Management of Congenital Cholesteatoma with Otoendoscopic Surgery: Case Report

Konjenital Kolesteatomun Otoendoskopik Cerrahi ile Tedavisi

Ayache STEPHANE, MD[a]

[a]Orpac Clinique Du Palais 25 Avenue Chiris 06130, FRANCE

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Yazışma Adresi/*Correspondence:*
Ayache STEPHANE, MD
Orpac Clinique Du Palais 25 Avenue
Chiris 06130, FRANCE
ayachestef@hotmail.com

ABSTRACT The author aims to describe the features of congenital cholesteatoma in adults and the value of the exclusive otoendoscopy in the surgical management. The congenital cholesteatoma is well known in children. This disease is uncommon in adults and is described here through a case-report in a 25-year-old patient. The forms localized to the upper posterior part of the tympanic cavity are the least common and can take the features of an acquired cholesteatoma. The surgical treatment is usually performed with otomicroscopy, with a canal wall-up tympanoplasty. In this case, the management was performed with a transmeatal approach with the exclusive otoendoscopic surgery. This procedure allows an access to anatomical areas hidden for the microscope. It belongs to a new surgical era in otology and is one of the minimal invasive surgical procedures of the future.

Key Words: Cholesteatoma, middle ear; endoscopy

ÖZET Yazar erişkindeki konjenital kolesteatomun özelliklerini ve cerrahi tedavide seçkin otoendoskopinin değerini tanımlamayı amaçlamaktadır. Konjenital kolesteatom çocuklarda iyi bilinmektedir. Bu hastalık erişkinlerde nadir görülür ve burada 25 yaşındaki bir hastadaki olgu sunumu vasıtasıyla anlatılmaktadır. Timpanik kavitenin üst arka kısmında yerleşmiş olan formlar en az görülür ve kazanılmış kolesteatomun özelliklerini alabilir. Cerrahi tedavi genellikle kapalı kavite timpanoplasti ile otomikroskopiyle yapılır. Bu olguda tedavi transmeatal yaklaşımla seçkin otoendoskopik cerrahiyle yapıldı. Bu işlem mikroskoptan saklanan anatomik alanlara ulaşılmasını sağlar. Otolojide yeni bir çığır açmıştır ve geleceğin minimal invaziv cerrahi işlemlerinden birisidir.

Anahtar Kelimeler: Kolesteatom, orta kulak, endoskopi

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Congenital cholesteatoma (CC) is a rare but well documented disease in the international literature. It occurs usually in the childhood, in the upper anterior part of the tympanic cavity. The surgical management is performed with the microscope with difficulties depending on whether the lesion is limited or not.

Through a case-report of a CC in a 25-year-old adult, features and the exclusive endoscopic surgical management of this disease are described. The primary otoendoscopy is a minimal invasive procedure which allows the removal of the disease with a transmeatal approach in anatomical regions usually hidden for the microscope.

CASE REPORT

A 25-year old man presented with a progressive right ear hearing loss without any previous otologic history.

The eardrum was remarkable by only a hyperhemic area on the malleus and the upper posterior part of the eardrum (Figure 1). The audiologic test indicated a right conductive hearing-loss, with an air-bone gap of 20dB. The computerized tomography (CT) of the petrosal bones showed a blurry postero-superior opacity with an extension between the incus and the pyramid of the stapes. The incudostapedial joint was eroded. The antrum and the mastoid were free from disease (Figure 2).

All the informations about the surgical procedure were explained to the patient: The objectives, the transmeatal approach and the possibility of an antromastoidectomy.

The surgical procedure was endoscope-assisted and under general anesthesia. A tympanomeatal flap was created from the 11 o'clock position to the 7 o'clock position (3 mm 0° endoscope, HD camera, Karl Storz, Germany) with a transmeatal approach.

