

## CHOLESTEATOMA-OTITIS MEDIA

Acquired cholesteatoma (the congenital form is rare) begins when the patient develops otitis media, usually in childhood. Pus in the mastoid and tympanic spaces erodes through the tympanic membrane or through the thin layer of bone (“scutum”) separating the epitympanum (attic) from the ear canal. In a few cases the perforation fails to heal and squamous epithelium then may migrate from the external auditory canal through the perforation and come to line the epitympanum and sometimes the mastoid antrum and air cells. Frequently, in spite of extreme squamous epithelization of the epitympanum, the mesotympanum or middle ear remains clear except for its posterior portion where the cholesteatoma may wrap itself about the auditory ossicles and cover the fallopian canal. At times, cholesteatoma is seen as a white mass behind the tympanic membrane filling the middle ear but not attached to its walls. More commonly, the cholesteatoma, always white except for surface debris, presents as a small mass (looking like a pearl) filling a perforation in the superior portion of the tympanic membrane or adjacent bony canal wall, where it often is associated with malodorous purulent discharge and a bead of granulation tissue.

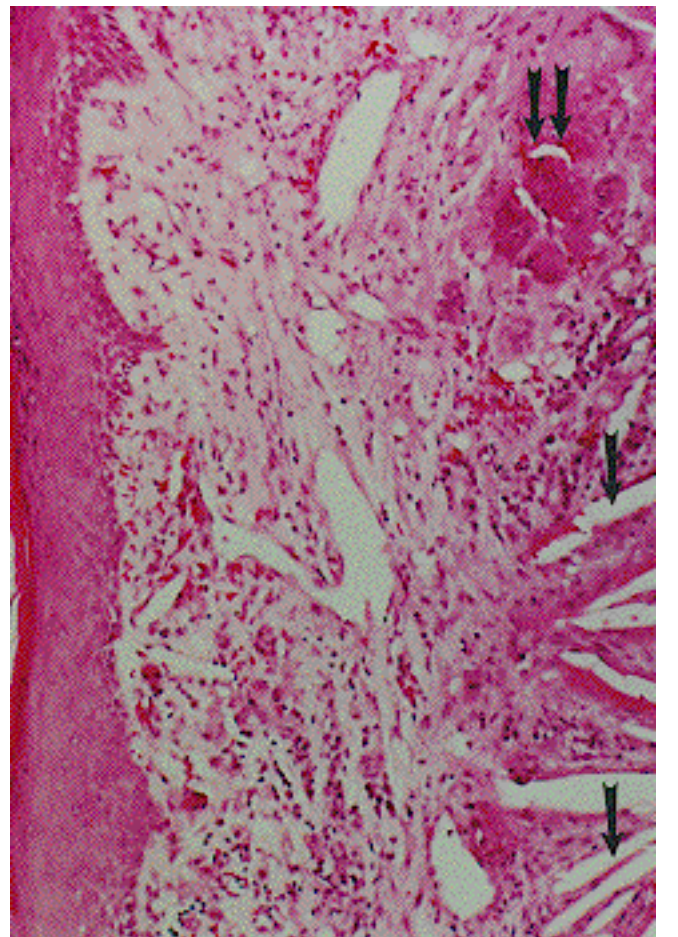
Symptoms develop when the cholesteatoma, by means of pressure erosion, destroys the ossicles, uncovers the facial nerve, erodes the horizontal semicircular canal, exposes dura of the temporal fossa or cerebellum, or causes other serious damage to important structures within or adjacent to the temporal bone.

Microscopically, squamous epithelium (called matrix by the otologist) forms a cyst of desquamating squames. The epithelium of a cholesteatoma and its cyst appears identical to that of epithelial inclusion cysts. The epithelium may rest on granulation tissue or fibrous tissue and sometimes a part of the cholesteatoma matrix overgrows normal mucosa but is not attached there.

