Disorders of the External Auditory Canal

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Abstract
The normal anatomy and physiology of the external auditory canal is covered, followed by detailed descriptions of disorders and treatments of known and unknown etiology. Included are asteatosis, bacterial and fungal external otitis, bullous myringitis, allergic dermatitis, keratosis obturans, canal cholesteatoma, exostosis, osteoma, ceruminoma, basal and squamous cell carcinoma, and adenoma. A review of malignant external otitis is included with special emphasis on the expanding role of outpatient treatment and the use of oral antimicrobials.

Key Words: Adenoma, cholesteatoma, dermatitis, external ear, otitis, squamous cell carcinoma

The external auditory canal of the adult is a complex structure that offers protection from trauma and bacteria to the delicate tympanic membrane and middle ear while (Table 1), at the same time, efficiently providing sound transmission. The canal can be conveniently divided into a medial, inner two-thirds, and a lateral outer third; the external ear canal terminates at the tympanic membrane. The outer third is cartilaginous and is covered with skin that is relatively thick; the medial, inner two-thirds is bony and covered with a very thin layer of skin that is exquisitely sensitive to pain. Adnexal structures, including hair follicles, sebaceous glands, and modified apocrine sweat glands, are located in the thick skin of the lateral cartilaginous third of the external auditory canal. Access to the tympanic membrane is restricted by the S-shaped configuration of the external auditory canal and by its narrowed isthmus, which occurs at the osteocartilaginous junction where the lateral third and middle two-thirds of the external auditory canal meet. Hair projecting into the lateral third of the external auditory canal traps foreign bodies and prevents their inward migration (Anson and Bast, 1980; Schuknecht and Gulya, 1986) (Fig. 1).

The external auditory canal has both afferent and efferent innervation from several different sources. The posterior and inferior regions

| Table 1 Protective and Antibacterial Characteristics of the External Auditory Canal |
|---------------------------------|--------------------------------------------------------------------------------|
| Protection from Trauma          | Length and configuration of external auditory canal protect tympanic membrane |
| Orientation of hair follicles retards inward migration of foreign bodies | Skin of medial external auditory canal is highly sensitive |
| Antibacterial Characteristics   | Low pH                                                                         |
| Cerumen                         | Lysozyme                                                                      |
| Fatty acids                     |                                                                               |

Figure 1 A normal external auditory canal. Small hairs can be seen in the lateral portion of the canal. These provide a protective function by limiting the ingress of foreign bodies, insects, etc. Reproduced with permission: Hawke M, Jahn AF. (1987). *Diseases of the Ear: Clinical and Pathological Aspects*. Philadelphia: Lea and Febiger.

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of the canal and the adjacent tympanic membrane receive afferent information from the auricular branches of the facial, glossopharyngeal, and vagus nerves. The anterior and superior aspects of the canal and adjacent tympanic membrane are afferently innervated by the auriculotemporal branch of the trigeminal nerve (Schuknacht and Gulya, 1986).

The pinna and external canal help to collect sound over large areas and concentrate it at the tympanic membrane. The external auditory canal contains a column of air approximately 2 cc in volume and can serve as a resonating chamber. Its resonance peak is between 2300 and 2800 Hz, which corresponds to a frequency range important for speech discrimination. Although there is no true amplification within this system, the sound intensification provided by this arrangement aids in overcoming the intrinsic impedance mismatch that occurs at the interface of the middle and inner ears.

The cerumen present in the external auditory canal is a mixture of secretions from sebaceous glands and modified apocrine sweat glands (Figs. 2–4). Secretions from the modified apocrine sweat glands are less viscous than the sebum secreted from sebaceous glands. Apocrine sweat glands are under the control of the sympathetic arm of the autonomic nervous system. Apocrine glands appear to respond to emotional states, certain drugs, medications, and mechanical stimulation.

Both the viscosity and volume of cerumen can change based on the degree to which apocrine glands are stimulated. The antimicrobial properties of cerumen, discussed elsewhere in this issue, are important in preventing infection.

