

Cholesteatomas in Children

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Key Words: Cholesteatoma- congenital cholesteatoma- acquired cholesteatoma- canal wall up – canal wall down – recidivism - endoscope - matrix – granulation tissue.

Introduction

The term cholesteatoma is itself a misnomer but continues to be used through a sense of tradition. Defining what is a cholesteatoma is a subject of debate. However, a working definition would describe it as a 3-dimensional connective tissue sac lined by stratified keratinizing squamous epithelium, which often grows at the expense of underlying structures (de Souza et al, 1989). It also has a tendency to regrow (Castle 2018) if a small piece of cholesteatoma is left behind. This is known as recidivism. Pediatric cholesteatomas in terms of gross histopathology resemble cholesteatomas in adults. However, in terms of behavior they differ considerably. There are 2 types of cholesteatomas, congenital cholesteatomas and acquired cholesteatomas. Cholesteatomas in children are thought to be aggressive and very destructive. Cholesteatomas in the pediatric population are also much more associated with intracranial and extracranial complications than cholesteatomas in adults.

Surgery for the treatment of cholesteatomas in patients who suffer from it is quite consequential, especially for children as it will affect their hearing.

HISTOPATHOLOGY OF CHOLESTEATOMA

Cholesteatoma is made up of three components: the cystic content, the matrix (often called the capsule of the cholesteatoma), and the perimatrix (also known as the cholesteatoma stroma) (Ferlito et al, 1997). Cholesteatoma consists of dead, fully differentiated anucleate keratin squamas. This is the outer corneal layer of the squamous cell epithelium. The matrix is composed of fully differentiated squamous epithelium resting on connective tissue called the perimatrix. There is a basal layer of small cuboidal cells above which is a malpighian layer composed of five or six rows of cells which are, from bottom to top: the stratum basale, also known as the stratum germinativum; the malpighian or spinous layer; the granular layer; stratum lucidum; and the keratin layer, with intercellular prickles. The deeper layers of the epithelium of the cholesteatoma matrix frequently show evidence of activity in the form of down-growths into the underlying connective tissue. These often separate into cholesteatoma lobules, thus suggesting vigorous extension and expansion. A thin granular layer lies between the malpighian layer and the corneal layer (Michaels, 1989). This histological picture is seen in all cholesteatomas. However, Frankel, Berson, Han, and Parisier (1993) have found that dendritic (Langerhans) cells are more profuse in an acquired cholesteatoma. They postulate that acquired cholesteatomas arise in an antigenically active environment, thus there are more dendritic cells. Congenital cholesteatomas arise in a relatively quiet antigenic environment and there are fewer dendritic cells. Quaranta, Resta, and Santangelo (1986) have found that the perimatrix of cholesteatomas found in children is richer in mononuclear inflammatory elements, with evidence of enzyme collagenase activity.

Is there any difference between the stratified keratinizing squamous epithelium of cholesteatoma, the squamous epithelium of the tympanic membrane, and the EAC? At this time this question cannot be answered with certainty. But current research does show that great similarities found in tissue cultures exist between them (Cody, 1977; Vennix, Kuijpers, Peters, Tonnaer, & Ramaekers, 1996). Michaels, (1989) finds these similarities to be indicative that cholesteatomas arise from the epithelium of the external canal skin. This is particularly pertinent for acquired cholesteatomas, where it is postulated that an abnormality in the migratory pattern of epithelium from the external canal results in epithelium migrating into the middle ear, resulting in cholesteatoma (Palva, Karma, & Makinen, 1982). In terms of histopathology there are no differences between cholesteatomas found in children and those found in adults (Damberg et al 2024).

Cholesteatomas are classified as congenital and acquired. Acquired cholesteatomas are further classified as primary, secondary, and tertiary.

Ghinst et al (2012) examined the expression of Galectin-1-3-7 in congenital and acquired pediatric cholesteatomas vis a vis that of external auditory canal skin. They found there was no difference in the galectin distribution pattern between congenital and acquired pediatric cholesteatomas.

Damberg et al (2021) in their study described the appearance and distribution of tissue remodeling markers like MMP-2, MMP-3, TIMP 2, TIMP-4, sonic hedgehog gene protein (Shh) pro and anti-inflammatory cytokines (IL-1, IL-10), factor (NFkB), proliferation marker (Ki-67) angiogenetic factor (VEGF), tissue defensins (HB D-2, HBD-4) in pediatric cholesteatoma. They note that the persistent increase in the Shh gene protein expression found in the perimatrix of pediatric cholesteatoma was responsible for the stimulation of growth of pediatric cholesteatoma. Palko et al (2018) found that the expression of KRT1 and KRT10 cytokeratin genes was higher in children as compared to adults. Chen and Qin (2011), Kan et al (2021) examined the association of matrix metalloproteinase-2(MMP-2) mRNA expression with subtypes of pediatric cholesteatoma. MMP-2 was thought to play an important role in the progression of cholesteatoma by causing and promoting bone destruction and keratinocyte infiltration. While they found no significant difference between congenital or acquired cholesteatoma in children, they noted that the level of expression of MMP-2 was related to the pathogenesis and aggressiveness of cholesteatomas in children.

Asher et al (2015) noted that subepithelial angiogenesis within the perimatrix of cholesteatomas in children was far more pronounced than that in adults contributing to the aggressiveness found in pediatric cholesteatomas.

Do genetics play a role in cholesteatoma? Jennings et al (2018) in their study hypothesize that congenital and acquired cholesteatomas have been demonstrated to be present within families in the

pattern typical of a monogenic or oligogenic disorder with incomplete penetrance. Evidence from syndromic cases suggest that genes controlling ear morphology may be risk factors for cholesteatoma formation. A small body of literature provides evidence of a heritable component for its etiology. Lee et al (2023): hypothesize that the candidate genes that they identified may be part of the key signaling pathways during mucosal response to middle ear infection. The occurrence of multiple rare variants may play a role in earlier onset of cholesteatoma formation in chronic otitis media. Franck et al (2022) postulate that the microbiota associated with cholesteatoma and chronic suppurative otitis media could be instrumental in the cause of cholesteatoma.

