All about Imagistic Exploration in Cholesteatoma

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ABSTRACT

Cholesteatoma is an expansive tissular process, non-neoplastic, well demarcated, developed in the temporal bone, with destructive effect. Cholesteatoma is diagnosed based on clinical, otoscopic examinations, sectional imaging examinations (computerized tomography in high resolution -HRCT and magnetic resonance imaging-MRI).

Examination HRCT on the temporal bone is an exam with high sensitivity, important spatial resolution, it can detect small tissue damage, describes the local architecture, complications, but have low specificity, unable differentiate between tissue masses from different origins.

MRI brings additional information, using conventional sequences in the preoperative evaluation about extension of the lesion, vascular complications but not in residual lesions or relapses. These new data are brought by using new type sequences Echo planar diffusion weighted. (DW-EPI) and non-Echo planar diffusion weighted (DW non-EPI), sequences that have high sensitivity and specificity even for small lesions (5 mm) with avoidance of a second-look surgery.

This article aims to show cholesteatoma aspects, including the definition, history and etymology, classification, histology, clinical signs and the most important imagistic aspects.

Keywords: cholesteatoma, diagnosis, CT, MRI

Cholesteatoma has been known for more than 300 years in the medical literature. Its incidence is 3/100000 to children (0.1% in Europe) and 9/100,000 adults with a predominance in males (1, 2). In the pediatric population, cholesteatomas account for 10% of chronic otitis media cases. It is a serious condition that involves functional prognosis of the ear, sometimes the vital one, by the complications (1-3). The diagnosis of a cholesteatoma at first presentations is based on clinical, but CT and MR imaging is indispensable for preoperative assessment and follow-up (1-4).

Cholesteatoma is of two types, congenital cholesteatoma (CC), specifically for children and acquired that can be shared on children and adults (2-4).

Treatment widely accepted is surgical and technique used is based on otoscopic data and CTHR that can determine the approach way. CT scanning is able to make a perfect descrip-
tion of the anatomy of the middle ear and mastoid space, it showing the relationship of lesion with vessels, facial nerve and inner ear. Preoperative CT can discover very small lesions and lytic effect on temporal bone. (2) CT is often performed when the patient has an infectious episode and middle ear cavities are occupied by inflammatory content, and then CT is unable differentiate between cholesteatoma, the inflamed mucosa, granulation tissue, fibrosis or mucoid tissue (1-4).

In cases where the ear cavities appear occupied by content with different densities, MRI can confirming the presence of cholesteatoma, its extension, giving relationships about local architecture and after surgery, give information about the presence of a residual lesion or looking relapses (2). Limitation of MRI are poor visualization of bony landmark.

**HISTORY AND ETYMOLOGY**

The first specification of a cholesteatoma was made by French anatomist Duverney in 1683, which describes a lesion in the temporal bone. In 1829 Cruveilhier described its features calling it: pearly tumour (2), referring to his shiny appearance. Cholesteatoma was defined in 1959 by Friedman, as a well-defined cystic formation, covered by a stratified squamous epithelium, developed over a fibrous stroma, with a variable thickness, which can contain some elements from the original mucous coating (1). In 1974 Schuknecht has termed it: accumulation of exfoliated keratin located in the middle ear, or in any pneumatic area of the temporal bone derived from the keratinized squamous epithelium (1,2,5). Although, it is a benign formation, it has locally invasive potential, and also can relapse (6).

Cholesteatoma term derived from the Greek (chole-cholesterol, steat-fat; oma - tumour) appeared in literature in 1838, when Johannes Muller, an German anatomical pathologist, had invented the term to describe a tumour that seemed to have comprises fat and cholesterol crystals (7). The term “cholesteatoma” is a misnomer, because the lesion contains neither cholesterol nor fat, is not likely neoplastic, he deriving from the keratinized squamous epithelium (1,2,5). Although, it is a benign formation, he has locally invasive potential, and also can relapse (6).