Antimicrobial properties are due principally to the presence of saturated fatty acids, lysozyme and, especially, to a relatively low pH. Typical cerumen from normal individuals has a pH around 6.1 (Brister et al., 1986; Raman and Lumpur, 1986; Hawke and Jahn, 1987; Mahoney, 1987; Myers and Pueschel, 1987; Pillsbury and Wilson, 1992).

Disorders of the ear canal can be categorized into those of known and unknown etiology. The major disorders of the external ear are reviewed below with treatments for each.
DISORDERS OF KNOWN ETIOLOGY

Atresia and Malformation of the External Auditory Canal

The pinna, external auditory canal, middle ear, and inner ear can experience disruption of their embryologic development either separately or together. Unilateral atresia is three to six times more common than bilateral atresia and the right ear is more frequently affected than is the left. When the external auditory canal is malformed in the presence of a normal pinna, the middle ear is generally normal because canalization of the external auditory canal occurs late in embryologic development. Deformation of the pinna, on the other hand, is usually associated with atresia of the external auditory canal. Even in the face of microtia and atresia of the external auditory canal, formation of the middle ear and mastoid is often relatively normal. Malformation of the ossicles is a relatively common accompaniment of microtia with aural atresia. The most common ossicular malformation is fusion of the malleus and incus into a single, primitive ossicle. The stapes is usually normal. Malformation of the inner ear is uncommon even when atresia of the pinna and external auditory canal is severe. The facial nerve, however, frequently takes an anomalous course, a point that must be borne in mind if surgical intervention is contemplated. Congenital atresia may occur as an isolated event or be associated with a variety of named syndromes (Table 2) (Mattox et al, 1991).

Treatment

Two separate issues need to be addressed when caring for individuals with microtia and aural atresia. First and foremost is assessment of hearing loss. When atresia is unilateral and hearing in the contralateral ear is normal, preferential seating may be all that is necessary. When the malformation is bilateral and associated with maximal conductive hearing losses, then a bone-conduction hearing instrument (either conventional or implanted) should be used. Surgical repair of external ear deformity is usually delayed until the age of 6. Surgical repair at a younger age is more difficult and less certain of success. Children are so frequently humiliated in a school setting that repair, however, is frequently initiated prior to schooling.

Decisions about surgery for hearing improvement in cases of unilateral atresia are usually deferred until the child is old enough to decide for himself (about age 18). The success of surgical correction of hearing loss associated with atresia is variable but postoperative hearing results of 15 to 20 dB can be expected in 30 to 80 percent of patients.

External Otitis

External otitis is an infection involving the external auditory canal. It develops when normal defense mechanisms fail. High ambient humidity, direct exposure to water, local trauma, chronic dermatitis, the introduction of exogenous bacteria, and prolonged exposures to elevated temperatures contribute to the failure of local defense mechanisms (Meyerhoff and Caruso, 1991).

Diffuse external otitis develops when the mixed bacterial fauna that normally colonize the external auditory canal are replaced by a single organism of pathogenic potential. Saprophytes that normally reside in the external auditory canal include Staphylococcus, Streptococcus, Micrococcus, some gram-negative bacilli, and some types of saprophytic fungi. The presence of these saprophytes helps suppress pathogenic bacteria by competing for locally available resources.

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Diffuse external otitis is often referred to as "swimmer’s ear" because moisture plays a significant role in its pathogenesis. High ambient humidity provides a favorable growth environment for a different subset of organisms from those that normally colonize the external auditory canal. Cerumen can be mechanically "washed out" and its protective effect thereby removed. Water in the external auditory canal, perhaps most importantly, may alter the pH. The alkaline pH often produced by exogenous water is much less effective in suppressing pathogenic microorganisms (especially Pseudomonas aeruginosa) than is the normal acidic environment of the external auditory canal. Pseudomonas aeruginosa is notoriously inhibited by acidic environments. The deleterious effect of water or moisture...
is compounded if there is an underlying chronic seborrheic or allergic dermatitis that weakens the integrity of the integumental barrier (Senturia et al., 1980; Hawke and Jahn, 1987).