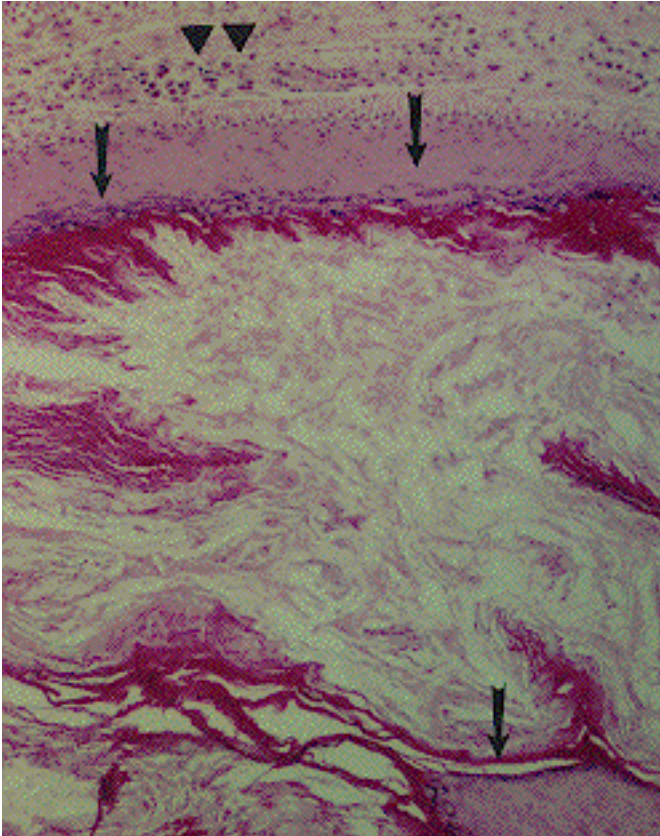
Extensive cholesteatoma that lined every mastoid air cell and wrapped about auditory ossicles destroying the incus and superstructure of the stapes, thereby causing a major conductive type hearing loss. Photograph shows a thickened keratinizing squamous epithelium.



Cholesterol crystals (single arrows) and reactive giant cells are a common finding in the granulation tissue associated with cholesteatoma. Here the giant cells (double arrows) are seen as part of the cholesterol granuloma but they can also occur as a foreign body reaction to keratin. Cholesteatoma itself does not contain cholesterol nor is it a tumor and, therefore, the name is a misnomer.







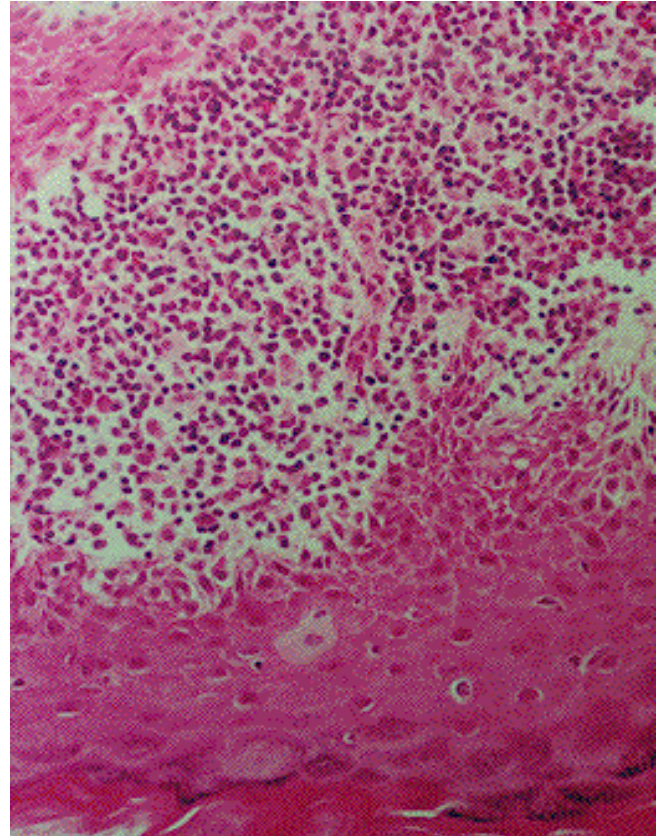
Cholesteatoma. Squamous epithelium or cholesteatome matrix (arrows) covers scar tissue (triangles) after mastoidectomy. If the mastoid cavity is exteriorized (“canal wall down” technique), then even though the bony wall of the drilled-out mastoid becomes epithelialized with squamous lining (desirable if the mastoid cavity is to remain dry), cholesteatoma does not reform since desquamating cells have ready egress to the enlarged ear canal.