CONGENITAL CHOLESTEATOMA

Cawthorne and Griffith (1961) and Cawthorne (1963) and are credited with describing congenital cholesteatoma, but it was Derlacki and Clemis (1965) who established criteria by which a cholesteatoma could be termed congenital. While most authors find the Derlacki and Clemis criterion useful, they disagree on the inclusion of the criterion that no attacks of otitis media should be present. Some authors (Levenson, Michaels, & Parisier, 1989) feel that this should be excluded, because otitis media is a very common and insidious problem in childhood. However, when otitis media is accompanied by otorrhea and perforation, then such a child should not be termed as suffering from a congenital cholesteatoma.

Embryology

Many theories have been presented regarding the formation of congenital cholesteatomas in the middle ear. We will discuss some of the more widely quoted and accepted of these.

Roth (1977) declares that congenital cholesteatoma is a disease of growth control. He states that ectodermal epithelium reaches a certain location and should receive a turnoff signal. Congenital cholesteatoma is a result of this signal failure. Aimi (1983) elaborates on this by describing the tympanic ring as a "stop" signal to the advancing ectoderm against its inherent potential to grow into the tympanic isthmus. Aimi (1983) notes that a majority of cholesteatomas occur near the tympanic isthmus of the middle ear. Since this is the junction between the 1st and 2nd branchial arch, the origin of congenital cholesteatoma could be linked to the migration of the external canal ectoderm into the middle ear at the early stage of development.

The tympanic ring plays an important role in limiting the medial extent of the external canal to the level of the tympanic annulus. Migration of the ectodermal tissue into the middle ear beyond the annulus is likely to be caused by the failure of this inhibitory function of the tympanic ring. The anatomical distance in the fetus between the tympanic ring and the internal acoustic meatus is extremely short. Petrous cholesteatomas could thus also be the result of failure of the inhibitory function of the tympanic ring.

Thus, congenital cholesteatomas are caused by the migration of external canal tissue into the tympanic isthmus, or into the petrous bone as a result of a developmental error in which advancing external canal ectoderm failed to receive the stop signal of the tympanic ring. This theory explains congenital cholesteatomas occurring in the posterior superior quadrant of the middle ear.

In another theory, Michaels (1986) found a squamous cell crest in the anterior superolateral wall of the tympanic cavity. This could be identified from 10 to 33 weeks of gestation. This structure has been termed "epidermoid formation" and is situated just anterior to the developing tympanic membrane. This epidermoid formation is present at a site where histological differences occur. Simple cuboidal epithelium lies just posterior to the epidermoid formation, while tall pseudostratified ciliated epithelium leading to the Eustachian tube lies anterior to it. The epidermoid formation is thus situated in an epithelial transformation point from the tympanic cavity to the Eustachian tube. After 33 weeks of gestation, the epidermoid formation is no longer present. The function of this formation is to act as an "organizer" for the development of the middle-ear cleft. Once its function of organizer is over, the epidermoid formation is expected to do so. It has been thought that its failure to do so would result in a congenital cholesteatoma. This would be most applicable in situations where the cholesteatoma is situated anterior to the handle of the malleus (Cohen, 1987).

The metaplastic theory has been proposed by Tumarkin (1938) and by Sade, BaBiacki, and Pinkus (1983) to explain the formation of cholesteatoma behind an intact tympanic membrane. They have offered convincing evidence that the middle ear mucosa undergoes transformation to stratified squamous epithelium in response to chronic inflammation.

However, in our studies of temporal bones in cats, chinchillas and human temporal bones we have described multiple epithelial changes during otitis media and we have never observed changes conducting to cholesteatoma formation. In all our cases the origin of cholesteatoma has been seemingly by squamous epithelial migration (Goycoolea 1999). This stratified squamous epithelium in turn leads to keratin formation. Paparella and Rybak (1978) theorize that congenital cholesteatomas could arise from ectodermal implants in the fusion planes of the first and second branchial arches. Maccarrone et al (2022) postulate that the tensor tympani is a possible site from which a congenital cholesteatoma occurs. They base this observation on the intraoperative observations that the congenital cholesteatoma sac arose from the tensor tympani tendon.

Signs and Symptoms of Congenital Cholesteatoma

As the patients afflicted are very young, there are usually very few signs and symptoms, since most patients are unable to articulate their problems. Some are discovered accidentally (Schwartz, Groundfast, & Feldman, 1984). Children suffering from craniofacial anomalies should be examined to determine if congenital cholesteatoma is present (Parisier et al., 1989). Awareness of these conditions will lead to a high index of suspicion. Congenital cholesteatoma may occur in an atretic ear, and these children are usually asymptomatic unless the lesion becomes secondarily infected (Sie, 1996). Schwartz et al (1984) found a significant number of congenital cholesteatomas associated with a middle ear effusion. Small epithelial pearls may be found within the layers of the tympanic membrane; that is, cholesteatoma within the layers of the tympanic membrane (Sobol et al, 1980).

Pneumatic otoscopy in experienced hands was often the first tool that led to the discovery of congenital cholesteatoma (Levenson, Parisier, Chute, Weing, & Juarber, 1986). CT scanning then provided documentation of the lesion along with other data (Arriaga 1994). Furthermore, CT scanning is a sensitive evaluation for the extent of middle ear, ossicular, epitympanic, and mastoid involvement by cholesteatoma (Bellet, Benton, Matt & Myers 1992; Liu & Bergeron, 1989).

Pneumatic otoscopy usually reveals a white mass behind the tympanic membrane. If the congenital cholesteatoma was located anterior to the handle of the malleus, a mild conductive hearing loss or normal hearing might be present. However, if the mass was detected in the posterior segment, then ossicular disruption was likely to be present, causing a definite conductive hearing loss. If the cholesteatoma was located in the petrous apex, then a SNHL was likely. Serous otitis media was also found to be simultaneously present. Bilateral congenital cholesteatomas are rare occurrences (Braganza & Kearns, 1993; Peron & Schuknecht, 1975).

Evaluation of a Patient suspected to have Congenital Cholesteatoma

Pneumatic otoscopy followed by ear microscopy are the most important tools in beginning an assessment. In experienced hands, they provide important clues to the pathology occurring in the ear. Audiological assessment includes pure tone audiometry as well as impedance audiometry for establishing hearing thresholds in older children. Evoked response audiometry is used to determine hearing thresholds in younger children. This will indicate if any damage has been caused to the conductive and/or neural mechanism of the ear.

