INTRODUCTION

External auditory canal (EAC) cholesteatoma is a lesion lined with stratified squamous epithelium containing proliferative keratin with bony erosion. It is an uncommon benign disease with an unclear etiology and pathogenesis. Some authors have reported that EAC cholesteatoma is a result of trauma to EAC, chronic inflammation, ear canal stenosis, or that it arises spontaneously [1, 2]. Symptoms of EAC cholesteatoma are indolent and nonspecific. Patients with EAC cholesteatoma commonly complain of unilateral otorrhea and otalgia; however, unilateral hearing impairment has rarely been reported [3, 4].

The locally invasive expansion of EAC cholesteatoma can cause the destruction of the adjacent structures, and if the mastoid cavity is invaded, facial nerve paralysis and labyrinthine fistula can occur [5, 6]. Destruction through the anterior wall of EAC can affect the temporal–mandibular joint [7], and advanced invasion of the skull base can induce meningitis or an intracranial abscess [8]. The mechanism of bony destruction in cholesteatoma remains unclear; however, most hypotheses of bony destruction or resorption in cholesteatoma involve pressure necrosis, osteolysis, and/or contact between the inflammatory granulation tissue and bone, which then causes enzymatic bony destruction [9, 10]. The aim of this study was to describe the clinical manifestations of EAC cholesteatoma and to present our experience of surgical management in such patients.

MATERIALS and METHODS

A total of 28 patients (30 surgical ears) were enrolled from January 2004 to December 2013. All patients underwent a detailed medical examination, medical history review, and a battery of tests including pure tone audiometry and high-resolution computed tomography (CT) of the temporal bone. This study was approved by the institutional review board.
The severity and staging of EAC cholesteatoma was based on the criteria proposed by Naim et al. [1] in 2005 and was based on the clinical presentation and level of destruction of the temporal bone (Table 1). Invasion was limited to the epithelium of EAC at an early stage. However, destruction of the bony ear canal or adjacent structures was at a late stage.

Operative Procedures for EAC Cholesteatoma
Every patient underwent eradication surgery with external auditory canaloplasty under general anesthesia. Surgery was performed based on the staging of EAC cholesteatoma and was first assessed using high-resolution temporal bone CT (Figure 1). The procedures were performed in a number of steps. First, 2% lidocaine with 1:100000 epinephrine was injected over the postauricular area, and after a few minutes, a postauricular incision was made. Subcutaneous and areolar tissues were then carefully dissected to identify the temporalis muscle fascia, after which a piece of temporalis fascia was harvested and compressed for reconstruction. The posterior flap was raised and Henle's spine identified and then a horizontal incision was made to approach the external canal. Subsequently, the extensive area of EAC cholesteatoma was identified and eradication was performed. To preserve a normal ear canal contour, irregular margins of the bony wall were smoothly drilled out (Figure 2). The inferior pedicled soft-tissue periosteal flap was harvested for the reconstruction of the external auditory contour, and canaloplasty was performed with the obliterating flap in the cholesteatoma cavity. During the canaloplasty, the temporalis fascia was placed under the skin of EAC and the inferior pedicled soft-tissue periosteal flap was placed under the fascia to fill the residual cavity after drilling the bone erosion to create a smooth ear canal contour (Figure 3). The final step of bony reconstruction was to set the inferior pedicled soft-tissue periosteal flap, temporalis fascia, and the full-thickness skin graft layer by layer.

Statistical analyses were conducted using Excel 2013 (Microsoft Corp., Redmond, WA, USA).

