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REVIEW

Imaging of non-operated cholesteatoma: Clinical practice guidelines

D. Ayache^{a,*}, V. Darrouzet^b, F. Dubrulle^c, C. Vincent^d, S. Bobin^e,
 M. Williams^f, C. Martin^g

^a Service d'ORL et de chirurgie cervicofaciale, fondation A.-de-Rothschild, 29, rue Manin, 75019 Paris, France

^b Service d'ORL, de chirurgie cervicofaciale et d'ORL pédiatrique, université de Bordeaux Segalen, hôpital Pellegrin, CHU de Bordeaux, place Amélie-Raba-Léon, 33076 Bordeaux cedex, France

^c Service d'imagerie médicale, hôpital Huriez, CHRU de Lille, rue Michel-Polonovski, 59037 Lille cedex, France

^d Service d'otologie et otoneurologie, hôpital Roger-Salengro, CHRU de Lille, 59037 Lille cedex, France

^e Service d'ORL et de chirurgie cervicofaciale, CHU de Kremlin Bicêtre, 78, rue du Général-Leclerc, 94275 Le-Kremlin-Bicêtre cedex, France

^f Service d'imagerie médicale, fondation A.-de-Rothschild, 29, rue Manin, 75019 Paris, France

^g Service d'ORL et de chirurgie cervicofaciale, CHU de Saint-Étienne, 42055 Saint-Étienne cedex 2, France

KEYWORDS

Ear;
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 Imaging;
 CT scan;
 MRI

Summary Middle ear cholesteatoma is an aggressive form of chronic otitis media requiring surgical therapy. The surgical strategy depends on the location of the lesion, its extensions to the middle ear and mastoid, the anatomical conformation of the tympanomastoid cavities and the health status of the patient (as well as his or her interest in aquatic leisure activities). For several years, imaging of the ear has been a routine test in the preoperative work-up of the disease. National guidelines for the topic "Imaging of non-operated middle ear cholesteatoma" were prepared in October 2010, for the annual congress of the French Society of Otolaryngology Head and Neck Surgery (SFORL), by a panel of experts from the SFORL, represented by the French Association of Otolology and Neuro-otology (AFON), and the French Radiological Society (SFR), represented by the French Society of Head and Neck Imaging (CIREOL). These guidelines are presented in the present article.

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Clinical situation

Middle ear cholesteatoma is a form of chronic otitis media considered dangerous due to the risk of evolution of potentially severe complications, making surgical management mandatory [1]. It is usually defined by the development of a

malpighian epithelium within the middle ear and/or mastoid cavities.

Modern imaging now plays a key role in pre- and post-operative management of middle ear cholesteatoma [1–4].

Objectives and indications for imaging

A review of the literature concerning the role of computerized tomography (CT) in the pre-operative workout for cholesteatoma was conducted according to the GRADE Work-

* Corresponding author. Tel.: +33 1 48 03 68 40;

fax: +33 1 48 03 65 17.

E-mail address: dayache@fo-rothschild.fr (D. Ayache).

ing Group guidelines [5] (strong professional agreement). No articles with “high or moderate” levels of evidence could be retrieved: there were no prospective comparative large-cohort studies; moreover, many articles dating back 10 to 15 years could be considered obsolete in light of the technological progress of recent years.

Most recent reports recommend a CT scan as part of the pre-operative work-up in middle ear cholesteatoma [1,3,4,6–11]. While each individual study had only low or very low levels of evidence on the usual classification systems, their convergence confers a much higher level of evidence on the item “role of routine CT scan in the pre-operative work-up in middle ear cholesteatoma”. Moreover, the working group experts considered that temporal bone CT without contrast enhancement should be part of the systematic pre-operative work-up in middle ear cholesteatoma (strong professional agreement).

Pre-operative imaging assessment is systematically recommended to determine cholesteatoma extension, screen for complications, assess tympanomastoid cavity anatomy (notably for variants entailing surgical risk), and confirm the diagnosis in those rare cases in which otoscopy proves inconclusive [1,4,6,8–11].

Which imaging technique?

In the pre-operative work-up in middle ear cholesteatoma, CT scan without contrast enhancement is the first choice imaging modality (strong professional agreement).

Nuclear magnetic resonance imaging (MRI) may sometimes be useful (professional agreement) to supplement CT data in certain limited indications (see below).

How to formulate the request for imaging?

The clinical context should be summarized (cholesteatoma is usually diagnosed by an ENT physician on otoscopic examination), detailing the main requirements of the surgeon (extension, complications, anatomy).

How to perform the examination?