High-resolution CT scanning of the temporal bone is the imaging modality of choice. CT scanning will delineate the extent of the disease and also demonstrate the anatomy of surrounding structures. CT Scanning will demonstrate problems like congenital aural atresia and / or stenosis of the external auditory canal (Kalmanson Francom, Darr and Hamilton 2023). This is important, since the dictum that where one anomaly is present, look for another, is particularly true (Bois, Nassar et al 2018) , (Mierzwinski et al 2018), (Vannesta, Page 2019) (Spinner A, Munjuluru A, Woolten C 2020). Thus, if another anomaly is present in the area scanned, it will also be well demonstrated. Furthermore, CT scanning will help determine if the lesion is indeed a cholesteatoma, or some other pathology.

Treatment of Congenital Cholesteatoma

Surgical removal is the best way to deal with this problem. When the lesion is located in the anterior quadrant, it has been found that the lesion is well encapsulated. This facilitates relatively easy total removal. However, cholesteatomas located in the posterior quadrant, by virtue of draping themselves on various structures, are more difficult to totally remove. Furthermore, ossicular destruction will also have occurred, necessitating ossicular reconstruction. In general, congenital cholesteatomas located and confirmed to be located anterior to the handle of the malleus are usually removed in a one-stage procedure, usually in the form of a tympanotomy, while congenital cholesteatomas located in the posterior quadrant are likely to need two-stage procedures (Sanna & Zinni, 1984), which might also involve the need to perform a mastoidectomy as well as a tympanotomy. The ossicular reconstruction is usually at a second stage.

Summary

- Congenital cholesteatoma typically presents as an expanding cystic mass with keratinizing squamous epithelium situated medial to the intact tympanic membrane,
- Congenital cholesteatoma is thought to be present at birth. However, it is commonly diagnosed during infancy or early childhood in patients with no prior history of otorrhea. In order for it to be termed 'congenital' there should be no history of perforation, or previous ear surgery.
- A history of previous bouts of otitis media or an effusion does not exclude the diagnosis of congenital cholesteatoma.
- The commonest site of congenital cholesteatoma is frequently at the anterosuperior quadrant of the middle ear.
- The clinical presentation of congenital cholesteatoma is determined by its location and extent.
- Congenital cholesteatomas present as a pearly white mass behind an intact tympanic membrane,
- Congenital cholesteatomas can present with a hearing loss (usually conductive hearing loss) when it enlarges to fill the middle ear or by eroding the ossicles,
- Children are usually not able to clearly articulate their symptoms.
- CT Scanning is the best radiological imaging tool to identify the dimensions of the lesion as well as determine bone erosion / destruction. MRI is also a useful radiological imaging tool to visualize soft tissue involvement as well as identify intracranial / extracranial involvement

ACQUIRED

CHOLESTEATOMAS

Etiology

Acquired cholesteatomas have been classified as primary, secondary or tertiary. Various theories have been postulated to explain the occurrence of these lesions. Primary acquired cholesteatomas are cholesteatomas that are situated in the attic and are the result of attic retractions caused by eustachian tube dysfunction (Sculerati & Bluestone, 1989). These retractions result in accumulation of debris, which may result in infection. This may cause ulceration with perforation, thus allowing the epithelium to grow into the attic. Some authors (Bartos, Urik, Buresova, Holochova P, Budinska and Linhartova 2025) postulate that retraction pockets could likely be a precursor (precholesteatoma) stage to the formation of a cholesteatoma.

Secondary acquired cholesteatomas are cholesteatomas that are found in the presence of a perforation, in the pars tensa or a retraction pocket in the pars tensa. In the case of perforations, it is thought that stratified keratinizing squamous epithelium migrates into the middle ear via the perforated edges of the tympanic membrane. In the case of a retraction pocket, it is thought that one of the following occurs:

- (1) Either the retraction progresses to such a stage that the epithelial debris can no longer have egress into the external canal, with consequent build-up of stratified keratinizing squamous epithelium and its debris and expansion into the middle ear, and the mastoid aditus and antrum.

(2) Or it is also thought that a retraction pocket could lead to collection debris, which may result in an infection. This infection results in an ulcer in the retraction pocket, leading to perforation. The epithelium is then permitted to migrate into the middle ear and mastoid.

Tertiary acquired cholesteatomas are the result of stratified keratinizing squamous epithelium being implanted into the middle ear or mastoid. This kind of cholesteatoma is associated with blast injuries, or following surgery on the ear, whereby the keratinizing stratified squamous epithelium is accidentally implanted into the middle ear. It is unclear at this time whether antimicrobial therapy and/or the placement of tympanostomy tubes in the management of otitis media has affected the incidence of cholesteatoma. (Padgham, Mills, & Christmas, 1989). Some children seem more susceptible to chronic otitis media and development of cholesteatoma, particularly children with cleft palates and those suffering from trisomy 21 (Dominguez & Harker, 1988; Pappas, Flexer, & Shackelford, 1994).

Surgery for cholesteatoma is divided into canal wall down and canal wall up strategies. Interestingly enough, both these strategies have common objectives as their goals. These objectives are a dry, disease-free ear and improvement in hearing. However, despite these common objectives, surgeons remain divided (sometimes strongly divided) on the technique to be used.

Signs and Symptoms

Children with acquired cholesteatomas are generally older than those with congenital cholesteatomas (Sie, 1996). Foul-smelling otorrhea, usually at first associated with upper respiratory tract infections, and later independently occurring, is the predominant common complaint. When a patient complains of foul-smelling otorrhea, the examiner must presume a cholesteatoma is present until proved otherwise (Edelstein, Parisier, & Han, 1989). Diminished hearing, otalgia, and upper respiratory tract infections are associated with this condition. Facial paralysis and profound SNHL can occur but fortunately are uncommon occurrences. Both these problems are usually associated with long-standing untreated or partially treated diseases. Aural polyps usually signify the presence of cholesteatoma (Gliklich, Cunningham, & Eavy, 1993).

Clinical examination is best done with the aid of an operating microscope. This permits a magnified view of the tympanic membrane, as well as cleaning of the ear. Debris obscuring critical areas can be removed to permit better visualization of the tympanic membrane. This helps in the better detection of cholesteatomas. Pneumatic otoscopy, although useful, does not achieve the same objectives as examination of the ear under an operating microscope.

