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Cholesteatoma of the Tympanic Part of the Temporal Bone

Traian S. Ataman

Otological Department, Phono-Audiological and ENT Functional Surgery Institute, Carol Davila School of Medicine and Pharmacy, and Functional Surgery Institute, Bucharest, Romania

Abstract: This article presents accounts of two patients with cholesteatoma of the tympanic part of the temporal bone, located immediately lateral to the tympanic annulus (and with an intact tympanic membrane). The lesions were located deep in the anterior and inferior walls of the canal, especially in the vaginal process of the tympanic part. These more severe cases required surgical correction (removal of the sac of cholesteatoma) with very good results. Pathogenesis and differential diagnosis are discussed.

Key Words: cholesteatoma; differential diagnosis; external auditory meatus; surgical removal; temporal bone; tympanic part of the temporal bone

The tympanic part of the temporal bone forms all of the anterior and inferior walls and a part (inferior and medial) of the posterior wall of the external acoustic meatus. This part is formed from the anterior and posterior horns and the vaginal process. During the intrafetal period of ontogenesis and sometimes in postnatal life, the vaginal process presents a small foramen (of Huschke) that represents a nonossified part of the plate.

The anterior horn is separated by the glaserian petrotympanic fissure from the petrous part and squama. The glaserian fissure consists of the chorda tympani nerve, Raw's anterior malleal ligament, and Nager's anterior tympanic artery and veins. The tympanomastoid fissure separates the posterior horn from the mastoid process, and it is a landmark of the fallopian aqueduct and facial nerve.

CHOLESTEATOMA OF THE EAR

Cholesteatoma of the ear is, from a topographical point of view, a syndrome, because it includes cholesteatoma of the middle ear (located in the tympanic cavity, in the mastoid process, or in both) and cholesteatoma of the petrous air cells (anterior air cells, posterior air cells, and apical cells). However, at the same time, the epidermal plug and hyperkeratinized epidermis of the external auditory meatus (EAM) belong to the same pathogenic and histopathological category [1–3].

Simultaneously, many categories of "common" middle-ear cholesteatoma exist, including congenital (or primitive) cholesteatoma with an intact eardrum, described by House in 1953 [4]. It is explained by embryological theory as remnant embryonic cells from the first branchial cleft. Thus, this is an epithelial dystopia in a mucous territory of the first branchial cleft.

The invagination theory explains the cholesteatoma that occurs in the case of the retraction pocket or adhesive otitis media. Sade in 1979 considered it like an epithelial migration defect with an attical retraction pocket [5]. Cole et al. in 1981 observed the existence of a metabolic disturbance with hyperkeratinization [5]. However, Ataman et al. [5] observed that only one-third of patients with adhesive otitis media have associated cholesteatoma. Therefore, cholesteatoma is not considered to be the last stage of adhesive otitis media; on the contrary, the two are considered to be different entities.

Secondary, acquired cholesteatoma, with a perforation of the eardrum, is explained by the epithelial invasion theory. The squamous keratinized epithelium invades through the tympanic membrane perforation into the middle ear along the contact guidance surface. The squamous metaplastic theory takes into consideration the

Reprint requests: Traian Ataman, MD, PhD, University of Bucharest Romania, Carol Davila School of Medicine and Pharmacy, Str. Poenari nr. 1, Bl. 13, SC.A, ap. 10, Sector 4, Bucharest, 040384 Romania. Phone: +40 21-331-3302; Fax: +40 4100278; E-mail: nora_ataman@yahoo.com

metaplasia of the mucous membrane of the middle ear into squamous keratinized epithelium. Sade in 1971 sustained the theory, showing that the mucous cells are pluripotent cells when they are stimulated by infection [5].

It has been shown that severe deficiency of vitamin A may provoke keratinized epithelium formation. The basal cell hyperplasia theory explained the occurrence of cholesteatoma as resulting from the invasion of the subepithelial tissue by proliferative epithelial cells. Ruedi in 1959 clinically confirmed this theory [5]. However, in this pathogenesis, the basal membrane must be altered so that the lamina propria may be invaded by the epithelium. The rupture of the basal membrane was also demonstrated. A special case is posttraumatic cholesteatoma, when trauma allows an enclave of the epithelial tissue into the middle ear.

Cholesteatoma in chronic otitis media and mastoiditis may accumulate the keratin mass slowly and may create some complications. Cholesterol granuloma is chronic granulation tissue containing cholesterol crystals [2,6]. No evidence exists to support the hypothesis that cholesteatoma can develop by metaplasia of the mucous membrane [7].

Cholesteatoma may be found in very rare cases behind an intact tympanic membrane without a prior history of otorrhea. This is a true congenital (or primitive) cholesteatoma. The tendency of cholesteatoma to cause bone erosion may lead to local destruction. The cholesteatomatous masses often are infected and, when accompanied by bone erosion, can lead to severe complications. A distinct category is cholesteatoma of the postoperative cavities (in radical mastoidectomy). Meatoplasty of the postoperative cavity is contrary to the concept of cholesteatoma definition.

