Idiopathic Inflammatory Medial Meatal Fibrotizing Otitis

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Background: Idiopathic inflammatory medial meatal fibrotizing otitis (IMFO) is rare. Only a few cases with unknown cause have been reported.

Objective: To report 3 cases of IMFO as a specific diagnostic entity.

Patients and Methods: Two adults and 1 child with bilateral IMFO were observed for several years at the Department of Otorhinolaryngology of Helsinki University Hospital, Helsinki, Finland.

Results: Only the osseous part of the external ear canals was affected by IMFO. The skin and skin organs over the lateral cartilage of the ear canals remained rigorously and constantly uninflamed through the active, relentless progression of the disease over several years, resulting in the formation of a fibrous plug of the medial meatal canal. The middle ears and mastoid air cells were not affected during the active inflammatory phase.

Conclusion: IMFO has its own specific pathophysiologic characteristics, and perhaps also etiopathologic characteristics, which are still unknown.


A CQUIRED ATRESIA of the external ear canal is relatively uncommon and may be postinflammatory, traumatic, postoperative, or neoplastic.\(^1\),\(^2\) Reflecting this heterogeneous etiology, numerous synonyms have been used to describe the postinflammatory category, including atresia meatus acusticus externus,\(^3\) chronic stenosing otitis externa,\(^4\) acquired external auditory atresia,\(^5\) postinflammatory acquired atresia,\(^6\) postinflammatory medial meatal fibrosis,\(^7\),\(^8\) and medial meatal fibrosis.\(^9\)

Katzke and Pohl\(^7\) described a series of 6 patients with postinflammatory medial meatal fibrosis and suggested that this progressive disease was a discrete clinicopathologic entity in which the granulation process produced the final fibrous plug. However, Slattery and Saadat\(^8\) found, in their review of 24 cases of postinflammatory medial meatal fibrosis, a condition that they subclassified as an idiopathic postinflammatory medial canal fibrosis: 4 patients with no readily identifiable cause for the development of the atresia. Following these reports, we describe an additional 3 bilateral idiopathic medial meatal atresia cases in which the disease presents as a defined entity.

METHODS

Two of the patients were referred to the Otorhinolaryngology Department of the Helsinki University Hospital because of continuous external ear canal infection and granulation. The third patient, when referred to the clinic, had already developed a medial canal fibrous atresia. Common for all of these patients was an infectious, chronic, granulative but painless process in both medial ear canals. Clinically, no patient had any ongoing middle ear or mastoid symptoms. Computed tomographic scans demonstrated variability in the medial meatal fibrotizing otitis: from somewhat shadowed ear canals and thickened tympanic membranes to medial meatal atresia (Figure 1). Individual times for the progression of the inflammation into the atresia phase and the preoperative and postoperative hearing status are shown in the Table.

Bacterial cultures grown out during acute infected phases have varied: *Escherichia coli, Serratia marcescens, Streptococcus β-hemolyticus* group G, but mostly *Staphylococcus aureus* or the bacterial cultures have remained negative (Table). Fungal cultures have been negative (Table). In more distinct inflammation periods, no particular bacterial over-

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growth in culture specimens has occurred in any of the patients.

Histologic specimens from the resected fibrous plug taken during surgery and the meatal tissue specimens tested during different phases of the ongoing inflammation have shown no specific pathologic conditions. They have shown dense fibrotic tissue or chronic lymphocytic inflammation cytologically, and proliferative small vessels, some fibroblasts, scarce collagen fibers, parakeratosis, and scanty calcification. Histopathologic findings from all 3 patients showed no tuberculous granulomas. In the florid inflammatory stage, histologic specimens taken from the meatal tissue lining the bone have shown no stratified epidermal lining or dermis. Except for ear canal symptoms, all 3 patients were healthy and had no immunologic deficiencies.

### RESULTS

#### CASE 1

A 5-year-old girl had a persistent aural discharge, meatal infection, and ear canal granulation bilaterally. Otherwise, her general health was good. She had previously had tympanostomy tubes inserted because of secretory otitis media and acute recurrent middle ear infections without tympanic membrane perforations. A year thereafter, when the middle ear symptoms had clinically resolved, the meatal infection and/or inflammation began.

On her first clinic visit, she had no discharge through the tympanostomy tube still in position on the left side, and both middle ears were clinically uninfected. Both tympanic membranes were thickened, and the medial ear canal was completely healed. The medial meatal inflammation and/or infection then reactivated. The patient underwent several medical interventions with varying combinations of treatments including topical applications of antibiotics and steroids in eardrops, ear packing, thorough and regular ear irrigation, a few sessions of surgical abrasion of granulation tissue, and oral antibiotic and steroid treatments in combination with local treatment or without local treatment. The medial ear canal skin in both ears never completely healed.