Pure tone audiometry helps in documenting hearing thresholds and in most cases, is useful in determining if ossicular discontinuity has occurred. In older children, it is useful and necessary to obtain speech reception thresholds and speech discrimination scores. Racca JM et al (2022) have clearly demonstrated a clear association between cholesteatoma and progressive hearing loss in children. Additional analyses demonstrated that mastoidectomy surgeries did not appear to contribute to the progressive hearing loss. They also showed that a high number of children suffering from cholesteatoma had normal hearing thresholds at their first test.

CT scans of the temporal bones with high-resolution images and with thin sections in the axial and coronal planes, are useful in demonstrating the extent of disease (Figure 1) and surrounding anatomy. CT scans will thus demonstrate the presence of fistulas in the labyrinth and cochlea, the status of the facial nerve and the size of the mastoid, and the position of the sigmoid sinus and internal carotid artery. While CT scans help anticipate certain situations, especially where revision surgery is contemplated, they are always to be correlated with clinical findings. Only hearing ears are imaged as well. The role of MRI is used only if disease has gone beyond the boundaries of the temporal bone.

MANAGEMENT

Surgery is the treatment of choice. The type of surgery to be performed is the biggest area of disagreement.

Canal Wall Up (Intact Canal Wall)

Surgeons employing the canal wall up technique do so because they desire to preserve the normal anatomy of the ear, while attempting to totally remove disease and improve hearing (Sheehy, 1988). They also state that improvement of hearing is much more feasible with this technique. The prerequisites of this technique are resectable disease, posterior tympanotomy, good follow-up, and often a second look at a later date to determine that they have been successful in totally removing cholesteatoma. The rationale given by those who prefer canal wall up techniques is that cavities are difficult to maintain (Charachon, 1988), both for doctors and patient. Tos (1983) finds that the temporal bone continues to develop throughout childhood. An open cavity becomes larger and the meatus thus gets smaller, as it does not keep pace with the growth of the cavity; this makes cleaning of the cavity difficult. Smaller children often resist cleaning of the cavity, adding to the difficulty of maintenance of the cavity. Cavities necessitate regular visits to the office. Sometimes the patient exchanges one cause of otorrhea for another when a cavity is created. In addition, certain activities like swimming may be restricted when cavities are created.

The reformation (recidivism) of cholesteatoma from cholesteatomata remnants left behind is the single most important hazard (Mills & Padgham, 1991) that occurs when the canal wall is preserved.

Disagreement has occurred around this issue. Rates of reformation vary from 17% to 30%. Despite this, Glasscock, Dickens, & Wiet (1981) still prefer to perform canal wall up surgeries.

In an effort to detect residual and hidden cholesteatoma certain surgeons advocate the use of the endoscope (Thomassin, Korchia, & Duchon Doris, 1993) to detect the presence of residual cholesteatoma in the sinus tympani. As the latter is notoriously difficult to visualize with the operating microscope, the endoscope provides better visualization of these recesses and is thus able to remove cholesteatoma that may have escaped detection. However, even with this technique, cholesteatoma can still be left behind. In some situations, cholesteatoma, though visualized, is still left behind to be removed at the second stage. For example, when the cholesteatoma is found to envelop the stapes, and removal of cholesteatoma might result in a permanent SNHL even when using laser techniques, the cholesteatoma is left behind to be removed at a later date. It is thought that after a while, the cholesteatoma will encapsulate itself, facilitating complete and safe removal (Gyo, Sasaki, Hinohira, & Yanagihira, 1996).

Yamada et al (2024) evaluated temporal bone pneumatization after canal wall up tympanomastoidectomy for the removal of middle ear cholesteatoma in children and found that pneumatization still occurs. They use this finding to justify canal wall up procedures for removal of cholesteatoma in children.

Second Stage Procedures

Timing

Primary reconstruction is preferred for well-encapsulated cholesteatoma with minimal to moderate disease affecting the middle ear mucosa and the ossicular mechanism. When the disease is diffuse and its impact significant, staging is recommended.

When indicated, a second stage is usually done 6 to 12 months following the original surgery. This gap of time permits complete healing of the ear from the ravages of cholesteatoma, allows the middle ear space to redefine itself, and allows adequate ventilation of the middle ear. Mucosal changes in children are more reversible than in adults. Tubal dysfunction caused by thickened mucosa is more likely to spontaneously resolve than in adults. Thus, the chances of reventilation of the middle ear in children are higher (Tos 1983). This interval also gives the hidden cholesteatoma remnant time to grow and expose itself. When the cholesteatoma grows and encapsulates itself, complete and total removal is then possible (Charachon & Gratacap, 1985). The need for second-stage surgery in children is greater (Sheehy, 1985) than in adults, because children are more likely to have an invasive cholesteatoma and total removal may not always be possible. This is true especially when the cholesteatoma is acutely infected. The bleeding during surgery would make total removal difficult. Cholesteatomas in children are more likely to be associated with large, pneumatized mastoids. This makes for difficult removal,

especially if the cholesteatoma is invasive. At a second stage, the epithelial remnants grow and are more amenable to removal.

Hearing Reconstruction

This is usually done at the second stage for the following reasons. If the patient's hearing status has not improved following initial surgery, the patient is more likely to be motivated to undergo second-stage surgery. Following removal of the cholesteatoma and repair of the tympanic membrane, it is likely that the middle ear space will redefine its boundaries. This interval of time allows for stabilization of the new middle ear space and its mucosa. Thus, when reconstructing the ossicular mechanism, the ossicle can be tailored to meet the new boundaries of the middle ear space, as anatomic relationships therein shared have stabilized by this time.

Techniques Employed at the Second Stage

The traditional aural incisions are used and the mastoid and middle ear spaces are inspected for the presence of cholesteatoma. In recent times, authors such as McKennan (1993); Takahashi, Honjo and Fujita, (1990); Gonzalez and Bluestone, (1986); and Nomura (1982) advocate the use of endoscopes to inspect the mastoid and middle ear for the presence of cholesteatoma. The mastoid is inspected via a small postaural incision under local anesthesia and the middle ear is inspected via a myringotomy. If no cholesteatoma is found, the procedure is ended and the patient can be discharged the same day. If extensive cholesteatoma is found, the traditional aural incisions are taken, and the procedure is continued in a standard mastoid procedure. If an epithelial pearl is discovered there will be an attempt to remove it endoscopically. If this fails, the procedure is converted to a standard one employing the postaural incision.