Diffuse external otitis typically starts with rapidly increasing aural pain over a period of several hours. Local tenderness around the tragus and pain associated with movement of the auricle develop rapidly. This pain is often sufficiently severe to require narcotic analgesics. As swelling continues, hearing may become subjectively diminished (Fig. 5). There is often an associated yellow or purulent exudate but it is generally scant. Physical examination shows edema of the external auditory canal. If the canal is sufficiently open for examination, a small amount of purulent exudate may be seen. There is almost always notable absence of cerumen. It is often not possible to examine the tympanic membrane but every attempt should be made to do so. The tympanic membrane, when visualized, is generally normal, except for a thin layer of exudate lying on its lateral surface. Occasionally, external otitis is secondary to an acute otitis media with rupture of the tympanic membrane and contamination of the external auditory canal with purulent drainage from the middle ear. When this is the case, physical examination of the tympanic membrane may show a perforation or other stigmata of otitis media.

**Treatment**

Effective treatment for acute external otitis can be deduced from its pathophysiology: restoration of the normal acidic environment of the external auditory canal will suppress pathogenic organisms and hasten the establishment of a balanced microbiological environment. This can often be accomplished with acidifying solutions alone. Acidified Burow's solution (Dombor®, a 1/1 mixture of isopropyl alcohol and 2 percent acetic acid, and 2 percent acetic acid in propylene glycol (Vosol®) are all effective acidifying solutions and generally sufficient to eliminate external otitis. If the external auditory canal is swollen closed or if mucopurulent exudate fills the lumen, then aggressive cleansing using a microscope and suction with or without placement of a wick will be necessary. Medications applied to a wick inserted into the external auditory canal draw medication into the inflamed canal. The patient should be re-examined in 24 to 48 hours in case additional aural toilet is needed.

Only rarely will mechanical debridement with reacidification fail to resolve an external otitis. But when these measures are insufficient, the use of antiseptics like gentian violet, Mercurochrome, or cresylate applied to the canal typically using a small cotton swab can suppress pathogenic organisms more aggressively. Local antiseptics frequently inhibit fungal as well as bacterial growth and consequently are less likely to promote the development of fungal otitis as a secondary infection.

Topical antibiotics applied as ototopical drops are also effective in the management of external otitis. However, they carry with them the potential for promoting resistant organisms and their broad spectrum bactericidal activity may promote fungal superinfection. Topical sensitivity is a common problem, especially with ototopical agents containing neomycin. Such sensitivity may be manifested only by continued pain and drainage and can easily be confused with ongoing infection. Persistent use of topical neomycin results in prolongation or exacerbation of pain, itching, weeping, and crusting of the external canal, conchal bowl, and lobule. More severe sensitivity reactions manifest themselves as the development of allergic rashes and swelling of the entire auricle. A patient who has been using antibacterial ototopical preparations for more than a couple of weeks and is thought to have “resistant” infection should be managed initially by removal of ototopical antibacterial...
drops, which are replaced with simple acidifying solutions. In the majority of cases, swelling resolves, drainage ceases, and pain disappears within a couple of days.

**Otomycosis**

Primary fungal external otitis is uncommon in the United States. Fungal external otitis occurs most commonly as a consequence of topical antibiotic therapy for acute bacterial external otitis. As mentioned above, such therapy can suppress all bacterial organisms within the external auditory canal, creating an empty "niche" into which fungal organisms can move. Aspergillus species are most typically encountered, although both Candida and Phycomycetes are occasionally identified.

Rare cases of primary fungal otitis usually begin with slight itching of the afflicted ear. As the infection progresses, edema, pain, and the production of mucopurulent exudate develop. Physical examination in the early stages may show only small amounts of erythema and edema but as infection intensifies, microscopic examination may reveal fungal filaments and spores. Ultimately, epithelial exfoliation and desquamation, combined with fungal debris and cerumen, produce sufficient debris to partially or completely occlude the external auditory canal. Primary invasive fungal external otitis should raise concern over the immune competence of the afflicted individual.

**Treatment**

Otomycosis is treated initially just as in bacterial external otitis: debridement of the external auditory canal and reacidification. If these methods fail, the use of antiseptic solutions usually are sufficient to eliminate the infection. Occasionally, topical antifungals in the form of drops may be necessary.