The classification made by the pathogenesis of the lesion splits it into: congenital (CC) and acquired cholesteatoma (CA), although its origin is indistinguishable, imaging and histologic, and only summing up more features can lead to a positive diagnosis: location, clinical signs, history, otoscopic examination which describe the appearance of the tympanic membrane (9-11).

CC (Figure 1) representing 4-28% of children lesions formed before birth, occurs in children with no history of otitis and to otoscopic examination eandrum is intact (4).

CA (Figure 2) develops exclusive in the middle ear, occurs after birth, most often after several episodes of chronic otitis and in childhood the most common cause is infectious. CA is more aggressively and commonly associated with sensory sequels and intracranial complications (4,5).

CA may be in its turn, primary representing 80% of the acquired and secondary (18%) occurred after various surgical procedures or otologic trauma. Location is in upper posterior part of pars flacida of the tympanic membrane (82%), or postero-superior of pars tensa, in disruption of tympanic membrane (18%) (9,11).

**CLASSIFICATION**

FIGURE 1. Axial (A) and coronal HRCT (B) scans demonstrate a round well-defined lesion (arrow) anterosuperior in the tympanic cavity, medial to the ossicular chain. Note the missing ossicular erosion. Based on the position of the lesion and the lack of bone erosion along with the clinical aspects, this is probably a congenital type.

Acquired cholesteatoma: (D) axial HRCT scan shows tympanic cavity, mastoid antrum enlarged, irregular walls, tissular content (white arrows) with bone lysis chain (thick arrow). (C) contrast-enhanced axial CT – lysis of the posterior petrous pyramid (white arrow) the presence of a small adjacent epidural abscess that has marginal contrast outlet (arrow head), Luc’s abscess or subperiosteal (thick arrow).
CA primary is the result of eardrum retractions that accumulates epithelium desquamation and lose the ability to self-cleaning (7,8). CA secondary is formed by epithelial migration through an eardrum perforation.

Other classifications based on localization criteria have been proposed, one in 1989 conducted by Tos and Lau, has been widely adopted (21) she comprising two categories: atical cholesteatoma that develops in pars flaccida (from Shrapnell membrane) and fill the space Prussak (Figure 3); sinus cholesteatoma from pars tensa that develops from retraction or adhesion of pars tensa (Figure 4).

In evolution, cholesteatoma may extend to a fosete located in posterior recess of the middle ear, (sinus tympani), in which case surgical excision becomes more difficult because of the difficulty of intraoperative visualization of this region. Tos argued that this classification system is useful for selecting surgical procedure and for prognosis, (the lowest recurrence is associated to atical cholesteatoma and the highest for cholesteatoma of sinus tympani).

In 2009, Telmesani and collaborators propose an ATM classification system based on cholesteatoma extension in atica (A), tympan (T) and mastoid (M) (26).

**HISTOLOGY**

Macroscopic appearance of cholesteatoma is: a round or ovoid lesion, pearl grey or yellow, well circumscribed, brittle appearance, and if it have also granulation tissue appears as a soft wax given by tissue inflammation. In electron microscopy cholesteatoma has three components: cystic content, matrix and perimatrix (3, 16). The primary component of lesions is the cystic content, containing keratin scales fully differentiated (anucleate), sebaceous material mixed with, pus or necrotic material. The matrix is made up of stratified squamous epithelium hyper-proliferating and it produces keratin lamellas that reach the cystic part. Perimatrix is the most external area, or lamina propria, consisting of collagen fibres, elastin, inflammation cells (lymphocytes, histocytes, neutrophils), granulation tissue and low bone fragments (3,4,12,16).

CA in childhood contain a higher proportion of inflammation cells in perimatrix than that in adulthood who containing more fibrotic tissue, (this is an indicator that lesions in childhood are more invasive and less reparative) (16). CA associate with bone erosion, reported in 80-90% of cases with a higher incidence in children than in adults.
CC cause less bone erosion chain, with extension that appears late, than those acquired (8-11).

**CLINICAL SIGNS**

Cholesteatoma develops clinical insidiously, there is often a non-aggressive state, remaining undetected for years before obvious clinical signs.