Can High-resolution CT Scans Determine if Cholesteatoma is Present as an Alternative to Surgical Exploration?

High-resolution CT scans pick up minute changes (Johnson, 1984). Unfortunately, they cannot distinguish between cholesteatoma, granulations, or fibrous tissue. However, serial CT evaluation has been advocated by Edelstein and Parisier (1989) for children with possible residual disease as an attempt to avoid staged surgery. Serial CT scanning will demonstrate a growing lesion when CT scans done at various intervals are compared with one another. An expansile lesion is thought to be indicative of a growing cholesteatoma, and thus necessitates surgery. When combined with endoscopic evaluation of the middle ear and mastoid, CT scanning might have more value (Baudouin R et al 2022). This area is yet to prove itself to be superior to traditional surgical exploration (Singh et al 2020). However, in recent times Arendt et al (2021) report that high resolution CT scans of today as compared to previous older generation CT scanners are much improved and sophisticated and can possibly help differentiate between cholesteatoma and middle ear infections (Figure 2).

Problems with Second-stage Exploration

Cost containment is a formidable foe to staging any procedure and is a likely obstacle to be encountered. Patient noncompliance is another factor that occurs. Does the presence of cholesteatoma at the second stage necessitate a canal wall down procedure? This will depend on the circumstances, extent of cholesteatoma, compliance of the patient, and the experience of the surgeon. A more experienced surgeon will be able to determine what will be best for the patient in that circumstance. However, when in doubt it would be better to opt for canal wall down techniques. Gyo and colleagues (1996) note that when an epithelial pearl formation is noted at second stage, it can be safely removed without fear of reformation. But when a diffuse cholesteatoma is seen, it would indicate that a canal wall down procedure would be the best option. An open flat cholesteatoma is much more aggressive and would likely reform. Stern and Fazekas-May (1992) note that in their series, cholesteatoma located in the sinus tympani has usually been found to reform.

When Can the Ear Be Termed Disease (Cholesteatoma) Free?

This is an area of controversy. If cholesteatoma is found and the canal wall is still preserved at a second stage, does this necessitate a third look to determine if cholesteatoma has not reformed? This question is largely unanswered by current available literature. Some surgeons feel reformation is directly proportional to time. That is to say, the longer the ear is followed-up, the more likely it is for a cholesteatoma to be present (Arriaga, 1994). Lau and Tos (1987) use an arbitrary figure of 10 years from initial surgery to term an ear free from cholesteatoma if the ear was disease-free during that time. As this is a gray area, the criteria used to determine if an ear is free from disease are ill defined. As the second-stage surgery is done up to 12 months following initial surgery, another 9 years must pass before terming the ear free from cholesteatoma. To properly claim that the ear is indeed free from cholesteatoma, another surgical inspection should be done. However, most of the surgeons who do canal wall up techniques do not do this. Advocates of the canal wall down technique point to this failing as another reason why they advocate canal wall down techniques.

Are Cholesteatomas More Aggressive in Children?

Pediatric cholesteatomas are thought to be more aggressive than adult cholesteatomas (Lynrah et al 2013). Thus, aggressiveness is determined by the higher rates of reformation and the rapidity with which it reforms. Reformation of cholesteatoma following canal wall up surgery is also more likely to occur in a shorter span of time in children as compared to adults (Glasscock et al., 1981).

Are Cholesteatomas in Children Different From Those Found in Adults?

Histologically, they appear the same, but they differ in behavior. Most available literature demonstrates that cholesteatomas are more extensive in children than in adults because children's mastoids are more cellular and this permits the cholesteatoma to be more extensive. (Edelstein & Parisier, 1989; Jansen, 1978). However, the destructive nature of cholesteatomas is directly related to the duration of time the cholesteatoma is present (Sade & Fuchs, 1994; Vartiainen & Nuutinen, 1992). The longer a cholesteatoma is allowed to reside, the more destructive it becomes. Pediatric cholesteatomas are more aggressive in the pediatric population and are associated with a higher incidence of complications both intracranial as well as extracranial (Figure 3, Figure 4). They are also associated with a higher rate of recidivism. Another contributing factor is that pediatric cholesteatomas have a higher presence of granulation tissue which is thought to be the contributing factor to its destructiveness. (Lynrah, Bakshi, Panda, Khandelwal 2013). James (2024) states that the younger the age of the child who presents with cholesteatoma the more likely the risk of cholesteatoma recurring.

Eustachian Tube Function

Eustachian tube problems need to be simultaneously treated. Sheehy (1985) advocates removal of disease as the best way to help -the eustachian tube regain its function. If eustachian tube problems are not treated and altered, cholesteatoma will definitely recur with an intact canal wall technique (Smythe 1977).

Results of Canal Wall Up Techniques

Reformation of Cholesteatoma

Reformation of cholesteatoma is higher in children than in adults. Glasscock, Dickens, and Wiet (1981) quote a figure of 23% who had reformation of cholesteatoma. Of these 10 patients, 7 were suspected to have disease, while 3 had disease that was unsuspected. Although recurrent disease was also found in 23% of their patients, 7 (16%) of the patients developed recurrence after 6 to 12 months and 3 (7%) patients developed recurrence at 12 months.

Recurrent Cholesteatoma

Recurrent cholesteatoma means that the original cholesteatoma has been removed and a new cholesteatoma has occurred all over again, independent of the previous cholesteatoma. It becomes difficult at times to be completely sure that the new cholesteatoma is actually a fresh one. Vartianen and Nuutinen (1992) had a mean follow-up of 7.1 years; 17% of their patients had canal wall down surgeries. They had a reformation of cholesteatoma in 11% of these patients. Jansen (1978) reports a reformation of 7.75%, Smyth (1976) reports 24%, Charachon and Gratacap (1985) report 50%, and Jahnke and Falk (1976) report a rate of 40%. Lau and Tos (1987) operated on 122 children between 1965 and 1978. They had a 98% follow-up. The median observation period was 11 years. In the years 1980/1981, they noted residual cholesteatoma to be in 10% of these patients while recurrence was 2%. By 1985/1986 the residual cholesteatoma was 12.2% and recurrent cholesteatoma was 4.8%. Palva, Karma, & Karja (1977) found residual cholesteatoma to be 5% in their series, with a follow-up ranging from 18 months to 10 years.

