

Cholesteatoma

A cholesteatoma is an abnormal growth or migration of skin from the ear canal into the middle ear. It has nothing to do with cholesterol, so its name is quite confusing. Skin can find its way into the middle ear because of chronic negative middle ear pressures associated with Eustachian tube dysfunction or by direct migration of skin into the middle ear through a perforation in the eardrum. Once the skin is inside the middle ear, it tends to accumulate and fill the spaces of the middle ear and mastoid (the bone behind the ear) and can cause many problems, including erosion of bony structures and chronic infections.

A cholesteatoma may be difficult to diagnose. This is because in many cases of cholesteatoma, you cannot see the cholesteatoma itself. You can only see the signs of its presence, such as a small opening in the top of the eardrum where the “mouth” of the cholesteatoma is present. At times, recurrent middle ear infections or hearing loss are the only signs of cholesteatoma. The constant infections make it impossible to see adequately into the ear and only a careful microscopic examination, sometimes done under anesthesia make it possible to identify that all of the trouble is caused by a hidden cholesteatoma cyst.

What causes cholesteatoma?

The ear canal is cleaned by the migration of skin from the center of the eardrum outward to the outer ear. If we mark the center of the eardrum with a spot of ink and look at the ear three weeks later, we will find that the ink has broken apart and migrated to the edges of the eardrum. Three weeks after that, the ink will be on the walls of the ear canal, and within 2-4 months, the ink will have migrated out of the canal altogether. This process of self-cleansing is necessary to prevent the accumulation of skin in the ear canals that could cause infection and hearing loss.

In some cases of Eustachian tube dysfunction (see the otitis media section for details), a hole does not develop in the eardrum, but chronic negative pressures in the middle ear cause the eardrum to be drawn into the middle ear. Just as plastic is “shrink-wrapped” around objects we buy at the store, the eardrum becomes wrapped around the bones of hearing and drawn deep into recesses of bone in the middle ear. When this occurs, the skin forming on the outer surface of the retracted eardrum cannot be cleaned by the natural process of migration outward, and instead accumulates slowly in the middle ear over a period of months or years. As this accumulation of skin grows, it erodes the bone around the ear and the bones of hearing. This accumulation of skin is called a cholesteatoma. Cholesteatoma can also form by growth of skin inward through a perforation of the eardrum.

In rare cases, skin from outside the eardrum can get trapped behind the eardrum when the ear is formed in utero (before birth). This can result in the silent growth of a cyst in an ear that is otherwise not prone to infection. This type of cholesteatoma is called a congenital cholesteatoma. Generally, a child with a congenital cholesteatoma may be three to six years of age before the cyst is

discovered because of its bright white appearance behind the eardrum and hearing loss.

Consequences of cholesteatoma

If cholesteatoma is left untreated, accumulation of skin under pressure will eventually cause it to erode pathways deep into the mastoid and skull. Fortunately, hearing loss caused by destruction of the ossicles or interference with their delicate movements occurs early. Eventually, the collection of dead debris will become infected, and the process of bone erosion will be accelerated. This infection tends to be resistant to antibiotics, because there are no blood vessels which carry the antibiotics in the debris-filled cyst. The only way to clear the infection is to remove the cholesteatoma.

Cholesteatoma has a tendency to erode bone. Bone erosion sometimes exposes the inner ear and facial nerve (nerve that controls the muscles of your face) directly to the cholesteatoma, causing injury by infection or making the possibility of injury during surgery higher. The cyst can also easily create a communication between the middle ear spaces and the cranial cavity, as these two areas are separated by only a millimeter of bone. An infection trapped by cholesteatoma in the middle ear or mastoid may find its way into the cranial cavity, causing meningitis. While this rarely happens in our medical culture, where patients have easy access to medical care, it is common to find patients hospitalized in clinics around the world with meningitis originating from neglected middle ear infections and cholesteatoma.

Surgery for cholesteatoma

Because of the far-reaching consequences of untreated cholesteatoma, when a cholesteatoma is suspected or discovered in an ear, surgery is generally recommended to eliminate infection, to minimize destruction, and to restore hearing. This surgery is generally known as tympanomastoidectomy. We will discuss the two parts to this procedure, mastoidectomy and tympanoplasty, separately.