Her right ear canal was medially sealed with fibrous tissue by age 6 years (Figure 2). The infection and/or inflammation stabilized in the right ear, and the skin epithelial lining over the atresia has not shown symptoms since the final fibrosing. The meatal skin lateral to the bony canal has been healthy in both ears. The boundary between the healthy and inflammatory skin has been distinct throughout her medical history in both ear canals.

At age 9 years, her left ear canal active process continues. In noninfectious stages she seemed to gain some benefit from a potent local steroid treatment (betamethasone). To prevent total fibrosing, treatment with oral prednisone and clarithromycin was started, but 4 months of this treatment has provided no permanent solution.

In pediatric consultations, the patient’s general health has been consistently excellent, and no immunologic deficiencies have been found. Test results for C-reactive protein have been negative; hemoglobin levels and complete blood cell counts, normal; erythrocyte sedimentation rates, less than 10 mm/h; and α1-antitrypsin values, normal. Immunoglobulin levels, including IgG subclasses, have been within normal limits.

#### CASE 2

A 55-year-old otherwise healthy man had continuous external ear canal infection and/or inflammation for 2 years. Clinical examination revealed purulent external ear canal infection with granulation tissue in both ear canals. Infection and granulation affected only the medial meatal skin granulated (Figure 1). The tube was extracted and protruding granulation tissue mechanically removed. Local treatment with topical antibiotic-steroid drops was started. Over the next several visits, the fulminating meatal canal granulative infection resolved. After tube extraction, the tympanic membrane perforation healed in a few weeks, but the medial meatal skin did not heal entirely (Figure 2).

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### Clinical Follow-up of Progression of Disease Into the Atresia Phase, Hearing Status, and Microbiological Findings From Ear Canals

<table>
<thead>
<tr>
<th>Sex/ Age, y</th>
<th>Time to Atresia, y (Right/Left)</th>
<th>Hearing, dB (Right/Left)</th>
<th>Bacterial Culture</th>
<th>Fungal Culture</th>
</tr>
</thead>
<tbody>
<tr>
<td>F/5</td>
<td>1.5/3*</td>
<td>30/5-30</td>
<td>Not operated on</td>
<td>13 × Negative, 6 × Staphylococcus aureus/epidermidis, 1 × Haemophilus influenzae, 1 × Streptococcus pneumoniae, 1 × Escherichia coli, 2 × Staphylococcus β-hemolyticus group G</td>
</tr>
<tr>
<td>M/55</td>
<td>3.5*/3.5*</td>
<td>10-15/10-15</td>
<td>Not operated on</td>
<td>3 × S aureus, 1 × Escherichia coli, 2 × Staphylococcus β-hemolyticus group G</td>
</tr>
<tr>
<td>M/44</td>
<td>11/12</td>
<td>45/45</td>
<td>25/10</td>
<td>1 × Negative, 4 × S aureus, 2 × E coli</td>
</tr>
</tbody>
</table>

*Continuing follow-up.

†Haemophilus influenzae and S pneumoniae have been found in the year 1997.

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atal canal and tympanic membrane. The granulation tissue was mechanically removed during the next several visits, and infection was treated with topical antibiotic and steroid eardrops. He never noticed any middle ear symptoms. A temporal bone computed tomographic scan revealed entirely healthy middle ears and mastoid cavities (data not shown).

The active infection and prominent inflammation resolved, but the epidermal layer of the medial meatal skin was in very loose contact with the scanty inflammatory derma, and the epidermal layer desquamated in large pieces under very gentle touch. The boundary between the healthy lateral canal skin and the affected medial canal skin has been very distinct at every stage of the disease: the cartilaginous area of the ear canal skin has remained normal even in the purulent discharge periods. Under thorough, active, local and periodic oral prednisone and antibiotic treatments (ciprofloxacin, clarithromycin), the recurrent infections have eased, but the basic inflammatory stage persists and progresses slowly but relentlessly (Figure 4). At this stage, the best response has been to a potent local steroid treatment (betamethasone). During local steroid treatment, no granulation has been seen, but the skin is permanently eczematous and defective with a white fibrinous cover (Figure 5).

CASE 3

A 44-year-old man presented with a history of continuous external otitis symptoms over the last 10 years, for which he regularly used topical antiseptic and antibiotic-corticosteroid drops. At age 40 years, stricturous atresia was noted in the middle of the ear canals, and medial to that, eczematous inflammation was found. The ear canals then experienced stenosis with fibromatous medial canal atresia.