**Furunculosis**

Furunculosis is a localized folliculitis that begins within the hair follicles of the lateral third of the external auditory canal. Generally, it commences with a single, infected follicle but infection can spread to involve other follicles, resulting in diffuse involvement of a lateral third of the external auditory canal. Staphylococcus is the most common causative organism. Pain is very intense. Physical examination shows erythema, edema, and, often, abscess formation around the hair follicles of the lateral third of the external auditory canal.

**Treatment**

Although topical antibacterial drops and creams are often useful adjuncts, the mainstay of treatment is systemic antibiotic therapy directed against Staphylococcal organisms. Occasionally, frank abscesses will form that should be incised and drained (Meyerhoff and Caruso, 1991; Roland et al, 1997).

**Allergic Imitative External Otitis**

Allergic dermatitis of the external auditory canal usually begins with itching, excoriation, erythema, and mild edema (Table 3). Sometimes a scant amount of otorrhea can be appreciated. Pain is generally absent. When pain does occur, it may herald the development of secondary diffuse bacterial external otitis. Etiologic agents include earplugs, hearing aid molds, soaps, detergents, or regular digital manipulation. True allergic contact dermatitis is a Gell and Coombs type IV, T-cell mediated inflammatory response to a previously sensitized inciting antigen. Topical neomycin, nickel, and poison ivy are all relatively common culprits. Approximately 15 percent of the population is sensitive to neomycin. Allergic contact dermatitis initially produces a maculopapular rash that ultimately becomes vesicular. Rupture of vesicles produces a macerated, irritative, and edematous skin surface. Eventually, fissuring of the skin of the auricle and lobule may develop.

**Treatment**

Both irritant and allergic dermatitis can be treated with topical steroid medications including drops and creams. The lowest dose of the least powerful topical steroid should be used in order to minimize local skin atrophy. Severe reactions may require a high, short-term tapering dose of systemic steroids. Both irritant and

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**Table 3 Causes of Allergic Dermatitis**

<table>
<thead>
<tr>
<th>Cause</th>
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<tbody>
<tr>
<td>Neomycin</td>
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<tr>
<td>Nickel</td>
</tr>
<tr>
<td>Rubber</td>
</tr>
<tr>
<td>Polymers</td>
</tr>
<tr>
<td>Chromium compounds</td>
</tr>
<tr>
<td>Poison ivy, oak, or sumac</td>
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</tbody>
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allergic external otitis may be compounded with secondary bacterial external otitis, which can both obscure diagnosis and complicate treatment (Hawke and Jahn, 1987; Shea, 1996; Tran et al, 1996).

**Seborrheic Dermatitis**

Seborrheic dermatitis is a chronic condition affecting the skin of the external auditory canal. It is characterized by frequent periods of exacerbation alternating with periods of remission. The fungal organism Malassezia furfur has been implicated in the etiology of the disease. The process often begins in childhood. It is commonly associated with seborrheic dermatitis of other anatomic areas, including the scalp, groin, axilla, and anogenital regions. Simple pruritis is its most ubiquitous and common manifestation. Visible drying and flaking of skin in the area of the conchal bowl, introitus of the external auditory canal, and lateral third of the external auditory canal develop as the severity increases. Ultimately, skin fissuring may develop. Decreased skin integrity associated with seborrheic dermatitis makes individuals with chronic seborrheic dermatitis vulnerable to acute bacterial external otitis. When a patient is seen with frequent recurrence of acute bacterial external otitis, a chronic underlying dermatitis of the external auditory canal should be sought as a predisposing condition.

**Treatment**

Low-potency steroid creams are effective in controlling seborrheic dermatitis even when used infrequently or once or twice a week. Nizoral cream is effective against this and can also be used to effectively eliminate symptoms.

**Psychogenic Dermatitis**

Psychogenic dermatitis (neurodermatitis) is produced by the patient himself, usually in response to primary or secondary pruritis. Scratching, scraping, and manipulating the external auditory canal in an attempt to relieve the pruritic sensation leads to drying, lichenification, and a scaly eruption in the external auditory canal. This is known as the itch-scratch cycle. Although sometimes of purely psychogenic origin, local and systemic diseases can produce pruritis, which results in neurodermatitis. Diabetes mellitus, renal disease, and lymphoma all produce itching of the skin.

