Introduction

Histology is similar to the initial stages of AOM. There is mucosal and submucosal inflammation. Presence of submucosal infiltrate of histiocytes, lymphocytes and mononuclear cells. There is increased vascularity. There is increase in the number of mucous secreting goblet cells.

Can occur without previous episodes of AOM

Need not be associated with ear drum perforation

Infection can spread beyond the confines of temporal bone causing distant complications

Chronic inflammation of Mastoid and Middle ear cavity

Infection/Inflammation
May resolve due to treatment / immunity

Healing process

The ear responds by attempting to heal causing formation of granulation tissue, aural polypi, and new bone formation.

Changes left by chronic inflammation that could persist include:
- Submucosal scarring / fibrosis
- Increased mucous producing goblet cells
- Bony erosion
- New bone growth
- Thinning / perforation of ear drum
Classification

Ongoing active inflammation

Active COM

Inactive COM

Resolved / Healed COM

Active Mucosal

Active Squamosal

CP with Active discharge

Active attic discharge

Dry CP

Retracted drum

Healed CP

Tympanosclerosis
COM with Cholesteatoma

History

1. Muller coined the term cholesteatoma in 1838 mistaking it for a neoplastic condition.
2. He mistook the lesion to be keratin flakes which appear like cholesterol crystals.

Definition

It is defined as a cystic bag like structure lined by stratified squamous epithelium on a fibrous matrix. It is also known as skin in a wrong place.

Cholesteatoma Types

Congenital

Acquired
**Congenital Cholesteatoma**

**Definition**

It is an expanding cystic mass with keratinizing squamous epithelium located medially behind an intact ear drum. Usually diagnosed in infant / childhood. These children have no history of ear discharge / surgery / ear drum perforation.

**Theories of Congenital cholesteatoma**

1. Metaplasia theory
2. Invagination theory
3. Epithelial cell rest theory (Now accepted as etiology of congenital cholesteatoma).
4. Implantation theory

Petrous apex cholesteatoma with intact drum should always be considered as congenital cholesteatoma.

If discharge and TM perforation is present in a patient with congenital cholesteatoma then it should be construed that the cholesteatoma has eroded the ear drum.

<table>
<thead>
<tr>
<th>Derlacki &amp; Clemis Criteria</th>
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<tr>
<td>1. No h/o of previous ear infections</td>
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<td>2. Normal &amp; intact ear drum</td>
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<td>3. Incidental finding</td>
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Location of epidermoid cell rests can be seen commonly in the Peri tubal area.
Congenital cholesteatoma Staging

**Derlacki & Clemis staging**

1. Petrous pyramid cholesteatoma
2. Cholesteatoma involving the mastoid cavity
3. Cholesteatoma involving middle ear cavity

**Potsic staging**

- Stage I: Single quadrant involvement with no ossicular / mastoid involvement
- Stage II: Multiple quadrant involvement with no ossicular / mastoid involvement
- Stage III: Ossicular involvement without mastoid involvement
- Stage IV: Mastoid extension

**Nelson staging**

- Type I – Involvement of mesotympanum without involvement of incus / stapes
- Type II – Involvement of mesotympanum / attic along with erosion of ossicles without extension into the mastoid cavity
- Type III – Involvement of mesotympanum with mastoid extension

Stage of CC at presentation is directly related to the risk of residual disease after surgery.

Degree of hearing loss at the time of presentation is related to the risk of residual disease following surgery.
Acquired Cholesteatoma

1. Similar to adult disease in terms of definition & etiology
2. It is not present at birth, but develops with keratin epithelium invading the middle ear and temporal bone
3. Secondary acquired cholesteatoma may develop as a result of ingrowth of keratin epithelium associated with ear drum perforation
4. Severe trauma like temporal bone fractures can lead to keratin implantation within the middle ear / mastoid
5. Iatrogenic implantation can occur after surgery

Acquired cholesteatoma in children

Even procedures like grommet insertion may lead to keratin implantation

Fracture temporal bone
Pathogenesis of Primary Acquired Cholesteatoma

Mechanisms
1. Metaplasia
2. Retraction
3. Immigration
4. Basal cell hyperplasia / papillary ingrowth

Retraction
Retraction can occur both in pars tensa and pars flaccida. This is thought to be due to dysfunction of regulation of middle ear pressure.

Various theories have been proposed
Retraction theory is commonly accepted

Retraction causes migration of squamous epithelium into the middle ear.

