin debris under microscopy, with or without the use of a topical agent, and cleansing the ear canal.

Various topical agents have been described in the literature. Chang et al managed 12 patients with spontaneous EECC and treated them topically with diluted vinegar and local cleansing under microscopy; they reported no recurrences during a follow-up of approximately 31 months.[17] Darr and Linstrom found that conservative management was an appropriate option for patients with advanced EECC, as they successfully treated 8 of 9 patients with recurrent debridements (the other patient had acute facial paralysis and was treated surgically).[14] They repeated local cleansing under microscopy at intervals of 3 months in the 8 patients, and they observed no recurrence or progression during approximately 18 months of follow-up.

We treated all of our patients with local debridement under otomicroscopy and repeat aspirations, and we used a topical antibiotic/steroid agent to treat infection when necessary. We scheduled our patients for a repeat aspiration approximately every 10 weeks after the first treatment. During a mean follow-up of 41 months, no recurrence of signs or symptoms and no disease progression were observed in any of our patients.

In conclusion, our findings indicate that signs and symptoms and disease progression in limited EECC can be controlled safely with local debridement under microscopy and repeated regular aspirations without the need for surgery. However, longer follow-up studies are needed to assess the long-term effectiveness of this treatment protocol.

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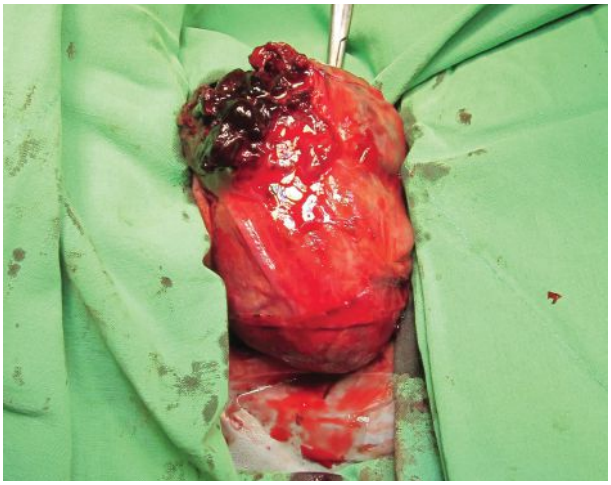

Figure 3. This perioperative photo shows the large goiter with focal capsular rupture; hemorrhage is seen on the right side.

aspiration rather than spontaneous bleeding.[2]

The cause of spontaneous thyroid hemorrhage remains unclear, with some authors speculating that the Valsalva maneuver might elicit venous pressure elevation and thereby lead to venous rupture and hemorrhage. Thus, straining, crying, and physical exertion may all be possible events preceding the hemorrhage.[3] In our patient, bleeding might have been induced by coughing and the underlying uncontrolled hypertension.

Without doubt, massive thyroid hemorrhage with airway obstruction warrants urgent management. The importance of definite airway establishment for ventilation and oxygenation cannot be overemphasized. Some authors advocate awake intubation with a small-caliber endotracheal tube since the patient's airway is already compromised by the thyroid mass. Intubation after the patient is anesthetized is associated with the risk of complete airway obstruction once the patient loses consciousness.[2] Subsequent thyroidectomy is required to relieve the airway obstruction. Tracheostomy is usually not necessary once the endotracheal tube is inserted, except in cases of tracheomalacia.[2]

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