**Neoplasia**

Although neoplasms of the external auditory canal are relatively uncommon, their proximity to intracranial structures makes their early identification a prerequisite for successful treatment. Pain, bloody otorrhea, exposed bone within the external auditory canal, and facial paralysis mandate a thorough evaluation to rule out a neoplastic etiology.

Squamous cell carcinoma is the most common malignancy of the external auditory canal. It is frequently associated with secondary infection and mucopurulent drainage. The mass in the external auditory canal is often indistinguishable from the type of granulation tissue that is so frequently associated with a chronic, draining ear. Pain, chronic and severe, is frequently a distinguishing feature. Mucopurulent drainage for more than a couple of weeks, associated with significant pain, should raise suspicion for malignancy. Moreover, mucopurulent drainage secondary to malignancy is unresponsive to usual medical management. Granulation tissue present in the external auditory canal for more than a few weeks warrants biopsy. Biopsy should occur even earlier if there is associated facial nerve paralysis or regional lymphadenopathy. Exposure of bone, especially when sequestra of dead bone can be removed from the external auditory canal, should also significantly raise the index of suspicion. Squamous cell carcinoma usually spreads along the external auditory canal with lymphatic spread through the fissures of Santorini to the preauricular and/or deep parotid lymph nodes.

**Treatment**

Surgical removal of the involved external auditory canal is frequently successful in curing squamous cell carcinoma when the disease is limited to the canal itself. Removal of first-echelon draining lymph nodes in the parotid or preauricular area with upper neck dissection is often
an essential part of the extricative procedure. But once the disease has spread into the middle ear, the chances of cure are considerably diminished. Total temporal bone resection is required, which is a large and potentially dangerous operation. Total temporal bone resection is invariably associated with permanent facial nerve paralysis. High-dose external beam radiation therapy is often used adjunctively and can significantly slow tumor growth. However, it is unlikely to produce a cure, even when combined with adjunctive chemotherapy, if disease is advanced (Kuhel et al, 1996).

**Basal Cell Carcinoma**

Primary basal cell carcinoma is more common in the auricle and the conchal area and less common within the external auditory canal. Primary basal cell carcinomas around the auricle, especially in the conchal bowl, are much more insidious and potentially dangerous than basal cell cancers in other areas of the body. Although basal cell carcinoma lacks potential for metastasis, it is locally invasive and can be very aggressive. Direct extension onto the dura mater and into the central nervous system (CNS) occurs in advanced cases.

**Treatment**

Again, early diagnosis is key to successful treatment. Any nonhealing lesion on the auricle or concha should be suspected and biopsied. Because of the occult nature of subcutaneous extension beyond surgical margins, basal cell carcinomas are generally best treated with subtotal resection of the temporal bone. Margins should be closely controlled with multiple, intraoperative frozen sections when local resection is performed. Regional lymphadenectomy is not necessary for control of basal cell carcinoma as lymphatic spread is very uncommon (Kuhel et al, 1996).

**Other Carcinomas**

The apocrine glands of the outer third of the external auditory canal can produce a variety of rare tumors, including adenocarcinoma, adenoid cystic carcinoma, and mucoepidermoid carcinoma.

**Treatment**

These lesions should be treated in the same way as squamous or basal cell carcinoma. The primary lesion is removed along with a margin of normal tissue and first-echelon draining regional lymphatics. In many cases, adjunctive radiation or chemotherapy is helpful.

**Osteoma**

Osteoma of the external auditory canal is a true neoplasm and it should be clearly distinguished from exostosis. Osteomas occur most commonly in young adults and they are classically unilateral. They are smooth, pedunculated, and covered with normal skin (Fig. 6). Osteomas are asymptomatic until they grow large enough to obstruct the external auditory canal. Obstruction can produce conductive hearing loss. More commonly, however, the mass prevents normal epithelial migration of desquamated squamous epithelium, which accumulates medial to the osteoma. This results in local infection and chronic drainage with or without pain. Such trapped epithelial debris can develop into a frank cholesteatoma of the external auditory canal medial to the obstruction. Pathological examinations of excised osteomas show normal trabecular bone (DiBartolomeo et al, 1991; Tran et al, 1996).