One-stage intact canal wall techniques have been reported by Tos (1983). He observed his patients for a period of 2 to 16 years and had a 97% follow-up. Fourteen percent of his canal wall up patients went on to develop reformation of cholesteatoma, while 13% of the patients on whom he had performed canal wall down surgery went on to reform their cholesteatomas. Austin (1989) and Hirsch, Kameron, and Doshi (1992) recommend single stage procedures. They state that 80% of patients can be controlled with single stage surgery. The remaining patients can be operated on as and when the cholesteatoma reforms. They have not encountered any life-threatening complications of cholesteatoma in their series of patients. Prasad et al (2014) in their study found that the rate of residual cholesteatoma tends to decrease as age advances. The type of cholesteatoma acquired, or congenital middle ear were not statistically related to the incidence of residual cholesteatoma. Analysis of hearing results demonstrated that hearing recovery was excellent with canal wall up procedures and remained stable for five years.

Canal Wall Down Techniques

Advocates (Cruz et al., 1990) of the canal wall down techniques offer the following rationale for doing these

procedures: removal of disease, exteriorization of the mastoid to prevent complications of cholesteatoma from hidden growth of cholesteatoma, and this is a single-stage procedure. The procedure is preferred in noncompliant patients and in those who do not find attractive the option of intact canal wall procedures, with its attendant conditions of follow-up and second-look surgery. In the canal wall down procedures, the cholesteatoma is removed and the mastoid exteriorized. However, as the sinus tympani is the area from which cholesteatoma reforms from remnants left behind at original surgery, cholesteatoma reformation can still occur. The advocates of both intact canal wall and canal wall down procedures do not advocate techniques because of technical ease with which they can be performed, but as a philosophy with which the disease is treated. Both intact and canal wall down techniques are equally technically demanding (Jackson, Glasscock, Schwaber, Nissen, & Bojrab, 1985) if they are to provide optimal results. Improperly performed canal wall down techniques can lead to an unsafe ear, resulting in otorrhea and infection, which can lead to complications with both intracranial, as well as extracranial, manifestations. The canal wall down technique requires continued follow-up as much as the intact canal wall technique. This is necessary to provide regular evacuation of wax that builds up. Lastly, literature states that intact canal wall techniques provide better hearing results than canal wall down techniques. Rambo (1965) recommends obliteration of the mastoid cavity, especially if the cavity is very large. In principle, this avoids the problems associated with large cavities. However, if the matrix of the cholesteatoma has not been completely removed, the cholesteatoma can continue to reform in a hidden manner under the material used to obliterate the cavity. Thus, this can have the same effect as a hidden cholesteatoma in an intact canal wall surgery. Miller KM et al (2025) in their paper on outcomes in pediatric cholesteatoma ardently recommend Tympanomastoidectomy with canal wall down (TM CWD) procedures stating that it was associated with a lower rate of recidivism and recurrence. TM CWD had fewer surgical procedures than the canal wall up procedure.

Modification of the canal wall down technique has been recommended by Paparella (1977). He recommends that the bridge be preserved so as to facilitate reconstruction of the hearing mechanism. When should intact canal wall procedures be performed in children?

We offer the following guidelines:

1. The parents and their relatives have fully understood the nature of the surgery and its implications and agree to abide by them.
2. The disease is resectable.
3. The presence of serviceable hearing which is of a conductive nature.
4. No complications of otitis media are present.
5. Adequate eustachian tube function is present
6. The patient's general health permits a second procedure.

When should a canal wall down technique be performed?

We offer the following guidelines:

1. The parents and their relatives choose this technique, fully understand the outcome of this surgery, and agree to accept its outcome.
2. Unresectable disease.
3. Nonhearing ear or an ear with poor hearing.
4. If the canal wall has been destroyed by disease, it may need to be lowered.
5. Small contracted mastoid.
6. Poor follow-up is anticipated.
7. Poor eustachian tube function is present.
8. The patient's general health does not permit multiple procedures.
9. Canal wall down procedures for patients in whom complications of otitis media have occurred.

When should an intact canal wall procedure be converted to a canal wall down procedure?

We offer the following guidelines:

1. Extensive disease discovered at second-look procedure.
2. Destruction of the posterior canal wall.

Mastoid Obliteration

(Kroon et al 2023) present the first pediatric study on the safety and efficacy of mastoid obliteration using S53 bioactive glass (BAG) for cholesteatoma surgery in children. Canal wall up surgeries were performed in 30% of children and 70% had canal wall down surgeries. All received BAG to obliterate the mastoid. In this MRI controlled study the safety and efficacy of S53 BAG was demonstrated. Complications postoperatively were minor and rare. A dry ear was achieved in a majority of patients. Problems identified were the high rate of recidivism which was 49%. Persistent hearing loss was also another problem that was encountered.

Relevance of modified Bondy mastoidectomy in children.

Udayabhanu et al (2020) in their study have demonstrated that when the modified Bondy mastoidectomy performed on appropriately selected pediatric patients suffering from cholesteatoma was found to be a reliable and effective way to manage this condition. None of their patients suffered sensorineural hearing loss. Just 5.5% of patients were found to harbor recurrent disease.

Canal plasty

Canal plasty is necessary in stenosis of the external auditory canal. Stenosis of the external auditory canal can lead to cholesteatoma formation in the external canal which can lead towards the tympanic membrane and into the middle ear with catastrophic consequences (Jamarun, Ong, Anastasius, Goh

2025). Canal plasty performed on a congenital stenotic external canal will prevent the formation of a cholesteatoma. (Elzomor, A, Firlie M, Orobello N, Murnick, J, Reilly BK 2023) and Kimura et al (2021) in their study on the effect of the condition (status) of the ossicular chain and choice of graft material following surgery for cholesteatoma in children note that preserving the ossicular chain confers significant hearing benefit. They also note that cartilage- myringostapedioplasty also confers significant improvement in hearing even when the incus is absent and even without partial ossicular prosthesis.