PATIENT 1

We admitted PI, a 12-year-old girl, to our clinic on September 20, 1999, for mucopurulent otorrhea in the right ear, which occurred 4 months earlier after repeated bathing in the swimming pool. We treated her for 4 months with drops into the ear canal of borated alcohol and rifampicin solutions, without any improvement of symptoms.

Otoscopic examination of the right ear revealed a huge granulation tissue that filled the entire external auditory canal and was covered with mucopurulent discharge. After otomicroscopical examination, we removed the granulation tissue and noticed a fetid, purulent discharge and massive keratin debris with bone erosion of the inferior wall of the EAM. The tympanic membrane was intact and mobile but thicker, matted, and bearing preserved landmarks.

Voice tests and tuning-fork tests emphasized normal audition. Pure-tone audiometry also emphasized normal audition. Microbiological assessment of the ear discharge revealed an infection with *Pseudomonas aeruginosa*, sensitive to gentamicin, carbenicillin, colistin sulfate, norfloxacin, and ceftriaxone. Routine laboratory study results were within normal limits.

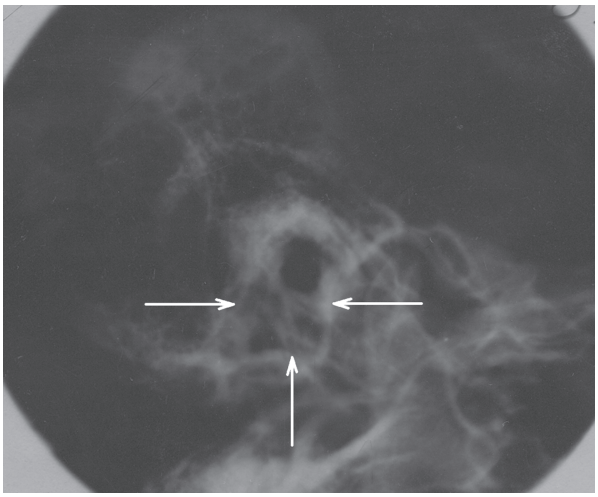

Figure 1. Radiological examination of the right ear of patient 1 (12-year-old girl) showed bone-destructive lesions of the inferior wall of the external auditory meatus.

Radiological and tomographic examinations of the right ear (in Schuller and Chausse III views) showed bone-destructive lesions of the inferior wall of the EAM (Fig. 1). We decided on a surgical approach and operated on October 4, 1999, with the patient under general anesthesia. We made a semicircular endomeatal incision on the inferior wall of the meatus and elevated and fully removed the entire cholesteatomatous sac. It penetrated into a huge cavity (2.5 × 3 cm) located in the internal and medial third of the right EAM.

We drilled the cavity (and a part of the inferior wall of the EAM) and removed a bloody granulation tissue. We also noticed and drilled osteitic lesions. Accidentally, we opened the canal of the chorda tympani nerve during surgery and damaged the nerve. For hemostatic purposes, we filled both the cavity and the EAM with compressed Gelfoam and added an auriculotemporal packing.

A long, postoperative course ensued, owing to recurrent granulation tissue that necessitated repeated removals. Reepithelialization of the postoperative cavity was slow and lasted for approximately 3 months. Since then, follow-up examinations over a 3-year period have revealed no local recurrence.

PATIENT 2

We admitted PS, a 28-year-old woman, to our clinic on February 12, 2002, for fetid, persistent otorrhea of the

right ear, associated with ipsilateral hemicrania and temporomandibular arthralgia with an important reduction of temporomandibular joint mobility. The apparent onset of the disease is placed in 1984, with violent right otalgia and fetid, mucopurulent otorrhea, apparently in the presence of an acute pharyngoamygdalitis. Antibiotic therapy was efficient in treating the right acute otitis externa and media.

Between 1984 and 2002, the patient had four other similar events, of which one episode in 1988 was noticeable, when trismus and functional impotence of the right temporomandibular joint were added to the initial symptomatology and feeding by a tube was imposed for 3 months. General antibiotic therapy, local suction, and local instillation of antibiotic agents were undertaken. In 1999, a tonsillectomy was performed, on the supposition that this was the point of infection.

In December 2001, the patient had the last episode with violent, permanent, and progressive right otalgia associated with ipsilateral hemicrania and trismus but without otorrhea and hearing loss. Despite massive antibiotic therapy, the symptoms did not abate; on the contrary, a fetid, pulsatile, green-yellowish, viscid discharge developed additionally.

In January 2002, we admitted the patient to our clinic for the aforementioned symptomatology. The physical examination revealed trismus due to the right temporomandibular arthralgia and right submandibular lymphadenopathy (1.5 cm in size). Otoscopic and otomicroscopical examinations revealed fetid, mucopurulent discharge in the right EAM, huge masses of whitish keratin debris, and a huge erosion of the anterior and inferior walls of the EAM in their medial third.