RESULTS
This was a retrospective study, and all patients received eradication of a cholesteatoma sac and reconstructive canaloplasty. Of the 28 patients (30 surgical ears), 9 (9 ears) were males and 19 (21 ears) were females, with mean ages of 53.7 (range 18–74) and 53.7 (range 22–83) years, respectively. There were 11 right and 19 left ears, and two of the females had previously undergone bilateral ear surgery. The most common clinical manifestations were unilateral otalgia (19 ears, 63.3%) and otorrhea (14 ears, 46.7%), followed by ear fullness (10 ears, 33.3%), tinnitus (5 ears, 16.7%), and hearing loss (4 ears, 13.3%). Four of the surgical ears (13.3%) were asymptomatic and incidentally discovered during otoscopy. The most frequent locations of EAC cholesteatoma with bony invasion were the posterior–inferior (12 ears, 40%), inferior (9 ears, 30%), posterior (6 ears, 20%), and posterior–inferior–anterior (3 ears, 10%) aspects. EAC cholesteatoma was staged using the classification as proposed by Naim et al. [1]; 26 ears (86.7%) were stage III and 4 ears (13.3%) were stage IV (Table 2). All patients received surgical management via a postauricular inlet, and the average length of postoperative follow-up was 61.5 months (range 8–131 months). One ear suffered from recurrence after surgery for 1 year 3 months. Pre and postoperative pure tone audiometry were performed in selected cases; there was no specific threshold change noted in these patients.
Case Presentation

A 40-year-old female suffered from right-sided otalgia and otorrhea for 6 months. Medical management at a local clinic with topical and systemic antibiotics combined with regular local treatment was successful. However, she was referred to our hospital for a further opinion, and otoscopy revealed erosions in the posterior and inferior aspects of EAC that contained bony debris and sequestra. A CT scan confirmed a soft-tissue mass with bony destruction in the inferior portion of EAC (Figure 4a). She underwent surgery to remove the EAC cholesteatoma and canaloplasty through the postauricular approach. The shape of the EAC wall was found to have healed well after 6 months of follow-up (Figure 4b).

DISCUSSION

Cholesteatoma is commonly found in the middle ear and mastoid cavity, with a reported annual incidence rate of 9.2 cases per 100000 individuals. However, EAC cholesteatoma is rare, with a reported incidence rate of approximately 1.2–3.7 per 1000 otological patients. The annual incidence of EAC cholesteatoma has been reported to be approximately 0.15 cases per 100000 persons.

The etiology and pathogenesis of EAC cholesteatoma are unclear. In general, the narrowing or occlusion of EAC is believed to be responsible. Holt proposed multiple etiologies, such as previous ear surgery/traua, congenital ear canal stenosis/obstruction, or spontaneous formation. The formation of EAC cholesteatoma is considered to be via “keratinization in situ,” which is a reduction in migratory capacity of epithelium. Same as the vascular distribution, the direction of epithelial migration is outward from the manubrium of malleus to annulus. Makino and Amatsu reported that the mi-

Table 2. Summary of reported cases with external auditory canal cholesteatoma

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Side</th>
<th>Location</th>
<th>Otalgia</th>
<th>Otorrhea</th>
<th>Hearing loss</th>
<th>Tinnitus</th>
<th>Ear fullness</th>
<th>Anatomical invasion</th>
<th>Naim’s stage</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>25</td>
<td>M</td>
<td>R</td>
<td>Post</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>+</td>
<td>Mastoid</td>
<td>IV</td>
</tr>
<tr>
<td>2</td>
<td>35</td>
<td>F</td>
<td>L</td>
<td>Post &amp; Inf</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Excavation</td>
<td>III</td>
</tr>
<tr>
<td>3</td>
<td>60</td>
<td>M</td>
<td>R</td>
<td>Post</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Mastoid</td>
<td>IV</td>
</tr>
<tr>
<td>4</td>
<td>53</td>
<td>M</td>
<td>L</td>
<td>Post &amp; Inf &amp; Ant</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>+</td>
<td>Excavation</td>
<td>III</td>
</tr>
<tr>
<td>5</td>
<td>67</td>
<td>M</td>
<td>L</td>
<td>Post &amp; Inf &amp; Ant</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>+</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>6</td>
<td>75</td>
<td>F</td>
<td>L</td>
<td>Inf</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Excavation</td>
<td>III</td>
</tr>
<tr>
<td>7</td>
<td>62</td>
<td>M</td>
<td>R</td>
<td>Post</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Mastoid</td>
<td>IV</td>
</tr>
<tr>
<td>8</td>
<td>59</td>
<td>M</td>
<td>L</td>
<td>Post</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>+</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>9</td>
<td>69</td>
<td>F</td>
<td>L</td>
<td>Inf</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>10</td>
<td>81</td>
<td>F</td>
<td>L</td>
<td>Post &amp; Inf</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>11</td>
<td>22</td>
<td>F</td>
<td>L</td>
<td>Post &amp; Inf</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Mastoid</td>
<td>IV</td>
</tr>
<tr>
<td>12</td>
<td>40</td>
<td>F</td>
<td>R</td>
<td>Post &amp; Inf</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>13</td>
<td>37</td>
<td>F</td>
<td>L</td>
<td>Post &amp; Inf</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>14</td>
<td>78</td>
<td>F</td>
<td>L</td>
<td>Inf</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>15</td>
<td>18</td>
<td>M</td>
<td>R</td>
<td>Inf</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>16</td>
<td>31</td>
<td>F</td>
<td>L</td>
<td>Post &amp; Inf</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>17</td>
<td>74</td>
<td>M</td>
<td>R</td>
<td>Inf</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
<tr>
<td>18</td>
<td>38</td>
<td>F</td>
<td>BR</td>
<td>Post &amp; Inf</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>EAC erosion</td>
<td>III</td>
</tr>
</tbody>
</table>