Staging of cholesteatoma

A number of staging systems for cholesteatoma have been devised. They were devised for cholesteatoma evaluation and for predicting outcomes following surgery. They were not devised for cholesteatoma in the pediatric population in particular. However, **Marchand 2025** reports that the Potsic, (Potsic et al 2002) CHoLE (**Linder et al 2018**), EAONO/ JOS (**Yung et al 2016**) and STAMCO cholesteatoma classification can be applied in children. He also states that their contribution remains limited when it comes to predicting outcomes. He and his colleagues found the Mod-Pot classification to be much more useful because it was simple and intuitive.

The endoscope and its application for removal of cholesteatoma in children

In recent times the endoscope has gained significant popularity for the surgical removal of cholesteatoma. Many authors advocate its usage citing magnification, ability to view areas which are not accessible with the microscope. Some authors cite using it along with the microscope in a hybrid manner, (Choi, , Kwak, Kang, Chung 2022), (Kim Yang, Cho 2022), (Nassif, de Zinnis 2024) (Ueda et al 2024).

The endoscope can be used in canal wall up procedures to evaluate the mastoid cavity for recurrence/ recidivism. Under suitable anesthesia a small incision is taken post aurally and the endoscope is inserted into the mastoid. If no cholesteatoma is seen the procedure can then be terminated. This avoids a large postaural incision which could increase costs and prolonged hospital stay. This is a day care procedure involving a short stay in hospital. In experienced hands it is a safe, reliable and bloodless method of evaluation for recurrent / recidivism cholesteatoma. If a cholesteatoma has been observed to be present then the surgeon can decide on a course of action necessary for the eradication of disease. (Dixon, James 2020). The safety and efficacy of the use of the endoscope in the surgery for the removal of cholesteatoma have been reported in several studies by numerous authors (Sun, Cen, Kuo, Cheng, Huang, 2025).

In addition, Hao et al (2021) report that the endoscope when used in combination with the microscope reduces the rate of recidivism / recurrence.

Summary

Cholesteatoma is classified into two general categories:

(A) Congenital

(B) Acquired.

- Acquired cholesteatoma in children present with clinical symptoms/signs that suggest advancement with/without destruction of the adjacent structures:

- It may be present with or without tympanic membrane retraction and/or perforation,
- Otorrhea is usually present and is foul smelling.
- Hearing deterioration, (either conductive or sensorineural or combined) is quite often present.
- CT/MRI findings will likely include soft tissue involvement, focal areas of bony erosion of the middle ear, and the mastoid

- A retraction pocket has the potential to develop into acquired cholesteatoma. This phenomenon takes place when the retraction pocket loses its ability of self-cleansing (removal of debris) and then starts to accumulate keratin debris.

- Acquired cholesteatoma can likely form from a retraction pocket located either in the pars flaccida, pars tensa, or both. This occurs when the basal cell layer invades via the basilar membrane that could likely be a result of the dysfunction of middle ear pressure regulation. Acquired cholesteatoma on occasion,

can also develop secondary to tympanic membrane perforation. Perforation could occur as a result of previous chronic otitis media, trauma, or iatrogenic causes.

- Acquired cholesteatomas do not present at birth.
- Cholesteatoma recidivism includes both residual and recurrent cholesteatoma. It is essential to differentiate them.
 - Residual cholesteatoma is the consequence of incomplete surgical removal of the cholesteatoma matrix.
 - Recurrent cholesteatoma is the consequence of the reformation of the retraction pocket following a complete previous surgical cholesteatoma removal.
- Acquired cholesteatomas are further subclassified into
 - retraction pocket cholesteatoma
 - pars flaccida (attic cholesteatoma)
 - pars tensa cholesteatoma
 - combination of pars flaccida and pars tensa cholesteatoma
 - non-retraction pocket cholesteatoma
 - cholesteatoma secondary to tympanic perforation (the so-called secondary acquired cholesteatoma)
 - cholesteatoma can occur following trauma or from otologic procedures.

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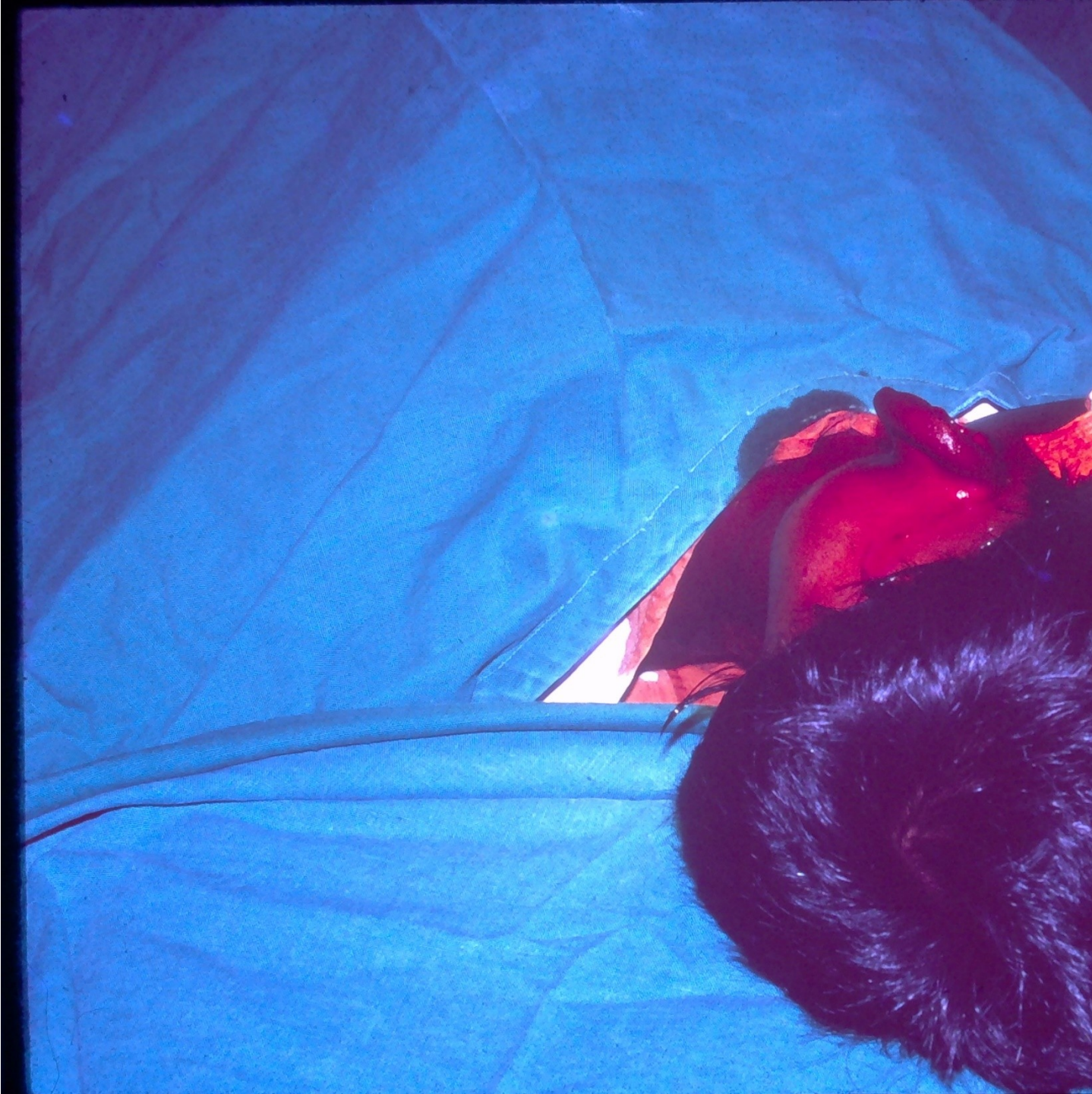