CC may be asymptomatic, discovered by chance in routine ear examination (Figure 3A). When the lesion grows it can cause a decrease in auditory acuity (by bone chain erosion, or by his mass effect) or may have symptoms of acquired cholesteatoma, (without any more infectious episodes), around the age of 5 years. (1,2,4,7,11,17,19,20).

AC are more frequent symptoms, begins in older children (around the age of 10) and adults (12). The main symptom is represented by recurrent painless otorrhea, (foul-smelling otorrhea), in 33%-67% (17) and when it complicates, with an infectious episode, this is difficult to treat to the systemic antibiotic, that’s cannot reach its center by it lack of blood flow. Topical antibiotics often surrounds a cholesteatoma, suppress infection, and penetrates a few into the centre. Despite aggressive antibiotic treatment, otorrhea is persisting or repeats (4,19,20).

Otoscopic appearance is frequently typical, with marginal tympanic perforation and specific location (upper and posterior in Schrapnell area). After long evolution, chronic erosion occurs attic wall at the level of osicular chain, with the opportunity to observe cholesteatoma on the otoscopy (Figure 3B). Through transparency of the tympanic membrane it can be observed a possible cholesteatomatous bag that fills the middle ear (Figure 3B). If otorrhea persists a long time, may occur polyp formation that stretching along or even cover edge of the tympanic perforation (a bag cholesteatomatous) (Figure 3C).

Hearing loss is also, a common symptom (60-87%) (11), which can be progressive conductive or sensorineural: by the occupation of the middle ear from cholesteatomatous process to which are added the infectious process and causing the lysis of osicular chain. Conductive hearing loss is due to the impaired movement of ossicles, and further damage to the cochlea can cause irreparable sensorineural hearing loss and the development of deafness.
Vertigo and facial nerve paralysis are symptoms relatively less frequent, but these can occur if bone erosion produce a labyrinthine fistula, is developed to the bottom of the ladder or occurs lysis facial canal. Occasionally, the patient may present symptoms of CNS complications: sigmoid sinus thrombosis, epidural abscess or meningitis (21).

FINDINGS AND PROCEDURE DETAILS

Aggressive characteristics of cholesteatoma justify a careful approach by clinicians, even when symptoms appear to be mild. If cholesteatoma is detected early and is not stretched can be used surgical methods less invasive, it can preserve hearing (important especially in children) and it can avoid complications that endanger the patient’s life. The temporal bone anatomy is complex, by its small size, three-dimensional orientation of structures, by existing nerves and vessels.

Cholesteatoma diagnosis is made based on otoscopic examination and clinic but confirmation is given by sectional exams type CT or MRI.

CT scanning is able to make a perfect description of the anatomy of the middle ear and mastoid space, showing the relationship of the lesion with vessels, facial nerve and inner ear, it can discover very small lesions (10) as well the lithic effect on temporal bone. CT examination is specific, it is used high resolution protocol (CTHR), fine axial sections of 1 mm or inframetric with coronal reconstructions, sagittal, axial, oblique, allowing the view of bone chain entirely, with the possibility of producing 3D reconstructions for bone chain. The analysis includes data about middle ear cavities, about its content represented by bone lesion chain and cholesteatomatous lesion. In 50-97% of cases the presence of erosions is confirmed intraoperatively, but may lack to CC or in very small lesions CA. Examination with iv contrast show, the absence of contrast intake or marginal enhancement, not always well visible, (especially if the lesion is small), exclude neighbourhood complications. However, in larger cholesteatomas, the absence of central contrast enhancements is a useful sign for the differential diagnosis (17). If the symptoms and otoscopic exam are conclusive in diagnosis of cholesteatoma, CT is the first and only preoperative imaging exam. CT is a quick examination that be able to demonstrate ossicular and epitympanic erosion and will show tegmen delineation. Facial nerve canal and lateral semicircular canal integrity can be evaluated as well. CT can exclude the presence of complications (in neighbourhood or at distance) (9,11,22,23).

CT is most useful in preoperative diagnosis and when the middle ear is pneumatized. CT is non-specific if the middle ear is occupied by tissue content with various origins. CT cannot determine the location and extent of residual lesion after surgery excision or the presence of relapses.

