Cholesteatoma

Cholesteatomas, like perforations and retractions, are another type of chronic ear disease. In this tutorial we will learn about how cholesteatomas form and how they present to the ear care clinician. Understanding cholesteatoma requires you to have a good knowledge of the anatomy of the ear and the physiology of the middle ear and eardrum. These have been covered in the tutorial on the middle ear cleft so you should revise it before continuing with this one.

The word ‘cholesteatoma’ comes from two Greek words meaning ‘fatty tumour’. It was called this because of its appearance but these days we know that it is made from skin not fat. It is not a tumour either. Ear, nose and throat diseases often keep their older names, which can be confusing. A more modern name would be ‘chronic suppurative otitis media of a squamous type’. That is a lot to say so cholesteatoma is very often simply called ‘squamous disease’. Just to make life more confusing for you sometimes cholesteatoma is called attico-antral disease. In this tutorial we will just use the word cholesteatoma but you will come across the other names during your studies.

DEFINITION

The simplest way of defining cholesteatoma is to say that it is ‘skin in the middle ear cleft’. We will discuss how the skin gets there later but for now just think of cholesteatoma as skin that has somehow grown inwards. The middle ear does not normally contain skin and its presence there causes destruction of the middle ear components. As it grows it causes more and more problems and gives rise to complications. These complications are discussed below.

Skin can grow into the middle ear in a number of ways so let us look at classifications of cholesteatoma.

CLASSIFICATION OF CHOLESTEATOMA

There are a number of different ways in which skin can get into the middle ear. In the table below there is some new terminology regarding cholesteatoma. The list starts with the commonest type and ends with the rarest type.

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
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<tbody>
<tr>
<td>Primary Cholesteatoma</td>
<td>This is sometimes called primary acquired cholesteatoma. It is the commonest type and is caused when a longstanding negative middle ear pressure creates a retraction pocket in the eardrum. The retraction pocket fills with migrating skin that becomes trapped.</td>
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<tr>
<td>Secondary Cholesteatoma</td>
<td>Sometimes called secondary acquired this form happens when skin grows into the middle ear through a pre-existing perforation. The perforation may have been caused by longstanding negative middle ear pressure or by recurring</td>
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Congenital Cholesteatoma
This form is the rarest and happens because of a problem during the development of the baby's middle ear during pregnancy. Cells are left in the middle ear that can become skin. These cells usually disappear so that by the time of birth there are none in the middle ear.

Traumatic Cholesteatoma
For completeness it should be noted that trauma to the eardrum, such as during surgery, can push skin cells into the middle ear that may grow into a cholesteatoma.

FORMATION OF PRIMARY AND SECONDARY ACQUIRED CHOLESTEATOMAS
The formation of a cholesteatoma is easy to understand when you know the normal anatomy and physiology of the eardrum and middle ear cleft. You should take some time to review these before proceeding.

One piece of physiology that we have not yet discussed is that of epithelial migration in the outer ear. To understand the importance of migration we must remember that all cells have a lifespan. They are 'born', mature and die in a preprogrammed way throughout our lives. Cells in the skin of the forearm or leg, for example, go through this life cycle without moving much. When they die the cells simply flake away and are lost into the environment and the cells below replace them.

In the ear things are slightly different. Not only do cells have a life cycle but they move outwards during their life as well. This is important because if they didn’t move the ear canal would be filled with dead skin cells in about a month or so. With a canal full of dead skin we would not be able to hear properly.

The sequence of diagrams below illustrates this.

In image 1 an ink spot has been placed on the umbo.

A few days later the spot can be seen to have moved out towards the annulus.

Later again it will reach the annulus and migrate out onto the canal wall (4).

Eventually it will reach the concha and die.
Epithelial migration like this is a life long event. It almost never stops although some diseases can cause problems with it. When the ear has a deep retraction the migration of skin can lead to cholesteatoma formation. Consider the diagrams below. In the first we see a normal ear with normal epithelial migration. In the second a shallow retraction has formed because of chronic negative middle ear pressure. Review the tutorial on retraction pockets for more details on these. Skin can migrate into and back out of a shallow retraction without difficulty.

The third diagram shows a deep retraction. Skin can migrate into a deep retraction but it can’t migrate out again. Skin builds up in the retraction and a cholesteatoma is born.

1 Normal ear with normal migratory pattern
2 Ear with shallow retraction. Skin can migrate into and out of the retraction
3 Ear with deep retraction. Skin can migrate into the retraction but cannot escape again. Skin keeps migrating inwards. This is a cholesteatoma.

In secondary acquired cholesteatomas there is a pre-existing perforation that allows skin to migrate into the middle ear. This is less common than a primary acquired cholesteatoma.
You may ask yourself why skin migrating into the middle ear is so important. After all, skin is a benign waterproof covering. Why should it cause problems when it is in the wrong place?