Mastoidectomy: Because an enlarging cholesteatoma generally finds its way into the mastoid and its air spaces, surgical opening of the mastoid process behind the ear is fundamental to surgery for cholesteatoma. We call this type of surgery a mastoidectomy. Now feel the mastoid. It is the hard bone behind your ear which descends to a broad, dull point behind your earlobe. Surgery to remove cholesteatoma is generally done through an incision behind the ear and involves removing bone until the margins of the cholesteatoma cyst can be seen. The entire cyst and infected material around it are then removed. Ear surgery on an infected ear can be hazardous because important structures like the inner ear and the facial nerve are imbedded in the very bone occupied by the cyst and the infection. These vital structures must be preserved and the infectious process completely removed to prevent future recurrences. This is done under an operating microscope by an experienced surgeon who will paint away bone using a high speed drill and continuous irrigation. The goal is to reveal the structures

within the bone so they may be preserved, just as an archeologist excavates a site without injuring the artifacts contained within.

In performing this type of surgery, attempts are made to preserve the normal ear canal by removing the cyst through the eardrum on one side, and through the mastoid from behind the ear. In some cases, removal of bone from around the cyst results in removal of some of the ear canal bone that separates the normal ear canal and eardrum area from the mastoid surgery area, and a teaspoon-sized cavity called a mastoid cavity is created. This cavity may require cleaning periodically after surgery by the ear surgeon because the cavity cannot clean itself of skin the way the normal ear canal does. When the mastoid cavity is formed, the appearance of the external ear is slightly changed by enlarging the size of the ear opening. This makes it easier to see into the cavity and to clean it, and helps the cavity to stay dry and uninfected. At times, the cyst can be removed completely with complete confidence that there is no microscopic residual disease. In other cases, the cyst may not come out cleanly and there will be a suspicion that microscopic pieces of skin remain. If ignored, these microscopic pieces may grow, invade bone, and cause the symptoms of cholesteatoma to reappear.

In these cases, a “second look” operation may be recommended approximately six months after the first operation. Six months is generally long enough for these slow-growing collections of skin to grow large enough to become visible, but not long enough for them to once again become invasive. At most “second look” procedures, residual disease which is discovered can generally be completely removed.

Tympanoplasty: Often, portions of the chain of hearing bones are found to have been eroded by the cyst, and at the end of the operation, the eardrum must be rebuilt and a new connection must be made between the eardrum and the inner ear. Because the middle ear is called the “tympanum”, operations on the eardrum or on the bones of the middle ear are called “tympanoplasty”, which literally means to reshape the middle ear.

In tympanoplasty surgery, hearing bones eroded by the cyst may be removed, sculpted, replaced, or rearranged to make a new connection between the new eardrum and the inner ear for hearing. A number of different kinds of prostheses may be used to reassemble the ossicular chain. These prostheses are artificial hearing bones, and may be made of different forms of plastic, artificial bone, stainless steel, titanium, or even bone itself. Although their job is to conduct sound and replace missing hearing bones, they do not look anything like the bones they are replacing. Success with this type of ear surgery depends heavily on the experience of the ear surgeon. The prostheses must be carefully fit to small tolerances with a fit that is tight enough to stay in place until new tissue fixes their position, but not so tight that the chain of hearing bones will be stiff, as stiffness is bad for hearing.

The eardrum often must be reconstructed as well. Usually we use materials from your own body—most commonly, we use the covering (fascia) of the muscle above the ear, the covering of the cartilage (perichondrium) from the

external ear, or a thin slice of cartilage from the external ear. Often, when eardrum reconstruction is done, the reconstruction is stabilized in a mass of gelatin packing that will hold it still and protect it from infection until new blood vessels grow into the graft and incorporate it into the new eardrum. After about 3 weeks, the packing from the ear canal is removed and after about 6 weeks, the packing behind the graft will have dissolved. For more details of eardrum reconstruction, read Fixing a Hole in the Eardrum at <http://www.dbi.udel.edu/MichaelTeixidoMD/patientInfo/TympanicPerforation.html>.