A cholesteatoma was discovered, without any contact with the deep side of the tympanic membrane, suggesting a congenital cholesteatoma. The lesion was in the upper posterior part of the tympanic cavity, reaching the second portion of the Fallopian canal, leaving the canal undamaged, and extended into the retrotympanum and especially the oval window. The long process of the incus was eroded. Only the anterior crus of the stapes remained, as well as a functional footplate (Figure 3).

Complete excision of the cholesteatoma was performed with the 0° and 30° endoscopes after the removal of the incus. There was no peroperative residual disease in the whole tympanic cavity (Figure 4). A titanium total ossicular prosthesis Kürz was placed between a thin tragal cartilage and the footplate. The tympanomeatal flap was restored to its initial position.

Two years postoperatively the otoscopy shows posterior cartilage reinforcement with a normal eardrum. The residual air-bone gap was 10dB. The CT-scan of the petrosal bones did not show any suspicious opacity that indicated a residual epidermal lesion (Figure 5).

DISCUSSION

CC develops as a result of epidermal remnants from the development of the temporal bone between the 3rd and the 5th week of embryonic life.[1] It mostly appears in young children,[2,3-5] mostly in boys.[2] It is usually a capsular cholesteatoma in the anterosuperior part of the tympanic cavity. The patient is asymptomatic most of the time. This type of CC, as described by Koltai,[6] involves the whole tympanic cavity if left undiagnosed. If operated early, its removal is usually easy and can be performed in a single surgical stage.

This study reports another kind of CC.

It is discovered in a 25-year-old man. Other authors have reported cases of CC in patients older than 50 years of age.[2,3,7] Such adult forms are very unusual. They probably have a different pathophysiology.[8] Their slow evolution explains the usual delayed management. The posterosuperior localization is the least common, usually blurry with the same evolution as an acquired cholesteatoma. Ossicular erosion is the most common complication. In this case report, the air-bone gap was small because the CC created a bridge between the eardrum and the footplate.

The surgical treatment of the capsular form of CC is a canal wall-up tympanoplasty with a transmeatal approach as far as possible, and usually a second stage is not planned.

Forms like in this case report require the same surgical procedure as the acquired cholesteatoma. Kuczkowski recommends open procedures,[9] well-known for their postoperative problems like otorrhea. Most of the authors agree on a closed, more conservative tympanoplasty, with planned surgical revision.[3,5,7,10]

Complete removal of a posterosuperior cholesteatoma like in the case report is achieved most of the time through a canal wall-up tympanoplasty using the otomicroscope with an antromastoidectomy and a posterior tympanotomy.

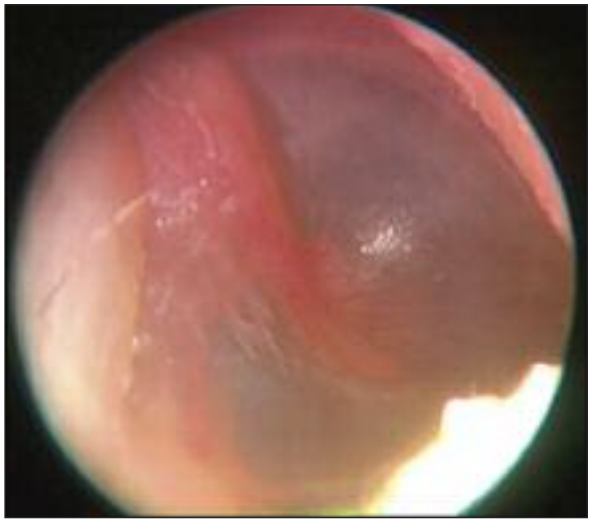

FIGURE 1: Pre-operative otoscopy.

