Cholesteatoma of the external auditory canal is rare in otologic practice. Current literature on that entity is generally limited and has been reported primarily in the White population; a significant underrepresentation has been noted in Asians. In this paper, we describe our Asian experience involving 4 cases of external auditory canal cholesteatoma that were diagnosed and subsequently managed in a tertiary-care hospital. We present our experience of the clinical presentation, diagnosis, and management of cholesteatoma of the external auditory canal to create an awareness of this rarity, especially in an Asian context.
External auditory canal (EAC) cholesteatoma is rare in otologic practice. Failure to diagnose and treat this condition appropriately can lead to serious complications, including erosion of the temporal bone, inner ear, and intracranial structures. In the following case studies, EAC cholesteatoma in 4 Asian patients is discussed and respective treatments are presented.

**CASE REPORTS**

**Case 1**

A 33-year-old Malay woman with a history of external ear polyps that were surgically removed during childhood complained that her left ear had been “blocked” for 3 to 4 months. Examination revealed granulation tissue in the EAC. Results of an audiogram showed a left conductive hearing loss with an air-bone gap of 20 dB at low frequencies and 50 dB at high frequencies. A computed tomography (CT) scan revealed a left soft tissue lesion that primarily involved the left EAC. The lesion had eroded the roof of the canal as well as the anterior and posterior walls and had extended into the middle ear with erosion of the head of malleus and the long process of incus (figures 1 and 2). A left tympanomastoidectomy was performed, and intraoperative findings revealed an extensive cholesteatoma eroding anteriorly into the temporomandibular joint, posteriorly into the mastoid air cells, inferiorly to the digastric ridge, and superiorly into the epitympanum. The head of the malleus and the body of incus were exposed. Medially, the cholesteatoma was pushing against the tympanic membrane (TM), eroding the bony annulus, exposing the fibrous annulus, and draping over the chorda tympani and left facial nerve, although facial nerve palsy was not detected.

**Case 2**

A 46-year-old Chinese woman presented with a chronic right ear discharge that she had experienced for 2 years. Examination revealed a right granuloma in the EAC. High-resolution CT showed bony erosion and scalloping of the posterior wall and the roof of the right EAC (Figure 3). The middle ear and ossicles were normal, and the mastoid was well-pneumatized with no opacification. The patient underwent canal-wall-down mastoidectomy, and the intraoperative
findings and the histologic report were consistent with a diagnosis of EAC cholesteatoma.

**Case 3**

A 41-year-old Chinese man presented with an intermittent right ear discharge and complained that his ear had been blocked for several years. He denied previous ear surgery or trauma. Results of an audiogram revealed a conductive hearing loss with an air-bone gap of 20 dB at low frequencies and 50 dB at high frequencies. A CT scan showed erosion of the bony sulcus canal (Figure 4). Intraoperative findings indicated a cholesteatoma that was eroding into the posterior ear canal. The TM was intact. An atticoantrotomy was performed, and the canal wall was drilled down with complete resection of the cholesteatoma. A meatoplasty was then performed.

**Case 4**

A 21-year-old Chinese woman presented with left-sided otorrhea that had persisted for 6 months and was associated with otalgia and decreased hearing. The patient denied prior otologic surgery or trauma. Granulation tissue in her left external ear canal was identified, and a left conductive hearing loss with an air-bone gap of 50 dB across all frequencies was noted. A CT scan (Figure 5) revealed soft tissue opacification that was confined to the left EAC and was associated with cortical bone erosion into the anterior mastoid air cells, the canal floor and roof, and the posterior canal wall. There was no extension into the middle ear cavity, and the ossicular chain, the inner ear, and the left facial nerve canal were intact. A canal-wall-down tympanomastoidectomy with meatoplasty was performed. Histologic analysis revealed inflamed granulation tissue with lymphocytes and plasma cells, multinucleated giant cells, keratin flakes, and an overlying squamous epithelium with keratin, all of which are consistent with a cholesteatoma.

**DISCUSSION**

EAC cholesteatoma is a rare occurrence that is estimated to occur in about 1 of 1000 new referrals presented for otological problems. Since 1980, this disease process has been described as a clinical entity distinct from keratosis obturans. The features that differentiate EAC cholesteatoma from similar entities are based on 3 main criteria. First, EAC cholesteatoma occurs in older patients, is usually unilateral, and follows an indolent course characterized by dull otalgia and otorrhea. Hearing is within the normal range in those patients. Keratosis obturans, however, is primarily a disease of young adults with acute otalgia and conductive bilateral hearing loss. Second, clinical examination in patients with an EAC cholesteatoma...
reveals an epidermal diverticulum arising from floor of an otherwise normal ear. In those with keratosis obturans, the ear canal is filled with keratin plug and appears widened, and hyperemia of the canal skin can be identified. Third, patients with keratosis obturans exhibit marked inflammation in the subepithelial tissue without bony erosion, and the EAC is filled with keratin squames arranged in a lamellar pattern. In patients with EAC cholesteatoma, a bony erosion of the canal wall with a blind diverticulum sac containing bony sequestra and keratin squames arranged in a random pattern can be identified. In the patients in our study, the diagnosis of EAC cholesteatoma was based on clinical presentation, the presence of focal bony erosion on CT scan, and (subsequently) on the results of histologic analysis.