**Figure 6** Large sessile osteoma of the external auditory canal. This is a true neoplasm. This osteoma has become sufficiently large so as to occlude the external auditory canal. Cerumen and epithelial debris produced more medially in the external auditory canal can become trapped, causing cholesteatoma of the external auditory canal with/without repeated infection. Exogenous water from bathing and swimming can become trapped medial to the osteoma. Reproduced with permission: Hawke M, Jahn AE. (1987). Diseases of the Ear: Clinical and Pathological Aspects. Philadelphia: Lea and Febiger.
Treatment

Treatment is required only when osteomas become large enough to produce hearing loss or obstruction of the medial canal. When treatment is necessary, surgical excision is definitive.

Exostosis

Formation of exostoses is a reactive rather than neoplastic process. Exostoses are commonly associated with exposure of the external auditory canal to cold water. In some parts of the country, it is referred to simply as “surfer’s ear.” The proposed pathogenesis assumes that vasoconstriction after cold exposure leads to subsequent reactive hyperemia with stimulation of local osteocytes and the deposition of new bone. As might be expected, exostoses tend to be bilateral and multiple. They are round, smooth, and, most commonly, sessile (Fig. 7). They occur only in the bony inner two-thirds of the external auditory canal. They are generally covered with a thin but normal layer of epithelium through which small vessels can be seen. Histological evaluation shows that they consist of highly stratified layers of lamellar bone. It appears that periosteal irritation leads to uniform, progressive layers of new bone deposition (DiBartolomeo et al., 1991; Tran et al., 1996).

Treatment

Just as is true of osteomas, exostoses require treatment only when they are so large as to produce conductive hearing loss or trap water or skin in the medial end of the canal.

DISORDERS OF UNKNOWN ETIOLOGY

Keratosis Obturans

Keratosis obturans is a disorder of unknown etiology in which squamous epithelium is circumferentially desquamated into medial portions of the external auditory canal. The normal cleansing process of the external auditory canal is unable to keep pace with the rate of desquamation and soon a large amount of squamous epithelium occludes the medial portions of the external auditory canal. As desquamation continues, bony remodeling occurs, which creates enlargement and flaring of the medial external auditory canal.
It is unclear whether the disease is caused by excessively rapid production and desquamation of epithelial cells, failure of the normal migratory cleansing process, or a combination of both. The disorder is most commonly bilateral and is associated with chronic sinusitis and bronchiectasis, which are disorders associated with abnormal or immotile cilia. Clinically, it is more common in young or middle-aged individuals and is frequently bilateral. Conductive hearing loss is a common symptom and is due entirely to occlusion of the canal with keratin debris. Otorrhea and pain can occur if the mass of impacted keratin debris becomes infected. The dilatation of the medial canal is circumferential and diffuse, eventually leading to the formation of a bottleneck at the lateral aspect of the bony canal (Senturia et al, 1980; Hawke and Jahn, 1987; DiBartolomeo et al, 1991; Hartley et al, 1995; Tran et al, 1996).

Treatment

Management consists of office removal of desquamated squamous epithelium and keratin debris. If the process has been a long-standing one, especially if there has been medial flaring of the external auditory canal, removal of impacted squamous debris can be difficult and painful. It is not uncommon for several office sessions to be required. Ototopical corticosteroids may reduce the rate of epithelial cell turnover and consequently decrease the necessity for mechanical removal. Repeated debridement with or without the use of corticosteroids seems to produce remission in some cases.

Canal Cholesteatoma

Cholesteatoma formation within the external auditory canal also results in accumulation of squamous epithelial debris in the medial canal. It is, however, quite a different entity from keratosis obturans. While deposition of keratin debris in keratosis obturans is circumferential and diffuse, formation of desquamated squamous epithelium in cholesteatoma of the external auditory canal is focal. The pathogenesis of this disorder appears to be secondary to trapping of epithelium beneath the skin in the external auditory canal, possibly as a consequence of previous trauma.