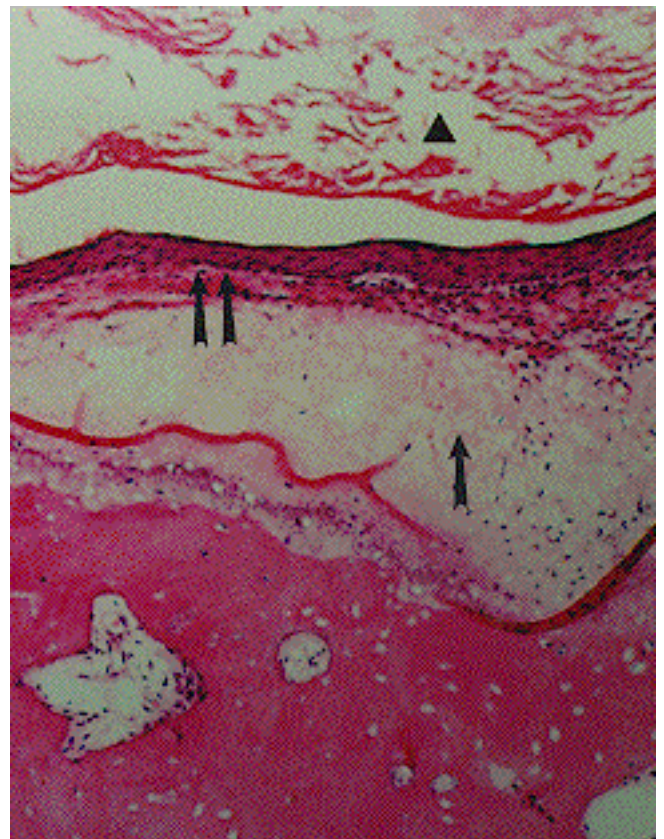
Cholesteatoma. Here the matrix (arrow) has been pulled intact from its attachment in the epitympanum. The cyst is comprised of desquamated squamous cells (triangles). One can understand how such a cyst, expanding over a period of years, could erode even the dense petrous portion of the temporal bone to produce, e.g., a fistula in the horizontal semicircular canal. Also such a cholesteatoma, after initially destroying the incus and causing a conductive hearing loss, may grow larger and actually serve to conduct sound from the tympanic membrane (which may be largely intact) to the stapes. Then, when the surgeon removes the cholesteatoma, hearing drops precipitously unless the ossicular chain is reconstructed. Microscopically, this cholesteatoma could pass for an epithelial inclusion cyst.



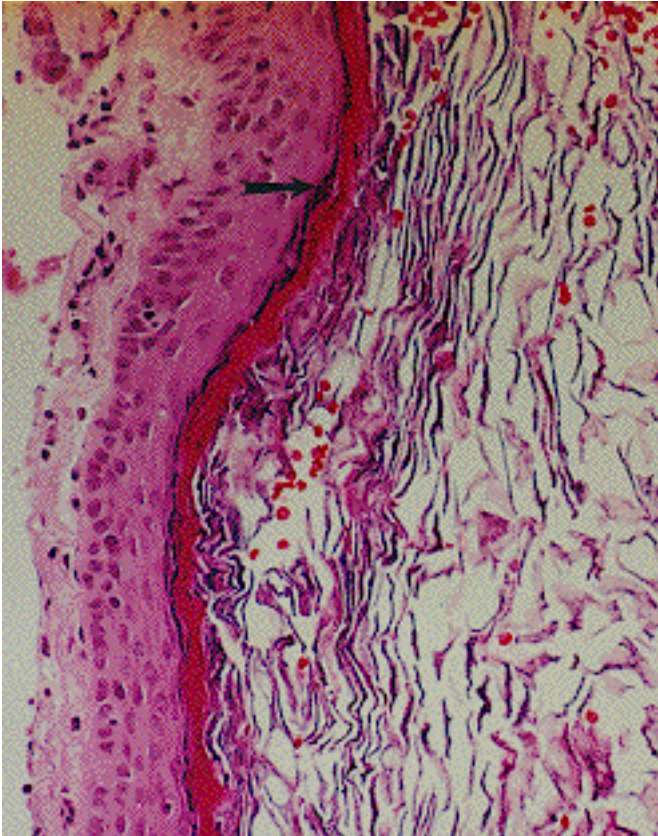
Cholesteatoma: Keratin and squamous epithelium rest on pyogenic granulation tissue. This combination accounts for the purulent discharge often associated with cholesteatoma. Whether or not cholesteatoma actually forms depends on whether squamous epithelium is "locked" in the mastoid or epitympanum or, just forms the outer surface of an aural polyp where squames cannot collect.