CT is recommended as part of the initial work-up in middle ear cholesteatoma. A high-resolution CT scan (HRCT) is performed with helical acquisition, inframillimetric slices and no contrast injection (professional agreement). Axial and coronal images should be bilateral, to allow comparative analysis.

Radiological semiology

Diagnosis of cholesteatoma is usually based on otoscopy rather than imaging, which, however, may provide confirmation in atypical presentations and is essential for diagnosing cholesteatoma developed behind a closed tympanic membrane (low level of evidence).

The two cardinal signs on CT are a classically nodular tissular mass, usually associated with neighboring areas of osteolysis (which witness complications, detailed below).

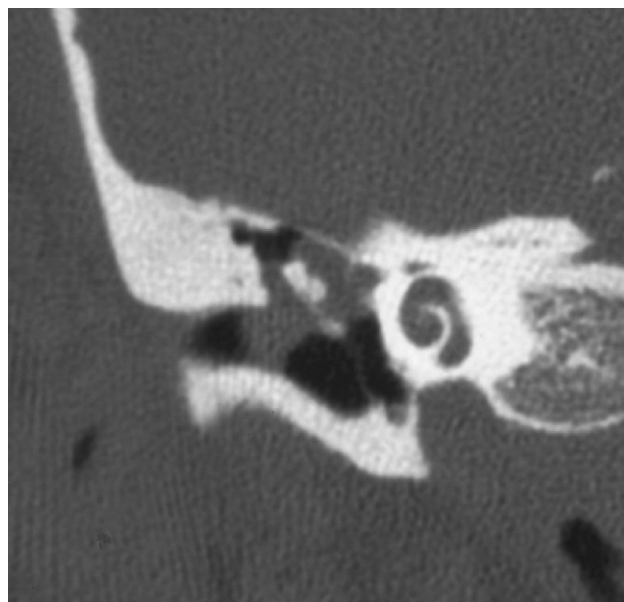


Figure 1 Right ear coronal CT scan: external and internal epitympanic tissue mass, presenting as nodular opacity with margins convexity, surrounding ossicles and causing erosion of the distal part of the epitympanic wall, highly suggestive of cholesteatoma.

The mass in the tympanomastoid cavity is homogeneous and non-calcified, with tissue-like density (Fig. 1). It is usually nodular, with a rounded, polycyclic or crenated convex contour. The contour of the opaque area is visible only where there is persistent perilesional aeration, and thus cannot be analyzed when the tympanomastoid cavity is completely filled by the mass. The lesion, especially when it extends to the external attic, may exert a mass effect, medially displacing the ossicular chain by pressure on the malleus head and incus body (Fig. 2). Exceptionally, this tissular mass



Figure 2 Left ear coronal CT scan, in a patient with epitympanic cholesteatoma; nodular opacity with tissue-like density, inducing erosion of the distal part of the external epitympanic wall and exerting a mass effect on the malleus, forcing it inward.

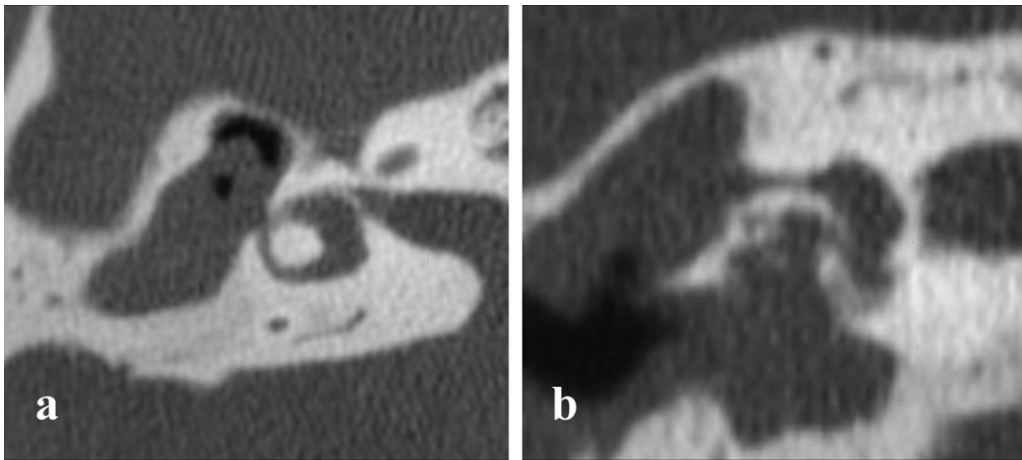


Figure 3 Right ear CT scan: lateral semicircular canal fistula (a: axial view; b: coronal view).

may be absent, if it has been removed by the ENT physician just before the CT scan.