After the stenosis progression of this inflammation was noticed, the right ear canal developed a total medial atresia within 1 year; on the left, progression to the stenosed quiescent stage took 2 years. After the formation of the fibromatous plug, the inflammation disappeared completely.

COMMENT

Postinflammatory meatal atresia is most often caused by chronic otitis media or by recurrent external otitis.1 Slattery and Saadat8 conclude that the small sample size...
in their own and previous reports precludes generalization in regard to the bilateral nature of the idiopathic medio canal atresia disease. The specific cause of the condition is unclear.7,10 It has been proposed that loss of the squamous epithelium on the lateral surface of the tympanic membrane results in exposure of its fibrous layer, and in the presence of ongoing inflammation, healing occurs through granulation formation.2,31 The stenosing process tends to continue up to the junction between the bony and cartilaginous canal.2 The active, relentlessly progressive stages are followed by a quiescent, mature fibrotic phase in which conductive hearing loss is the main (and often only) symptom.

In most published series, etiologies show variability even in a single patient. This poses a challenge to exact diagnosis and treatment, although the final status is fairly distinct and consistent regardless of possibly differing pathophysiologic etiologies. The literature also includes a few cases in which chronic otitis externa is considered the primary and only etiopathologic factor in the fibrotizing process.3,6

The middle ear status of our patients is normal or healed, with no major chronic purulent middle ear symptoms (currently or previously). Clinically and radiologically, the inflammation and infection have been found to be restricted to the medio canal skin over the bony part of the canals and tympanic membranes in all 3 patients. In children, acquired fibrosis is even more rare than among adults. Keohane et al9 described a child who underwent surgery (tympanomastoidectomy) for suspected chronic middle ear disease, but the middle ear and mastoid were found to be normal at surgery in both ears. The child in the present study developed medio canal atresia without ongoing middle ear disease, as verified clinically and by computed tomography.

Our 3 patients share several features. The disease is a bilateral, local fibrotizing inflammation of the medio canals with poor response to conservative and surgical interventions. The symptoms are “quiet” throughout the history: even in acute phases with purulent infectious discharge and granulation, there is usually no pain. The only complaint besides the discharge in these phases is that hearing is abnormal. The infections, like the chronic inflammatory reactions, are completely restricted to the medio canal skin covering the tympanic membrane and the bony part of the ear canal. The inflammatory reaction does not affect the skin with its organs covering the cartilaginous part of the ear canal, even at the time of the more suppurative stages of the disease. Clinically, the boundary between inflammatory and healthy skin is astonishingly sharp. The first signs of atresia reaction are a circular web formation at the border of the cartilage and the bony ear canal. One common peculiarity is that the atresia in these patients does not obliterate the canal; instead, the medio canal is blunted and filled up with a fibrous plug.

The etiology seems to be not purely infectious in that the process continues even when, clinically, the infection has stabilized. Additionally, the inflammatory reaction is best controlled by treatment with a potent corticosteroid rather than antibiotics. The use of topical antibiotics alone in the inflammatory stage seems to give no benefit, and when used in combination with corticosteroids, they give no auxiliary benefit.

The medio canal skin of the ear canal has certain peculiar characteristics that distinguish it from skin anywhere else on the body: the dermis here is attached directly to the bone with no subcutis; the skin is very thin and has no skin organs; it has a lateral migratory keratin desquamation capacity; and this area is the only locus (excluding the natural orifices of the body) where the endodermal and ectodermal epithelia are so close after birth that under pathologic conditions they may come into direct contact and interfere with each other.

A histologic specimen taken from the affected skin area in the nongranular and/or noninfectious stage shows no epithelial lining, which is very interesting. The inflammation and/or disease stabilizes by itself when the skin over the bony canal has been replaced by the fibrous plug.

The chronic progression and the response, although not permanent, only to corticosteroid treatment raise the suspicion of an autoimmune disease or a dermolytic skin defect prone to secondary purulent infections and granulation formation. As Stoney et al11 speculate with regard to granular myringitis, “The initiator is almost certainly some form of local trauma or microbial infection, which destroys the epithelium, exposing the fibrous layer. The inflammation is then confined to the epithelia and underlying fibrous layers.” In our patients, the process seems to be a continuous inflammatory condition and not primarily the result of repeated fulminating infectious episodes, although an infection seems always to have been a triggering factor. Also, the inflammation predisposes the tissue to recurrent infections, recurrent granulation formation, and scarring. Based on these cases, we suggest that idiopathic inflammatory fibrotizing chronic otitis of the medio canal is its own clinical and/or pathologic entity.

Accepted for publication May 20, 2002.
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REFERENCES