If in the middle ear cavity is found content with different densities examination magnetic resonance imaging (MRI) can provide further information characterizing tissue damage found at HRCT examination, confirm the presence of cholesteatoma, its extension, gives important relationships about local architecture and postoperative give information about the presence of a residue or the occurrence of relapses (2,4,24-27). Classic sequences used are T1,T2-weighted sequences where cholesteatoma have a signal close to the cerebellum (hypo signal T1, moderate hyper signal T2), round or oval form, can be edging by a hyper signal in T2 (Figure 5A, B).

At these sequences is adde the late T1 sequence after contrast administration iv (to 45-60 minutes), (Figure 5C) which highlights the absence of contrast from the avascular cholesteatoma in opposition to uploading granulation tissue and inflammatory marginal tissue.

In the last few years increasingly using more new sequence type diffusion (echo-planar images EPI-DWI - where appears a hypersignal (Figure 5D) due to restriction in water diffusion, possibly by oily consistency on the part cystic given by keratin (17,28-30). In the study of Vercruiysse has been demonstrated that the sequence has a high sensitivity of 81% and a specificity of 100% in non-operated colesteatoma detection (lowest sensitivity is due to lesions small then 5 mmm or wall lesion) (31).

False positive results can occur in acute ear infections, scar tissue, granulation tissue, bone powder, cholesterol granuloma (31). The sequence has limitations in detecting residual lesions or relapses at tegmen timpanii level by magnetic susceptibility artefacts appeared at the air/bone interface (Figure 5D), motion artefacts given by pulsation vessels and thick sections and low resolution can hide an injury (32).
Greater specificity is achieved by using sequences DW-non EPI or DWI-PROPPELLER that can detect lesions up to 2 mm, with high sensitivity and specificity (Figure 5E), important in detecting residual lesions after surgery or in small dimensions recurrences. It is a sequence less sensitive to magnetic susceptibility artefacts, with thinner section thicknesses, higher spatial resolution (33).

DW-non EPI is a longer sequence which successfully replaced T1 sequence postcontrast delayed, also help to avoid unnecessary second-look operation at 6-18 months after surgery (31-33). Follow relapses are made up to 2 years in agreement with the surgeon.

Special sequences of diffusion have sensitivity and high specificity for residual or recurrent lesions and are used for their detection, but does not offer anatomic landmarks of the temporal bone and this sequence was a drawback. De Foer et al. showed the value of classic MRI for the assessment of possible complications such as erosion of the lateral semicircular canal, invasion of the labyrinth, and invasion of the middle cranial fossa through an eroded tegument, which is why an examination is made complete with classical and diffusion sequences (2,32-34).

CONCLUSION

First intention on imaging examination in the diagnosis of cholesteatoma remains CT with thin sections and high resolution; it makes the diagnosis, appreciate the extension and complications.

In cases of middle ear opaque, preoperative MR exam brings new information confirming the presence and location of cholesteatoma appreciates presence of complications.

Postoperative MR examination (using new sequences type DW-EPI and DW-non-EPI) is important for assessing the residual lesions or in the detection of relapses with time tracking up to 2-3 years especially in children to reduce their exposure to ionizing radiation (35), replacing second look surgeries.

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REFERENCES

5. Robinson JM – Cholesteatoma: skin in the wrong place. JRSM 1997; 90:93-96
8. Semaan MT, Meguerian CA – The pathophysiology of cholesteatoma.
13. Tos M, Lau T – Late results of surgery in different cholesteatoma types. ORL 1989;51:33-49
20. Caponetti G, Lester D, Thompson R – Pathology Clinic Cholesteatoma. ENT 2009;234:3-6

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28. Fernández Taranilla M, Herrera Herrera R, Moreno de la Presa R – Magnetic Resonance Imaging (MRI) and High Resolution Computed Tomography (HRCT): Can they improve the evaluation of Middle ear cholesteatoma. EPOS – ECR 2013:C-1249


33. Takahashi H – Cholesteatoma and Ear Surgery, Amsterdam 2013© Kugler Publications