CT is often able to determine extension [low level of evidence], especially when tympanomastoid cavity is not completely filled and still partially aerated. Analysis is more difficult when middle-ear cavity filling is diffuse, as CT fails to distinguish reliably within the tissular opacity between the cholesteatoma and the frequently associated neighboring inflammatory reaction [8,12–14] (low level of evidence).

CT is the examination of choice for identifying areas of osteolysis and screening for the main complications associated with cholesteatoma (strong professional agreement):

- erosion of the external wall of the epitympanum. This is an early sign frequently found in cholesteatomatous pathology (Figs. 1 and 2). Likewise, mastoid cholesteatoma leads to disappearance of mastoid cell trabeculation (low level of evidence);
- ossicular chain damage, ranging from partial lysis (especially of the long process of the incus) to disappearance of all ossicular structures (low level of evidence). Ossicular

chain damage is not specific to cholesteatoma, and can be found in other forms of chronic otitis media;

- labyrinthine structure damage, mainly involving the lateral semicircular canal or, more rarely, the posterior or anterior canal or cochlea (Figs. 3 and 4). MRI can supplement CT data when intralabyrinthine extension is suspected, revealing obliteration of labyrinthine fluid on T2-weighted inframillimetric slices facing the bony dehiscence, by an unenhanced tissue on post-contrast T1-weighted images [15] [low level of evidence];
- erosion of the facial canal along its pathway through the temporal bone (Fig. 5) –although radiologic analysis, especially of the tympanic segment of the canal, may be difficult to interpret [7,8,12] (low level of evidence), in which case particular caution is advisable in case of labyrinthine fistula, where association with facial canal dehiscence is known to be significantly more frequent [10,16] (low level of evidence);
- erosion of the tegmen or bony capsule of the sigmoid sinus (Fig. 6). CT assessment should be supplemented by MRI when meningocephalic infection, intracranial extension or sigmoid sinus thrombosis is suspected [17] [very low level of evidence]. Depending on the clinical presentation, venous angio-MRI or venous angio-CT may be

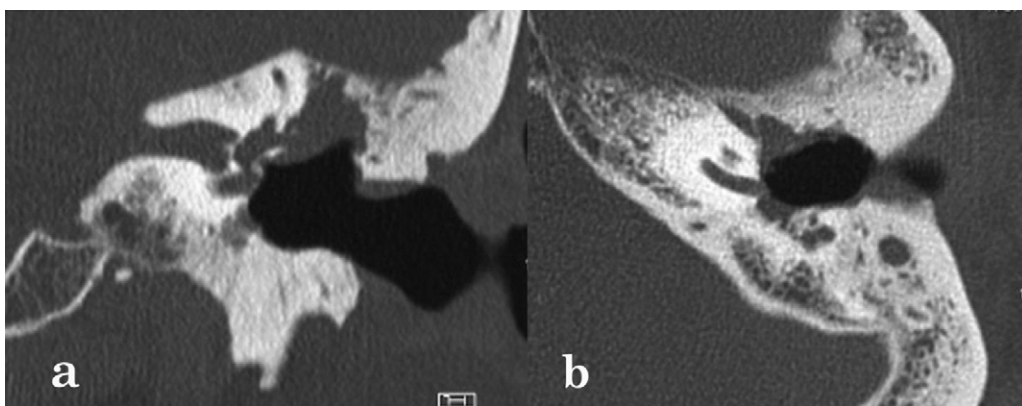


Figure 4 Left ear CT scan: fistula of the basal turn of the cochlea (a: coronal view, also revealing a lateral semicircular canal fistula, with non-visualization of the tympanic segment of the bony facial canal; b: axial view).