The cause of EAC cholesteatoma is unknown, although possible pathophysiologic mechanisms include the retention of squamous debris in the medial portion of the EAC as a result of occlusion or the narrowing of the external meatus; the implantation of squamous epithelium cyst formation deep in the skin of the external canal; or erosion of the bone of the external canal, which permits the ingrowth of the squamous epithelium into the defect. Makino and Amatsu noted that the migratory rate of the epithelium in the inferior wall of the external ear canal was slower in patients with an EAC cholesteatoma than in those with a normal ear. This theory attributes the cause of EAC cholesteatoma to the overproduction of epithelial cells or to the faulty migration of the epithelium. Holt classified the aetiologies as follows: congenital, posttraumatic, iatrogenic, spontaneous, postobstructive, or postinflammatory.

Our experience with the clinical presentation of cholesteatoma in Asian patients seems to mirror that in the White population with respect to differences in the age of the patient at presentation and the presence of hearing loss. All of our patients presented with a long history of otologic complaints, which is typical of individuals with an EAC cholesteatoma. Spontaneous EAC cholesteatoma reportedly occurs most often in the elderly (most common patient age, 40-75 years). Another case series reported that the mean age of patients who presented with an EAC cholesteatoma was 61 years (age range, 34-83 years). The 2 cases of spontaneous cholesteatoma described in this article are highly unusual because they occurred in a 21-year-old woman and a 41-year-old man. The clinical and anatomicopathologic features in our patients were consistent with those of people who have an EAC cholesteatoma rather than keratosis obturans, and both patients with spontaneous cholesteatoma denied having prior ear disease.

Although hearing loss in patients with an EAC cholesteatoma is uncommon because of the noninvolvement of the TM and the ossicular chain, conductive hearing loss can occur because of the obliteration of the EAC or the erosion of ossicles and the TM in advanced stages of disease. In our second, third, and fourth patients, we noted a conductive hearing loss caused by the lesion’s having filled the EAC without involvement of the TM or ossicles. Vrabec and Chaljub reported that in 12 patients with spontaneous EAC, no hearing loss was attributable to the lesion, because a cholesteatoma tends to cause bony erosion and to render the TM and ossicles relatively uninvolved until late in the course of the disease. In the first patient described in this report, the cholesteatoma had eroded through the TM and the ossicles to produce a conductive hearing loss.

Radiologic investigation is important in helping the clinician confirm the diagnosis, assess the extent of disease, and plan the surgical treatment needed. The typical CT appearance of an EAC cholesteatoma is that of a localized bony erosion of the auditory canal. A cholesteatoma can invade and involve the surrounding structure, as illustrated by the first and fourth patients described in this report. Usually, an EAC cholesteatoma arises in the inferior part of the auditory canal, which was involved in only 1 of the 4 patients described in this article. The posterior wall and roof of the auditory canal was involved in all 4 patients, and anterior wall involvement was noted in 2 of the patients studied.

Bone erosion can progress to involve the temporomandibular joint anteriorly, the
hypotympanum and the jugular dome inferiorly, and the mastoid posteriorly and upward to the facial nerve (11). Although the TM is not usually affected, advanced stages of disease may be characterized by some involvement. In the first case presented in this article, the cholesteatoma had invaded the middle ear with erosion of the malleus and incus. In the third case, part of the TM had to be sacrificed for cure because the cholesteatoma was in contact with the TM. A temporalis fascia graft was used to cover the defect.

The reported complications of EAC cholesteatoma include facial dehiscence or palsy, invasion of the cholesteatoma into the mastoid cavity, ossicular erosion, and/or the development of a labyrinthine fistula that causes vertigo (6, 12-14). Conductive hearing loss can also occur after obliteration of the EAC or invasion into the mastoid cavity with mucoid accumulation in the middle ear and a normal TM and ossicles (15).

**TREATMENT**

The treatment of an EAC cholesteatoma, which can vary from conservative management to modified radical mastoidectomy, depends on the extent of disease. When the cholesteatoma is localized and the entire extent of the bony defect can be visualized, debridement and applications of mineral oil to the ear canal have been used as therapy, and the topical application of 5-fluorouracil has also been used successfully (16). If the defect is deeper, meatoplasty is required to remove diseased skin and bone and to exteriorize the recess. The necrotic bone must be drilled away, and the resultant raw area can receive a graft of the temporalis fascia or a split-skin graft (7). If the mastoid cavity is involved, a mastoidectomy is needed to eliminate the disease while preserving the TM and the ossicles. Vrabec and Chaljub (5) reported that most 12 of their patients with a spontaneous EAC cholesteatoma were treated with simple debridement. However both of our patients with a spontaneous EAC cholesteatoma had to undergo tympanomastoidectomy because of mastoid cavity involvement. In all our patients, the canal wall had to be drilled down to ensure disease clearance, and a tympanomastoidectomy with meatoplasty was needed for disease clearance in 3 patients. An atticoantrotomy surgery was performed for the patient whose disease did not affect the mastoid cavity.

The long-term recurrence rate of EAC cholesteatoma is not well-defined because it is a rare disease, but most researchers advocate long-term follow up. At the time of this writing, none of the patients described in this article has experienced a recurrence of disease.

**CONCLUSION**

Although EAC cholesteatoma is often reported to affect the elderly White population, we present 4 cases of younger Asian patients with this rare disease. It is important to recognize and treat this type of cholesteatoma appropriately because of its propensity to invade vital auditory structures.

**REFERENCES**