This is an area of much research, however, it seems likely that once bacteria invade the cholesteatoma they form a biofilm and provoke an inflammatory response. It is this response that causes bone to be dissolved. As bone is dissolved the cholesteatoma can expand more. Some researchers also believe that the cholesteatoma can expand by the effects of pressure on bone but this is less likely.

**CLINICAL FEATURES - SYMPTOMS**

Of all the ear diseases cholesteatoma is considered the one that you should not miss. This is because it has the potential for causing significant complications and may even cause death. In the old days we called an ear with a cholesteatoma ‘unsafe’ but this is not a term that we use so commonly now.

Cholesteatoma is a silent disease until it causes a complication. In other words patients don’t notice that they have one until they get some hearing loss. In the early stages cholesteatoma does not cause pain, discharge or hearing loss. However, as the cholesteatoma grows it starts to cause symptoms. The symptoms depend on how much damage the cholesteatoma has caused and whether or not an infection sets in.

The commonest symptom is hearing loss and this happens as the retraction pocket extends and erodes the long process of the incus and suprastructure of the stapes. Clinical examination will reveal this to be a conductive hearing loss. Tinnitus often accompanies the hearing loss. Sensorineural hearing loss is a much rarer problem in cholesteatoma and only occurs when infection spreads into the inner ear.

The next commonest symptom is discharge. Usually the discharge is small in volume but it smells foul. Pseudomonas bacteria growing within the cholesteatoma cause the smell. The same bacteria also cause the discharge to be green in colour. The discharge does not usually run out of the ear unlike that of an infected perforation.

Other symptoms include facial weakness, headache and photophobia, vertigo, deformity of the pinna and swelling behind the pinna or in the neck but these only arise with more serious complications.

Symptoms are what the patient complains about. What about physical signs? Again, these will depend upon how extensive the disease is and what complications have arisen.
CLINICAL FEATURES - SIGNS

Cholesteatoma has many appearances. Here are some pictures of them with a brief commentary on each.

The first picture shows a posterosuperior crust that is golden in colour. There is some tympanosclerosis as well.

The second is the same ear but I have removed the crust under anaesthesia. Now you can see a posterior and superior retraction with skin within it – a cholesteatoma.

The first picture shows attic erosion with collection of keratin. The ear appears dry.

The second shows an attic polyp with accumulations of skin deep to it. Both of these are cholesteatoma.
The first picture shows an attic erosion with pus inside it. This cholesteatoma has destroyed the scutum and the head of the malleus and body of incus.

The second picture shows granulation tissue in the posterosuperior quadrant overlying a cholesteatoma. Granulation tissue arises when bone becomes inflamed as a result of the infection within the cholesteatoma.

The other common physical sign is a conductive hearing loss.

**COMPLICATIONS OF CHOLESTEATOMA**

The pictures shown above are common signs of cholesteatoma and so is a conductive loss. However, the disease can extend beyond the attic and ossicles to cause damage to other structures. This leads to signs and symptoms that are less common.

You must learn to ask for the symptoms of these complications when you see a cholesteatoma and you must also record the physical signs that you see. In this section we will look at the complications one by one and discuss both the signs and symptoms.

The commonest three complications of cholesteatoma in the UK are facial paralysis, mastoiditis with subperiosteal abscess and intracranial infection so we will look at these first.

**1. Facial Paralysis**

The facial nerve travels through the ear on its journey to the face and this makes it susceptible to damage from cholesteatoma. The nerve is covered by bone (the Fallopian canal) but this bone can be eroded by cholesteatoma and the nerve can become inflamed.

The patient develops a partial or complete lower motor neuron paralysis. This can
be treated and reversed if the patient has urgent surgery to remove the cholesteatoma.

The picture below shows an ear that has a very severe retraction and destruction of the ossicles. The horizontal section of the facial nerve is clearly seen travelling through the middle ear.

2. Mastoiditis with subperiosteal abscess

Mastoiditis is an infection within the mastoid bone and can be caused by acute suppurative otitis media that has extended posteriorly into the mastoid or by infected cholesteatoma.

In both these diseases pus builds up inside the mastoid air cells and pushes its way out of the mastoid in a lateral direction towards the skin behind the ear.

This creates a swelling behind the pinna that is fluctuant. The swelling pushes the pinna forwards and downwards.

The diagrams below show an axial section through the middle ear and mastoid. The first image shows normal anatomy. The second shows pus spreading backwards into the mastoid from the middle ear and the third shows the pus pushing through the bone over the mastoid and forming an abscess under the shin behind the ear. Note that this abscess pushes the pinna forwards.
This shows a normally pneumatised mastoid as it might appear on axial CT scanning. The black colour represents air in the mastoid cells.

This shows pus in the mastoid cells arising from an acute suppurative otitis media or cholesteatoma.

In this picture the pus has escaped from the mastoid cells and has pushed its way out under the skin behind the ear.

You can see an abscess pointing behind the pinna over the mastoid bone. The skin is red and the pinna is pushed forwards.