EAC: external auditory canal; M: male; F: female; R: right; L: left; B: bilateral; Post: posterior; Inf: inferior; Ant: anterior
through the anterior aspect of EAC [6, 15, 16]. In addition, rare cases of sinus [3, 5, 6]. The temporal–mandibular joint would then be affected it may damage the fallopian canal, semicircular canals, and sigmoid advanced diseases. If EAC cholesteatoma invades the mastoid cavity, motion of EAC and involvement of adjacent structures can occur in ad-}

Other medical problems affecting blood circulation can also cause EAC cholesteatoma, such as diabetes mellitus, heavy smoking, and repeated microtrauma [4, 13]. Some authors have also proposed that syphilis, scarlet fever, and rheumatoid arthritis can also cause EAC cholesteatoma [1]. More female patients had EAC cholesteatoma in our study (19/28), which is in agreement with other studies [1, 14]. However, these studies have only included a small number of cases of EAC cholesteatoma, and future studies should include more cases with a long follow-up period.

The manifestations of EAC cholesteatoma include accumulation of keratin debris and bony destruction as inflammation progresses. The most common symptoms are unilateral otalgia and hearing loss, followed by otorrhea and tinnitus [15]. However, one metaanalysis of EAC cholesteatoma reported that unilateral otorrhea with otalgia was the most common symptom, followed by hearing impairment [6]. In our study, the most common manifestations were unilateral otalgia (19 ears, 63.3%) and ototorrhea (14 ears, 46.7%), with relatively fewer patients complaining of ear fullness (10 ears, 33.3%), tinnitus (5 ears, 16.7%), and hearing loss (4 ears, 13.3%). Four ears (13.3%) were asymptomatic and incidentally discovered during otoscopic examinations.

External auditory canal cholesteatoma can be limited to the epithelium without bony invasion, in which case keratin debris and epithelial hyperplasia are found during otoscopy. However, the bony destruction of EAC and involvement of adjacent structures can occur in advanced diseases. If EAC cholesteatoma invades the mastoid cavity, it may damage the fallopian canal, semicircular canals, and sigmoid sinus [5, 9, 13]. The temporal–mandibular joint would then be affected through the anterior aspect of EAC [6, 15, 16]. In addition, rare cases of skull base invasion have been reported in advanced diseases [17].

Clinical examinations alone are not sufficient to accurately detect the spread of EAC cholesteatoma to the mastoid cavity and adjacent structures. Currently, high-resolution CT of the temporal bones is the gold standard to assess lesions and make decisions for the treatment of EAC cholesteatoma [15, 17]. In temporal bone CT, EAC cholesteatoma is shown as a soft-tissue tumor within the auditory canal, bony invasion, or destructions of the middle ear and adjacent structures [5, 18]. All our cases received high-resolution CT of the temporal bone to detect lesions and guide management.