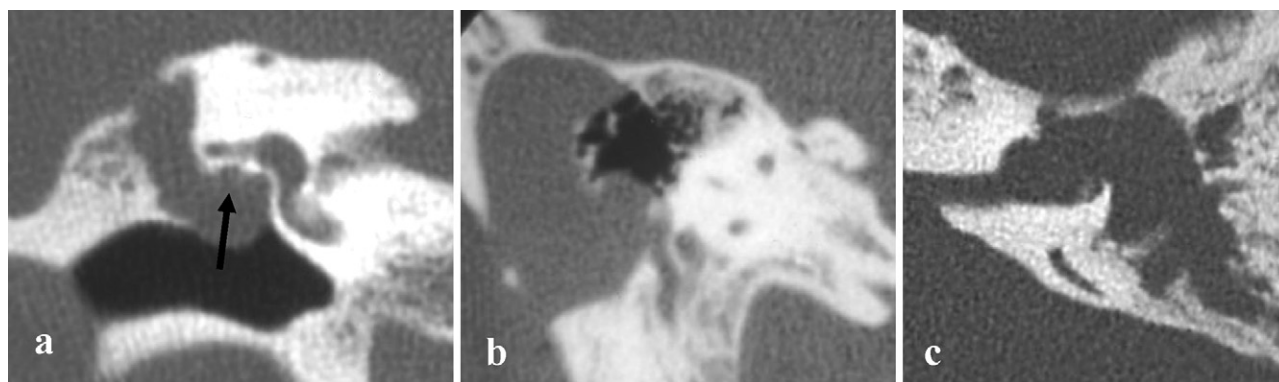


Figure 5 Various types of facial canal involvement in cholesteatoma (a: tympanic segment; b: mastoid segment; c: labyrinthine segment).

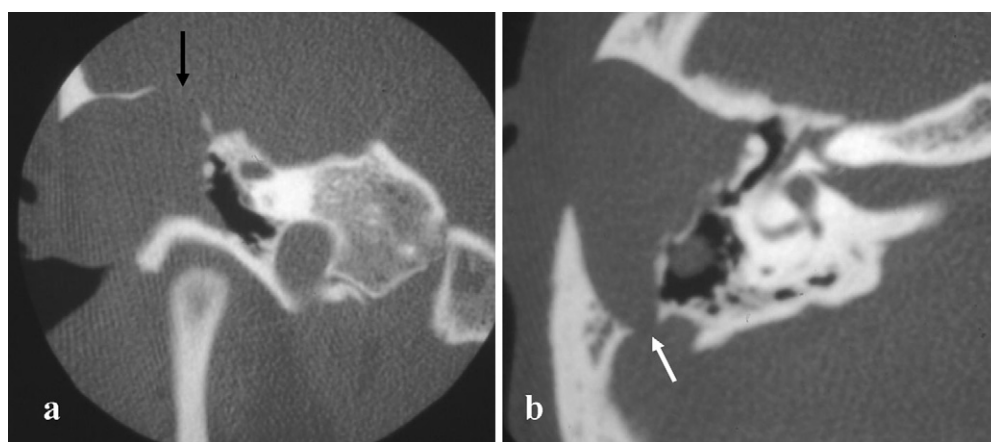


Figure 6 Iatrogenic right-ear cholesteatoma: a. coronal view, showing tegmen erosion (black arrow); b. erosion of bony capsule of the sigmoid sinus on axial view (white arrow).

used to detect sigmoid sinus thrombosis. Cerebral abscess facing tegmen erosion may be detected not only by classic sequences (T1-weighted with and without gadolinium enhancement, FLAIR, T2) but also by a diffusion-weighted sequence (which, in doubtful cases, can distinguish a cerebral abscess from a necrotic tumor).

CT is also very useful for determining tympanomastoid cavity anatomy. Mastoid volume and pneumatization may directly influence the choice of surgical technique [1,4,8,9,18] (low level of evidence). Finally, certain anatomic variants should be systematically explored for and screened in pre-operative imaging, as they may incur additional surgical risks: superficial or prolapsed sigmoid sinus, prolapsed tegmen, prolapse of the tympanic segment of the facial canal or variant trajectory of the mastoid segment, and prolapse and/or dehiscence of the bony capsule of the jugular bulb or intrapetrous carotid [3,6,19] (low level of evidence).

Conclusion

Following pre-operative work-up, clinical, audiometric and imaging data enable the otologist to confirm diagnosis of cholesteatoma, screen for the main complications and plan

surgery according to lesion extensions and the patient's anatomy.

Summary guidelines

Diagnosis or suspicion of middle ear cholesteatoma on clinical examination requires imaging assessment.

The first-line imaging modality is CT scan without contrast injection.

It allows the clinician:

- to confirm diagnosis;
- to explore for osteitic complications (bony labyrinth, facial canal, tegmen, sigmoid sinus, ossicular chain);
- to determine extension;
- and to assess middle-ear and mastoid cavity anatomy.

According to the clinical and CT presentation, assessment may be supplemented by MRI or angio-MRI: large labyrinthine fistula, tegmen erosion with suspected meningocele or intracranial extension, or neuromeningeal complications.

Imaging, and especially CT scan without contrast injection, is part of the systematic pre-operative work-up for middle ear cholesteatoma and contributes directly to drawing up the treatment strategy.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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