It is usually unilateral and occurs most commonly in elderly patients. Physical examination shows accumulation of keratin and epithelial debris around a focus of denuded bone, usually in the posterior/inferior portions of the canal. When a canal cholesteatoma is longstanding, it is frequently possible to remove the small bony sequestra from the area of involvement. Canal cholesteatomas become infected and are associated with granulation tissue, inflammation of surrounding tissues, and tissue edema. Focal bone destruction should always raise the suspicion of neoplastic disease and biopsy is mandatory (Senturia et al, 1980; Hawke and Jahn, 1987; Hartley et al, 1995; Tran et al, 1996; Roland et al, 1997).

Chronic Hypertrophic External Otitis (Sclerosing External Otitis)

Chronic hypertrophic external otitis (sclerosing external otitis) is a disease of unknown etiology that is often bilateral. It begins with minimal irritation in the medial portion of the external auditory canal, intense pruritis, and a very thin, scant exudate. Over months or years, fibrous tissue is deposited within the dermis and subdermis, which slowly narrows and finally and completely obliterates the medial end of the external auditory canal. The endstage of this disorder is a 3- to 4-mm plug of fibrous scar tissue starting at the lateral surface of the tympanic membrane. Surprisingly, cholesteatoma formation due to trapping of squamous epithelium on the lateral tympanic membrane does not seem to occur. Other than the intense pruritis, the principal symptom is conductive hearing loss, which develops as the stenosis nears completion. Forty- to 60-dB losses are common (Senturia et al, 1980; Hawke and Jahn, 1987). Little can be done to arrest this process. There are some reports that topical or injected steroids may slow or arrest the progression of stenosis.
Table 4  Risk Factors for Malignant Otitis Externa

<table>
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<tr>
<td>Diabetes mellitus</td>
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<td>Advanced age</td>
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<td>Aural irrigation with water</td>
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<tr>
<td>Swimming</td>
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<td>Immunosuppression</td>
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Instrumentational manipulation of canal tissues appears to accelerate the process. Amplification is an effective form of remediation. Resection of the stenotic areas along with the involved skin of the medial external auditory canal and skin grafting has been used. Reported outcomes from surgical intervention are variable but generally not encouraging (Senturia et al., 1980; Hawke and Jahn, 1987).

Malignant Otitis Externa (Osteomyelitis of the Temporal Bone)

Pseudomonas osteomyelitis of the temporal bone can develop as a consequence of external otitis in diabetic patients. Although this disorder is occasionally seen in immunologically compromised individuals, the vast majority of cases are in elderly diabetics. When initially described by Ryan Chandler, the disease carried with it a mortality as high as 60 percent. Over the last decade, death from malignant otitis externa has become uncommon but treatment remains prolonged and, at times, difficult. Almost all cases in diabetics are caused by Pseudomonas aeruginosa. Case reports of malignant otitis externa due to gram-negative bacteria, Staphylococcus aureus, or fungi have been reported, but mostly in individuals with immuno-compromise secondary to HIV infection, chemotherapy, or iatrogenic immunosuppression (Meltzer and Kelemen, 1959; Chandler, 1968).

Diabetics have impaired glucose metabolism, which degrades systemic and local defense mechanisms in several ways (Table 4). The ability of white blood cells to phagocytize and kill some types of invading bacterial organisms is significantly diminished in the presence of high sugar levels. Occlusion or obliteration of small blood vessels secondary to diabetic microangiopathy decreases blood flow to a variety of tissues. Decreased blood flow impairs the delivery of infection-fighting white blood cells as well as systemically administered antibiotics. Perhaps critical to the development of malignant otitis externa is the altered character of diabetic cerumen. Cerumen taken from the canals of diabetics shows consistently elevated pH. Since the acidic environment of the normal external auditory canal is especially important in controlling the growth of Pseudomonas, the alkaline environment found in diabetic individuals appears to be a principal etiologic factor in the development of acute malignant otitis externa (Barrow and Levenson, 1992; Farr et al., 1992).

The introduction of exogenous water into the external auditory canal is also a very important etiologic factor. The disease was first described in south Florida, an area where larger than usual numbers of elderly diabetic individuals are likely to swim. Moreover, several recent studies have implicated the deliberate introduction of water into the external auditory canal for the purpose of cerumen removal in the development of malignant otitis externa. Thus, it appears that the disease is often partly iatrogenic in etiology. Introduction of water into the external auditory canal of diabetics should either be avoided entirely or followed by the installation of acidifying solutions to restore normal pH (Fabricant and Perlestein, 1949; Chandler, 1972; Anson and Bast, 1980; Reid and Porter, 1981; Calderon and Mood, 1982; Schuknecht and Gulya, 1986; Goidl, 1987; Driscoll et al, 1993).