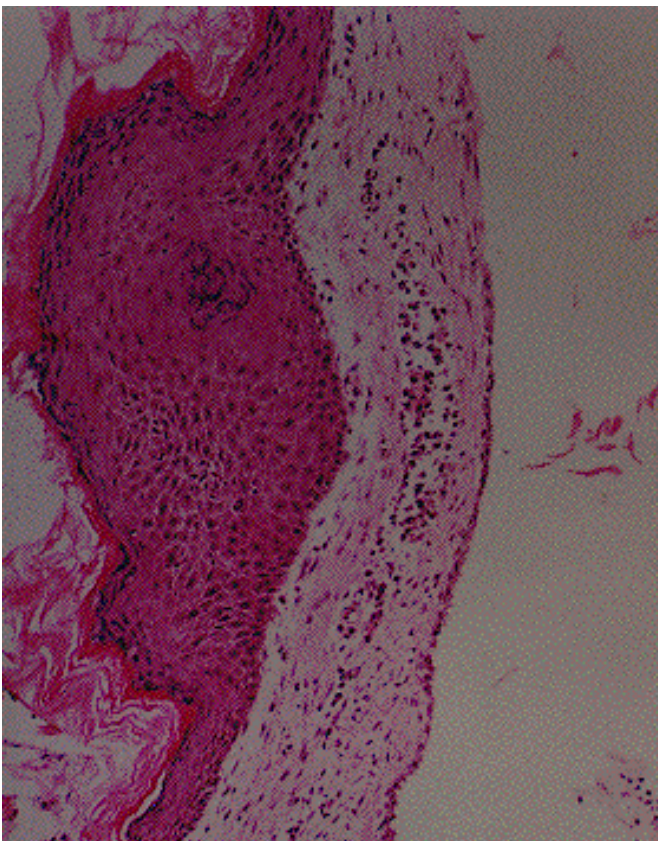
Cholesteatoma; the larger portion of the photomicrograph shows mastoid bone with edematous tissue (single arrow) between bone and squamous epithelium (double arrows); then there is an artifactual cleft and finally squamous debris (triangle). This is a plastic embedded section and the red line that looks like a blood vessel is an artifact.





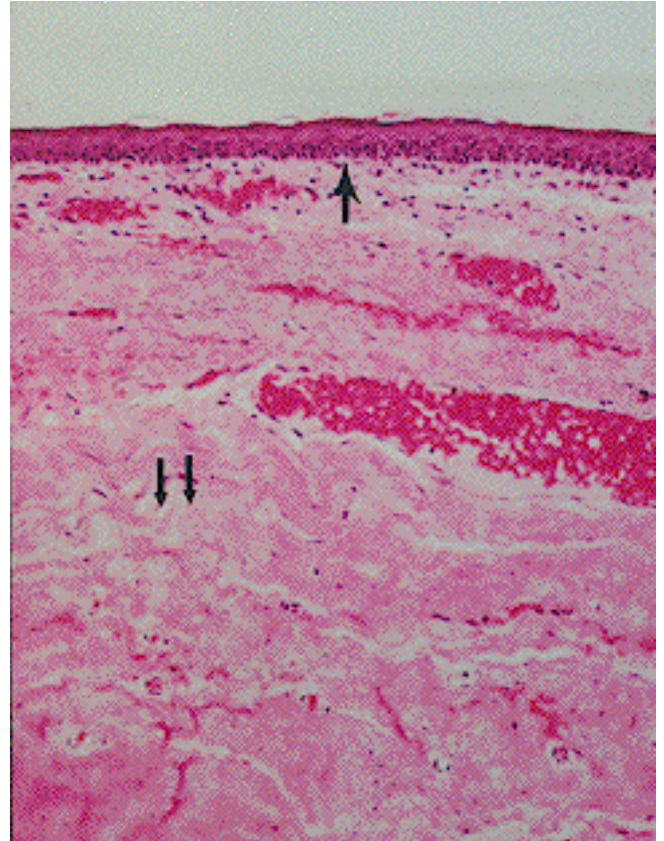


Cholesteatoma; even though there usually are associated inflammatory changes, dysplasia is uncommon. Rete pegs are usually absent and the epithelium, although fully developed, is thin. Here, all layers including the granulosa layer (arrow) are present. If a biopsy shows well developed rete pegs and dysplasia, one should consider the possibility of squamous carcinoma. Plastic section.