Temporal bone CT also plays an important role in the diagnosis and extension of EAC cholesteatoma. However, soft-tissue abnormalities in EAC can include atresia, edema, hemorrhage, keratosis obturans, cholesteatoma, adenoma, ceruminoma, fibroma, mixed tumors, as well as squamous cell, basal cell, and adenocystic carcinoma [18]. Naim et al. [1] recommended performing routine histological investigations of operative specimens to rule out malignancy in EAC cholesteatoma. They reported two patients with EAC tumors, who had persistent otorrhea and similar clinical presentations to EAC cholesteatoma. In these cases, squamous cell carcinoma was diagnosed with histological analysis [1]. In our study, each surgical specimen was pathologically examined and then diagnosed with EAC cholesteatoma.

External auditory canal cholesteatoma was staged using the classification as proposed by Naim et al. [1], which is based on pathological, clinical, and radiological evidence (Table 1). In the earlier stages (stages I and II), there is no destruction of the bony canal, whereas bony destruction of EAC and involvement of adjacent structures are seen in advanced stages (stages III and IV). In our study, all surgical cases were at advanced stages [26 ears (86.7%) stage III, 4 ears (13.3%) stage IV].

With regards to the management of EAC cholesteatoma, the main goal is the same as that for middle ear cholesteatoma. It is very important to eradicate the cholesteatoma and preserve normal structures as far as possible to restore epithelial migration in EAC. In earlier stages, EAC cholesteatoma without bony destruction should be conservatively treated. The first choice of therapy is to remove keratin debris and locally apply gauze with salicylate and cortisone ointment [1, 11]. If these procedures are not sufficient to control the clinical symptoms, or the patients have the advanced stage with the destruction of the bony canal or adjacent anatomical structures, surgery should be considered. The goal of surgery is to excise defective areas with a clear margin of epithelium and carefully remove eroded skin and destructed bone. The irregular bony surface should then be saucerized using a diamond burr to achieve a smooth contour. Some studies have proposed surgical techniques to fill the bony defects, such as a soft-tissue flap, ear conchal cartilage, and temporalis [11, 18, 20].

In large defects of EAC and mastoid cavity, Tos [21] recommended obliteration with an inferior-based subcutis muscle peristomeum flap, harvested along the posterior edge of the auricular incision. Some authors have suggested using bone with perichondrium for reconstruction [22]. Anthony and Anthony [11] reported the use of two surgical techniques to manage EAC cholesteatoma. First, careful removal of the EAC cholesteatoma with one to two margins of normal epithelium was performed, followed by smoothing and saucerizing the bony contour. Second, set harvested normal skin over the exposed bony defect, and then fill EAC with Gelform®.

Based on the satisfactory removal of the EAC cholesteatoma and reconstruction of defects, we modified the procedures suggested by Anthony in our patients. After eradicating the EAC cholesteatoma,
In conclusion, EAC cholesteatoma is an uncommon benign disease with multiple etiologies and pathogenesis. The clinical manifestations are not always obvious, and most patients suffer from advanced disease requiring further consultation. Such patients require surgical management. In our clinical experience of treating patients with EAC cholesteatoma, canaloplasty and obliteration with an inferior pedicled soft-tissue periosteal flap was a reliable procedure.

**Ethics Committee Approval:** Ethics committee approval was received for this study from the ethics committee of KMUHIRB-E(II)-20160161.

**Informed Consent:** Written informed consent was obtained from patients who participated in this study.

**Peer-review:** Externally peer-reviewed.


**Acknowledgements:** The authors thank the help from the colleagues of Department of Otolaryngology, Kaohsiung Medical University Hospital.

**Conflict of Interest:** No conflict of interest was declared by the authors.

**Financial Disclosure:** The authors declared that this study has received no financial support.

**REFERENCES**

7. Martin DW, Selesnick SH, Parisier SC. External auditory canal cholesteatoma with erosion into the mastoid. Otolaryngol Head Neck Surg 1999; 121: 298-300. [CrossRef]
11. Anthony PF, Anthony WP. Surgical treatment of external auditory canal cholesteatoma. Laryngoscope 1982; 92: 70-75. [CrossRef]