Malignant otitis externa begins as a simple external otitis. Consequently pain, otorrhea, and conductive hearing loss are the principal symptoms. Unlike simple external otitis, however, malignant externa does not respond to treatment. Pathologically, infection appears to enter the temporal bone through the small fissures of Santorini on the inferior/anterior canal wall near the bony cartilaginous junction. A tuft of granulation tissue can frequently be seen in this area and is an important diagnostic hallmark of the disease (Fig. 8). As disease spreads throughout the temporal bone and skull base, cranial nerve paralysis often develops. Facial paralysis is most common. Paralysis of the lower cranial nerves is associated with tongue paralysis, hoarseness, dysphagia, and aspiration. Advanced cases may result in spread of the disease through the clivus to the contralateral temporal bone or in intracranial extension (Chandler, 1972; Senturia et al., 1980; Seyfried and Fraser, 1980; Hawke and Jahn, 1987; Strauss, 1990).

Treatment

Any diabetic individual who has external otitis that does not respond to usual medical management has malignant otitis externa. The
diagnosis is a clinical one. Patients almost invariably have a significantly elevated erythrocyte sedimentation rate (60 mm/hour or above). An elevated erythrocyte sedimentation rate can help confirm diagnosis but, more importantly, falling erythrocyte sedimentation rates are an encouraging sign that the disease is responding to treatment. Technetium 99m bone scan is quite sensitive to osteocyte activity and consequently to bony involvement. It can be useful in determining the extent of the disease. Since bone scans will remain positive for many years after infection, they are of little use in monitoring the response to treatment. Computed tomography scans can help identify areas of bony erosion and abscess formation, which may occur in more severe infections (O'Sullivan et al, 1978; Parisier et al, 1982; Rubin et al, 1990).

Using a combination of clinical indicators, including cranial nerve paralysis, and radiographic evaluations, the disease can be staged as follows (Bopp and Reed, 1987):

**Stage I:** infection of the external auditory canal and contiguous soft tissues with or without facial nerve involvement.

**Stage II:** extension of the infection to include osteomyelitis of the skull base and involvement of other cranial nerves, especially cranial nerves IX–XII.

**Stage III:** intracranial extension.

Effective treatment requires administration of appropriate systemic antibiotics long enough to eradicate the infection. Surgical debridement with drainage of abscess plays only an adjunctive role.

The mainstay of treatment for many years has been intravenous double antibiotic therapy directed at Pseudomonas. Selection of antibiotics should be based on specific sensitivities against the organism cultured from the infection. It is frequently necessary to reculture because Pseudomonas aeruginosa can develop resistance to specific antibiotics quickly. Treatment should be continued for a minimum of 6 weeks and much longer treatment periods are frequently required.

Recent data suggest that it is frequently possible to treat Stage I disease with oral fluoroquinolones alone. Ciprofloxacin appears to be the most effective. Approximately 80 percent of individuals will be cured of infection using only a protracted oral course of fluoroquinolone therapy. Patients need to be carefully monitored for development of resistance and treatment failure. Those individuals whose infection does not appear to be responding should be switched promptly to intravenous therapy with two appropriate antibiotics.

It is frequently difficult to determine when therapy can be safely stopped. Technetium scanning is not helpful because it will remain positive for years. Gallium and indium radionucleotide scanning, however, remains positive only in the face of active infection. Treatment should be continued until gallium or indium scans show no evidence of active disease. The erythrocyte sedimentation rate should have returned to normal before therapy is terminated. While the recovery of cranial nerve function is a promising sign, some nerves are so damaged that recovery does not occur (Parisier et al, 1982; Bopp and Reed, 1987; Strauss, 1990).

Individuals with Stage II or Stage III disease that do not respond to even prolonged administration of systemic antibiotics may benefit from the use of hyperbaric oxygen (Land et al, 1991).

**REFERENCES**