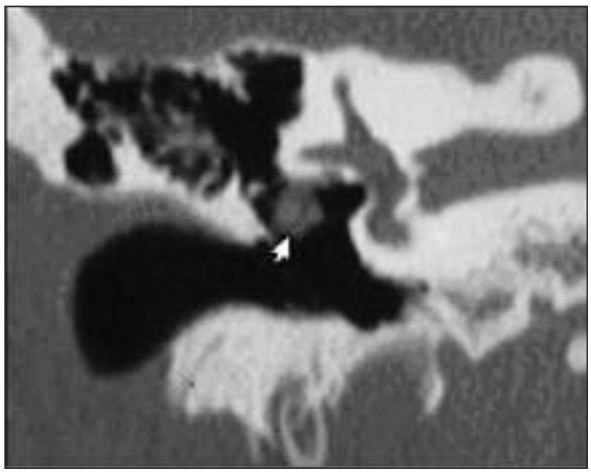

FIGURE 2: Pre-operative CT-scan (coronal plane) *(white arrow : congenital cholesteatoma).*

Endoscope-assisted surgery allows the best access in the more inaccessible regions of the middle ear. The posterosuperior recess of the tympanic cavity, the retrotympanum and particularly the sinus tympani are hidden regions for the microscope and sometimes even after a wide posterior tympanotomy. The value of an additional endoscopic surgery with lateral vision has already been published,[11,12] as well as the primary endoscopic surgical management of the epitympanic cholesteatoma.[13,14]

The tympanoplasty was performed under exclusive otoendoscopy.

As the otoscopy is normal, the first step is diagnostic tympanotomy with a transmeatal approach using a Karl Storz 3 mm 0° endoscope. After performing a tympanomeatal flap, the observation of a poorly defined posterosuperior cholesteatoma without any connection to the tympanic membrane confirms the diagnosis of CC. This step can also be performed with a microscope. However, it may be impossible to perform a complete removal without an antromastoidectomy with a posterior tympanotomy, especially because of the extension of the disease to the sinus tympani. The otoendoscopy allows a very conservative tympanoplasty, without

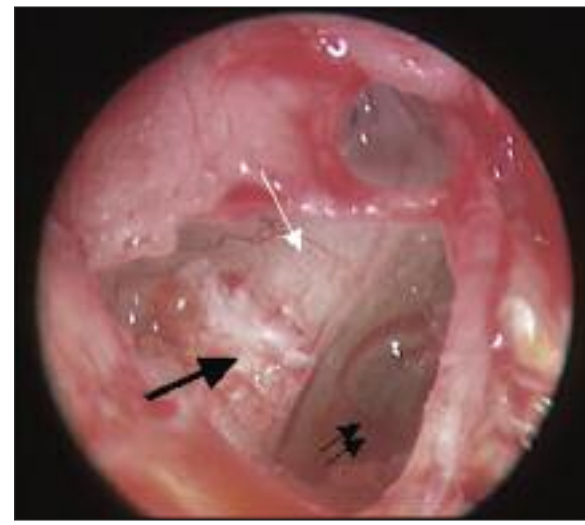

FIGURE 3: Per-operative view *(white arrow: second portion of the facial nerve, black arrow: residual cholesteatoma of the facial nerve, double arrow : stapedial footplate).*

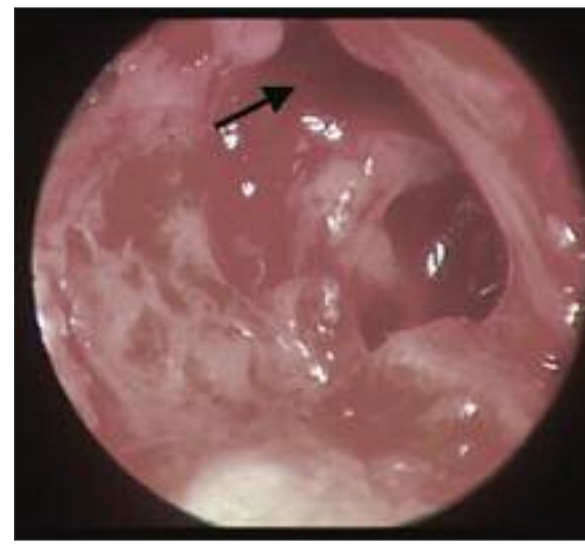

FIGURE 4: No preoperative residual disease after the removal of the incus *(black arrow: posterior epitympanum).*