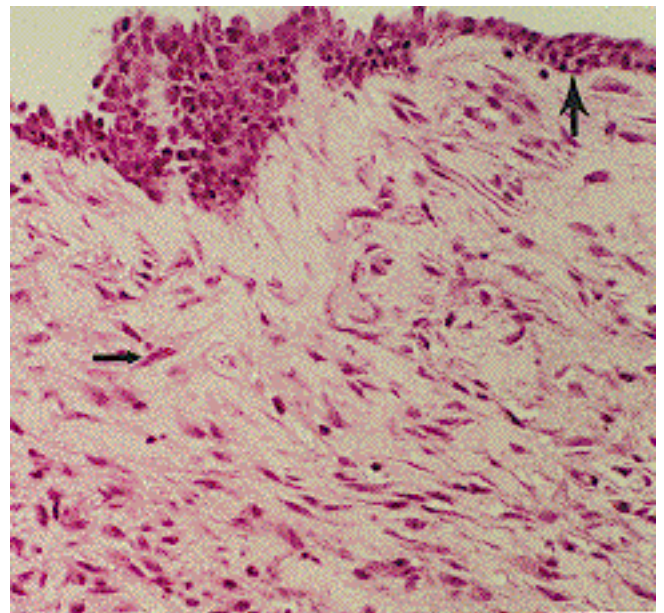


Cholesteatoma; tangential cut gives appearance of dysplasia, but this is normal epithelium as evidenced by the orderly arrangement of cells.

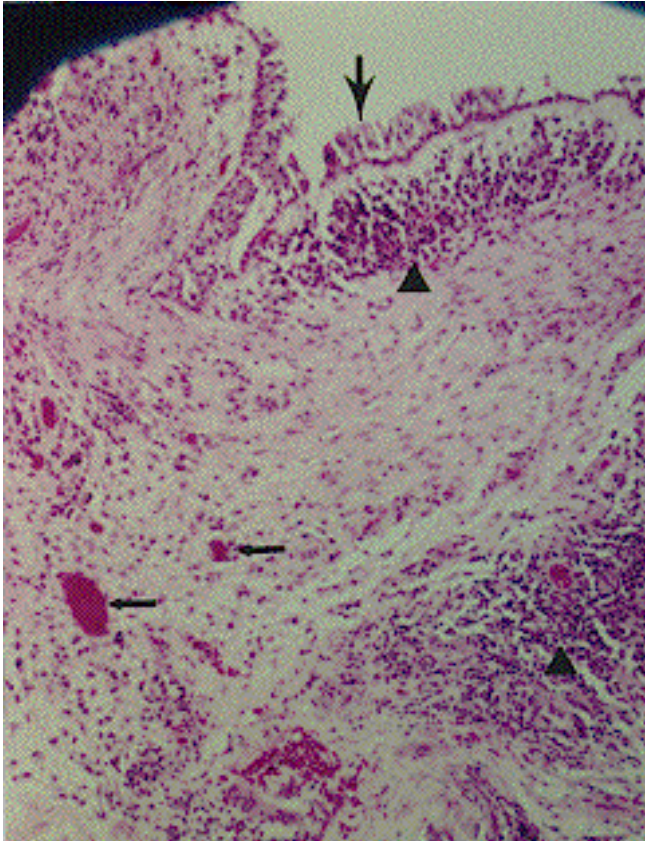
Cholesteatoma; matrix covering dense scar (double arrows) in epitympanum. Note how thin and atrophic the epithelium (single arrow) appears and that it is devoid of rete pegs. The usual ball of keratin debris has been lost in processing.



Mastoid, chronic inflammation. Fibrosis and chronic inflammatory reaction is seen in this specimen from the mastoid of a patient who had a chronic draining ear. A squamous type epithelium (large arrow) replaces the usual mastoid epithelium. Fibroblasts (small arrow) are predominant.







Chronic inflammation (otitis media), middle ear. Respiratory epithelium (large arrow) overlies marked chronic inflammation (triangles) and granulation tissue. Blood vessels (small arrows) are prominent. Patient had a draining ear for several months; specimen represents a polyp that protruded through a perforation of the tympanic membrane.

### CLINICAL ASPECTS:

Keratitis obturans, an infrequent but also frequently overlooked condition, is the accumulation of keratin debris in the ear canal just external to the tympanic annulus. It erodes the floor of the ear canal and causes pain. Treatment is by removal of all debris and if the condition is recurrent, it may be necessary to drill the floor of the bony ear canal so that it no longer represents a deep pocket external to the tympanic membrane but is flush with the entire canal.

Cholesteatoma may form in the middle ear (as contrasted to the mastoid) as the result of a perforation of the tympanic membrane that permits ingrowth of squamous epithelium from the external ear canal. It often is associated with chronic otitis media and an accompanying purulent otorrhea. Cholesteatoma of the middle ear and mastoid requires surgical removal of the squamous epithelium and often reconstruction of the ossicular mechanism destroyed by the cholesteatoma.

Cholesterol granuloma is not the same as cholesteatoma but may also cause middle ear destruction and require similar treatment as cholesteatoma.