3. Intracranial infection

The commonest intracranial complications that you will see in Cambodia are meningitis and abscesses. Other complications such as otitic hydrocephalus and
sigmoid sinus thrombosis will also occur but will be difficult to diagnose. However, in the interests of completeness I shall describe them here.

**Meningitis.**

Infection spreading superiorly from the middle ear and mastoid through the tegmen or middle fossa plate comes to the meninges that surround the brain. These can become infected and if they do they produce symptoms and signs that are typical of the condition. The patient will complain of severe headache, neck stiffness, photophobia, fever, vomiting and sleepiness. Your examination may show a depressed level of consciousness, photophobia, neck stiffness and the patient may have epileptic fits.

These patients require emergency admission to hospital for antibiotics, fluids and careful monitoring.

**Brain Abscess.**

Abscesses are collections of pus surrounding a part of the brain or inside the brain itself. The symptoms are much the same as those for meningitis but there may also be double vision, paralysis, and ataxia.

An MRI or CT scan will diagnose the abscess. Again these patients require emergency management in hospital and the abscess may need to be drained.

**Sigmoid Sinus Thrombosis.**

In this disease the sigmoid sinus, one of the large veins draining blood from the brain into the internal jugular vein, becomes blocked by an infected blood clot. Patients present with fever, headache and a discharging ear. They may also suffer raised intracranial pressure particularly if the right side is affected, as this is the larger of the two sigmoid sinuses.

In very severe cases cranial nerves 9, 10 and 11 may become weak and cause hoarseness, difficulty swallowing and difficulties with moving the shoulder. Hydrocephalus may result in severe disease (otitic hydrocephalus).

**MANAGEMENT**

The risk of a serious complication from cholesteatoma (apart from conductive hearing loss which is almost always present) is probably one percent per year. Even though this is a low risk the complications are all severe so the treatment is almost always surgical removal of the disease.

Operations to remove the disease are divided into two basic types: those that remove the posterior canal wall partly or totally (canal wall down procedures), and those that leave the posterior canal wall intact. These are called canal wall up techniques.
The canal wall down operations are: atticotomy, attico-antrostomy, modified radical mastoidectomy and radical mastoidectomy. As a general rule small diseases limited to the epitympanum are dealt with by atticotomy while those were the disease has spread back through the aditus into the mastoid antrum are called attico-antrostomies. Cholesteatomas that have spread down into the mastoid are removed by modified radical mastoidectomy. Radical mastoid surgery is rarely performed these days.

All of these operations create an open mastoid cavity that is clearly visible during otoscopy. The cavity can be cleaned easily and the surgeon can easily see if the cholesteatoma returns. Having a cavity like this means that patients will need to be careful with getting water in the ears and the ear will sometimes discharge if the cavity does not heal properly.

Canal wall up operations do not leave a visible mastoid cavity and otoscopy will show a normal posterior canal wall. This has the advantage that patients may be able to get the ears wet and may not need to see their surgeon very often. However, a recurrence of the cholesteatoma is less easy to see and for this reason almost all patients will require a second operation to look into the mastoid about a year after the first operation – a second look procedure.

In developing countries the open cavity operations are more sensible as the expertise and equipment required to do canal wall up surgery is less likely to be available. Restrictions in CT scanning, difficulties with patient re-attendance for long term follow up and the need for second look surgery all make this type of surgery less attractive in these circumstances.

The image above is an axial section through the ear canal and it shows the position of the mastoid cells. It is these cells that are removed during a mastoidectomy. The next image shows the same axial slice during and after a modified radical mastoidectomy which is a canal wall down procedure.
An incision is placed behind the pinna and the skin is lifted off the bone and pushed forwards.

A drill is used to clear the bone from the mastoid and the disease is removed.

It is possible to do the operation with a hammer and gouge and this is how it is done in some developing countries but a drill is more precise.

Once all of the disease has been removed from the mastoid the skin incision is closed and the pinna is put back into its correct position.

When you look into the ear of the patient you will see that the posterior canal wall has been removed and that there is a large space lined with skin. This is the mastoid cavity.

Here is a photograph of a left ear that has had a canal wall down procedure.

The patient has also had a meatoplasty. This is the name of the operation to widen the cartilaginous portion of the external ear canal.

It allows easy access to the mastoid cavity for cleaning and helps to encourage healing and stability of the skin.

**COMPLICATIONS OF SURGERY**

Mastoid operations have some risks. The greatest of these are a dead ear (loss of all hearing) and facial paralysis. Fortunately these complications are rare. Other risks
are more common and include wound infection, wound breakdown, change in the sense of taste, cosmetic change to the pinna, tinnitus and imbalance.

Usually surgical management is successful and the cholesteatoma has been removed without damaging the patient. There is a small chance that it will return and this is one reason why most ear surgeons like to see their patients from time to time to check the ear.

Unfortunately some ears do not heal properly following surgery. This means that the skin that lines the cavity becomes inflamed and discharges. This is not usually dangerous for the patient but it can be annoying and may require revision surgery to cure it. Many patients will simply have their infections treated in the usual way rather than have surgery again.