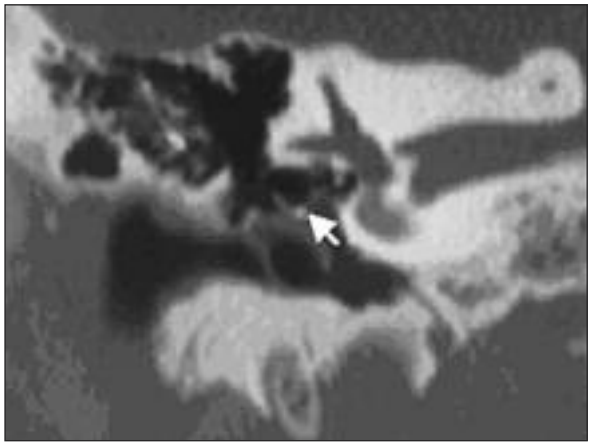

FIGURE 5: 2 years post-operative CT-scan (coronal plane) *(white arrow: total titanium prosthesis).*

any posterior access, particularly a posterior tympanotomy, dangerous for the facial nerve. The analysis of the preoperative CT-scan is decisive in choosing this procedure. The aditus ad antrum is the posterior limit of an endoscopic removal of a cholesteatoma. An opacity of this region on the CT-scan is always searched. The surgeon must follow the disease in the tympanic cavity until the aditus. In case of an extension beyond this limit, an antromastoidectomy must be performed.

Exclusive endoscopic ear surgery allows the surgeon "to introduce his eye" into the middle ear. This procedure is very useful in case of a tympanal procidence, particularly for the posterior tympanal in case of a retrotympanal cholesteatoma.

The procedure is performed with a transmeatal approach. The diameter of the external ear canal can be a limit in case of the presence of wide osteomas or a major procidence of the posterior ear canal wall. Three or 2.7 mm endoscopes can be useful but introducing the instruments may be difficult. In those cases, a canalplasty would be performed.

The ossicular chain reconstruction was performed with a total titanium prosthesis between the cartilage and the footplate. The insertion is performed without aspiration and needs a perfectly dry auditory canal to avoid the prosthesis to stick in blood on the canal.

Endoscopic ear surgery is a "one-hand" surgery. That means that there is no aspiration during the dissection. One hand carries the endoscope and the camera, the other one the instrument. The bleeding is often described as a limit to this procedure. However in fact, bleeding can occur during the dissection of the tympanomeatal flap, but never in the tympanic cavity. The rules of the otologic surgery are also valid in endoscopic surgery : pre-operative preparation of the ear against inflammation of the cavity and cooperation with anaesthesists to avoid any per-operative variation of the blood pressure.

Following the removal of the CC, a cartilaginous reinforcement of the posterosuperior part of the tympanic membrane was performed. The localization of the CC in the posterosuperior part and its non-capsular aspect give to this CC the features of an acquired cholesteatoma and therefore needs the same tympanic reconstruction pattern. Critics of systematic cartilaginous reconstruction could argue the difficulties of otoscopic examination. Therefore most of the authors advocate a second surgical stage. A CT-scan of the petrosal bones plays an essential part in investigating the cavities of the middle ear where the mucosa does not normally present reactional hyperplasia in a CC. A systematic surgical revision will not be planned as a rule for the CC.

CONCLUSION

Endoscopic ear surgery is a new philosophy in the management of the middle ear disease. This is a "one-hand" surgery, which needs a progressive learning curve. Well-known as an additionnal tool, otoendoscopic surgery has become an exclusive procedure, allowing the surgeon to see inside the tympanic cavity without a microscope and avoiding antromastoidectomy or posterior tympanotomy. Although more aggressive, these procedures are often limited to remove the cholesteatoma in hidden anatomic regions. Otoendoscopy is also useful to perform ossiculoplasties or cartilaginous myringoplasties with a transmeatal approach.

This is a minimal invasive surgical procedure of the future.

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