Failure of cellular debris in the retraction pocket to migrate out may lead it to slowly invade the middle ear cavity.

Multiple factors could work to cause cholesteatoma like retraction and basal cell hyperplasia.
Theories of Primary acquired cholesteatoma

1. Cawthrone theory
2. Tumarkin theory
3. Toss theory of invagination
4. Wendt’s theory of metaplasia
5. Habermann’s epithelial invasion theory

Ear drum invaginates into the middle ear. Squamous debris migrate to this pocket from external canal.

Cawthrone theory
Suggested by Cawthrone in 1963. Cholesteatoma originated from congenital embryonic cell rests present in various areas of temporal bone.

Wendt’s theory
Proposed by Wendt in 1873. Pavement epithelium of attic undergoes squamous metaplasia in response to infection and aids cholesteatoma formation.

Tumarkin theory
Cholesteatoma occurs due to migration of squamous epithelium from the deep portion of external canal into middle ear through a marginal perforation or total perforation of ear drum. Total perforation is seen in acute necrotizing otitis media.

Habermann
Attic perforation caused squamous epithelium to migrate into the middle ear cavity.
Toss theory of Invagination

Toss postulated that persistent negative pressure cause invagination of pars flaccida forming a retraction pocket.

Retraction pocket later becomes filled with desquamated squamous epithelium from external canal.

Toss graded attic retraction into 4 grades.
Epidemiology

Incidence of cholesteatoma in children varies between races.

Caucasians / Africans have the highest incidence.

In 10% children it is bilateral.

8-15/100,000 Danish study.

Contra lateral cholesteatoma can develop during follow up.

Congenital cholesteatoma makes up 25% of pediatric cholesteatomas.

Children with congenital cholesteatoma present earlier than those with acquired cholesteatomas.

Grommet

Use of ventilation tubes may cause a decrease in the incidence of pediatric cholesteatoma because of improved middle ear ventilation.
Differences between pediatric & adult cholesteatomas

Similarities
1. Nature of disease process
2. Microscopic pathology
3. Treatment options

Pediatric temporal bone is well pneumatized when compared to that of adult thereby facilitating routes for spread of cholesteatoma.

Congenital cholesteatoma
- Commonly arises from anterior middle ear
- Completely fills middle ear cleft including ET
- Only later it spreads to attic and mastoid cavity

Areas of temporal bone affected

Temporal bone anatomy

Ossicular erosion common

Petrous bone involvement is not that common

Erosion of lateral canal and facial canal uncommon

Extent of Eustachian tube dysfunction is severe

Inflammatory reaction is more in children than adults

Tolerant to therapeutic interventions

Increase in the rate of recurrent / residual disease

Treatment limitations

More aggressive than adult type

Congenital cholesteatoma

Pneumatization
Reasons for increased recurrence / recidivism of pediatric cholesteatoma

- Middle ear disorders like AOM / OME are common
- ET function is poor
- ET immaturity persists even after treatment of cholesteatoma
- More prone to URI
- Extensive pneumatization makes disease clearance difficult
Symptoms

1. Hearing loss could be the only symptom in congenital cholesteatoma
2. Foul smelling scanty ear discharge
3. Complications can cause other symptoms like giddiness, facial palsy etc.

Difficult to examine the ear of a child

Signs & symptoms of cholesteatoma may be non existent in congenital cholesteatoma

Attic cholesteatoma as visualized in otoscopy. Destruction of outer attic wall is classic feature along with whitish flakes

Discharge from attic
Similar to that of adult cholesteatoma

Microbiology

Pseudomonas aeruginosa

Proteus Mirabilis

Staph aureus

Anaerobes
CT Imaging

1. CT is the preferred imaging modality
2. Reveals the anatomy of temporal bone in great detail
3. It also reveals breach of bony barriers if any due to cholesteatoma
4. Delineates the extent of cholesteatoma in great detail

Assess the degree of pneumatization of temporal bone

MR Imaging

1. Very useful in assessing soft tissue lesions
2. Very useful in assessing intracranial complications following dural erosion by cholesteatoma
3. Helps in assessment for the presence of residual cholesteatoma after surgery

Erosion of bony labyrinth and facial nerve canal can be visualised

CT coronal showing attic cholesteatoma

MR Image showing erosion of lateral canal
Treatment

Non surgical

Medical therapy will delay the complications till definitive surgery can be undertaken.

Risk of intracranial complication is 1 in 10000 every year.

During acute infections surgery should be postponed allowing conservative management to take place.