Figure 1: CT scan, Coronal View of the temporal bones demonstrating opacification of the middle ear. A congenital cholesteatoma encompassing and destroying the ossicles.



Figure 2: CT scan of the temporal bones, demonstrating a massive cholesteatoma in a child with sigmoid sinus thrombosis. The sigmoid sinus thrombosis presents as the typical delta sign.

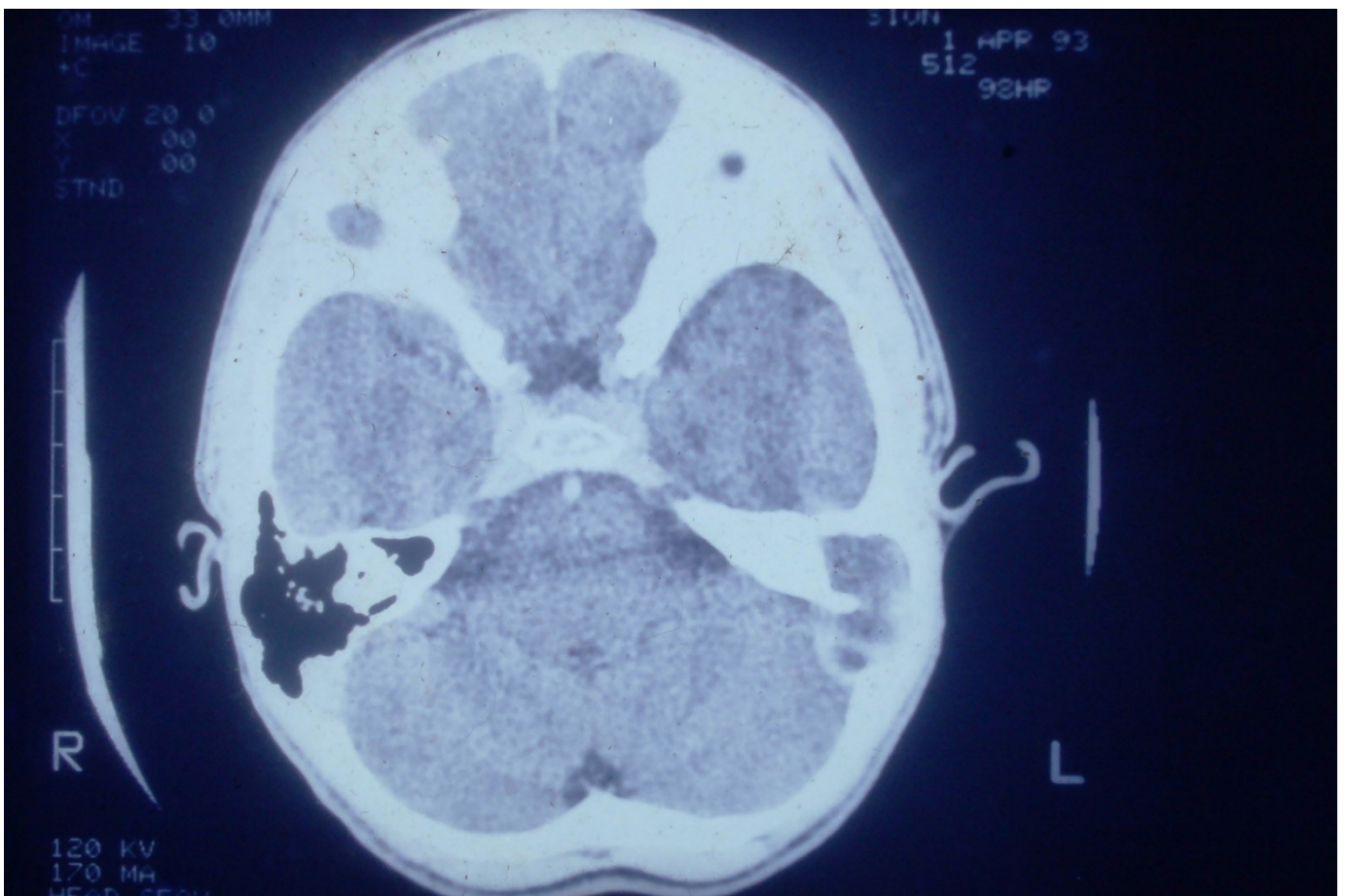


Figure 3: A massive postaural abscess in a child. The cause was a cholesteatoma that was infected.



Figure 4: A massive extracranial complication in the form of a postaural abscess caused by an infected cholesteatoma. The Pinna has been pushed forward.