After the suction of discharge and removal of the cholesteatoma, the tympanic membrane became visible and was seen to be intact and mobile (but a little thicker), with its landmarks present. Tuning-fork tests and audiometric examinations revealed normal audition. Routine laboratory study results emphasized a nonspecific chronic inflammatory syndrome.

Radiological and tomographic examinations (in Schuller and Chausse III views) revealed normal pneumatization of both mastoid processes and the presence of a large bone erosion, relatively well delimited in a double sac, of the right tympanic part of the temporal bone, suggestive of cholesteatoma (Fig. 2).

On February 12, 2002, we performed surgery with the patient under general anesthesia. We made a semicircular incision of the inferior wall of the right EAM and removed the entire sac of cholesteatoma. We found a large cavity (some 1.5–2 cm toward the anterior and inferior walls of the right EAM). We found the inner one-third of the anterior wall of the tympanic part of the temporal bone both completely eroded toward the parotic region and changed into a fibrous wall.

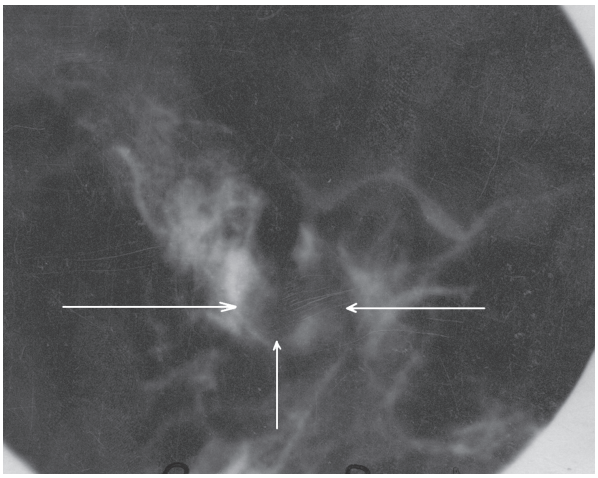

Figure 2. Radiological examination of patient 2 (28-year-old woman) revealed normal pneumatization of both mastoid processes and the presence of a large erosion of the right tympanic part of the temporal bone, suggestive of cholesteatoma.

We blindly elevated and totally removed the sac of cholesteatoma, which necesitated massive drilling of the inferior wall of the EAM. Surgical exploration of the cholesteatomatous cavity confirmed the imaging examination: The cavity had no communication with the tympanic cavity and mastoid process.

For hemostatic purposes, we filled the entire postoperative cavity and the EAM with compressed Gelfoam and added auriculotemporal packing. After a couple of weeks of antibiotic therapy, a very good—and surprisingly short—postoperative course followed (with the disappearance of trismus and of all the symptoms). During the 8-month follow-up period, we have noticed no local recurrence.

DISCUSSION

Cholesteatoma of the EAM is a rare pathological finding that entails some different entities. Keratosis obturans is, for some authors, considered to be in the same field as cholesteatoma. Keratosis obturans is caused by a combination of hyperkeratosis and failure of normal epithelial migration of the EAM's skin. Zuehlke described pseudocholesteatoma of the EAM in patients with osteoma of the EAM [5]. Ombredanne and Porte in 1962) [8] and Schucknecht in 1989 [9] described cholesteatoma of the EAM occurring in association with congenital aural atresia.

Cholesteatoma of the EAM on a nonatretic ground is extremely rare. The pathogenesis of cholesteatoma of the tympanic part of the temporal bone involves some hypotheses. Aberrant air cells that have migrated into the plate of the tympanic part might be a plausible

explanation for this kind of cholesteatoma. The persistence of Huschke's foramen could be another explanation. The foramen of Huschke is normally present during growth before the age of 2 years. Sometimes, it persists into adult life as an anatomical variation, and the skin of the EAM is invaginated into the opening and under the inferior wall of the meatus [10].

Another hypothesis involves an atypical coloboma auris as a point of departure, developing into such a cholesteatoma. The differential diagnosis could include the following entities: cervical Bezold's otomastoiditis, Mouret's jugodigastric otomastoiditis, cancer of the EAM, ceruminoma of the EAM, glomus jugulare tumor, and epidermal plug.

Exploration of cholesteatoma of the EAM advances our knowledge in this field. Gray's definition of cholesteatoma ("skin in a wrong place") appears to be obsolete [5]. The skin of the meatus is relatively thick (similar to the integument of the auricle) in the cartilaginous part of the meatus. However, the skin in the bony portion of the meatus is very thin, and it appears completely different, like a mucous membrane. The changes in the behavior of this EAM integument could be the starting point of the cholesteatoma of the tympanic part of the temporal bone. A positive diagnosis can be confirmed during surgical exploration. Surgery must also be adapted to the local situation, often requiring a laborious and iterative operation.

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