1. Drug therapy is not standard in cholesteatoma management.
2. Since cholesteatoma has no blood supply systemic antibiotics cannot reach the center of the lesion.
3. Topical antibiotics just surround the lesion, permeating just a few millimeters towards it center.
4. Chronic infections manage to persist even after aggressive antibiotic therapy.
**Goals of surgery**

- **Complete removal of cholesteatoma matrix**
- **Prevention of further erosion & complications**
- **Provide dry ear**
- **Provide a self cleansing ear**
- **Prevent recurrence**

**Secondary Goal**

Preservation / improvement of hearing is only a secondary goal.

- Normal hearing is only remotely possible
- Paediatric cholesteatoma is a difficult entity to treat
Surgical approaches

Canal wall up

In this procedure posterior canal wall is retained. This enables the middle ear depth to be maintained. Hearing reconstruction gives encouraging results.

Canal wall down

1. Also known as modified radical mastoidectomy
2. Used to clear extensive disease
3. May be used if petrosectomy need to be combined
4. Sometimes may be combined with obliteration of external auditory canal

Endoscopy assisted

1. Tympanotomy / Tympanoplasty
2. Atticotomy +/- reconstruction
3. Canal wall up mastoidectomy with combined approach tympanoplasty

Decision making

1. Extent of cholesteatoma
2. Whether long term follow up is assured
3. Presence of complications
4. Anatomy of temporal bone
5. Age of the child
6. Ability to tolerate microsuction during OP procedures
7. Ideal surgery should remove all cholesteatoma

A surgeon who operates should be able to perform a wide range of procedures.
Canal wall down procedure Indications

**Indications**

1. Extensive disease / aggressive disease
2. Erosion of canal wall
3. Need to remove outer attic wall
4. Anatomical reasons like poor mastoid pneumatization, low tegmen, and anteriorly placed sigmoid sinus

14% of patients who underwent CWU procedure needed CWD secondary procedure after follow up at a later date

43% of CWD procedures were performed due to anatomical causes

10% of patients need CWD procedure

Nearly half of the patients who underwent CUD procedure was due to the extent and severity of the disease
**Benefits**

- Normal ear canal / reconstructed ear drum heals well and hence it is dry
- Rapid healing is possible as only a small area needs to be re-epithelized
- Ear packing is needed only once immediately after surgery and can be removed as an outpatient procedure
- Normal hearing aid can be used if hearing outcome is not satisfactory
- Periodical suction not needed

**Downsides**

- There is a need for second look operation
- Recurrence is a possibility
- Technically more demanding
Canal wall down surgery (CWD)

Posterior canal and outer attic wall are removed to provide access to middle ear.

Provides best intraoperative access to all portions of middle ear cavity.

The Eustachian tube and remainder of middle ear cavity is sealed off from outside world.

Ideally a dry bony cavity lined by dry migratory squamous epithelium is created.

In 5% of these cases the ear could be still be wet.
Benefits & Downsides of CWD surgery in children

Benefits

- Only one major surgery is needed
- Post-op cavity can be inspected easily on a regular basis
- Reduction of volume of middle ear and mastoid. This leaves the ET with lesser space to ventilate hopefully reducing the chances of recurrence

Downsides

- Larger mastoid cavity leading to cavity problems
- If the cavity does not heal well then it leaves a persistent discharging ear
- Cavity needs to be kept dry. Cold water may stimulate labyrinth causing giddiness. Swimming should be restricted
- Sometimes difficult to use hearing aid in the cavity
- In cases of middle ear inflammation which is common in a child it can cause continuing ear discharge
Incidence of residual/recurrent cholesteatoma is common in children

Residual cholesteatoma is the keratin epithelium the surgeon has failed to remove during the initial surgery.

Recurrent cholesteatoma occurs when the squamous epithelium begins to grow into the middle ear cleft at a new site.

The term recidivism is a combination of residual and recurrent cholesteatoma.
1. Children with Down’s syndrome have increased incidence of cholesteatoma due to poor Eustachian tube function and mid face hypoplasia. The anatomy of the ear also could be very challenging because of narrow external canal, poor pneumatization of mastoid and low tegmen. Access to middle ear cavity via CWU procedure could be limited. CWD procedure need to be performed in these children. Facial nerve in these children could also be superficial.

2. Children with craniofacial syndromes have increased incidence of cholesteatoma.

3. Child with di George syndrome also have increased incidence of cholesteatoma