

Cholesteatoma (Part 5)

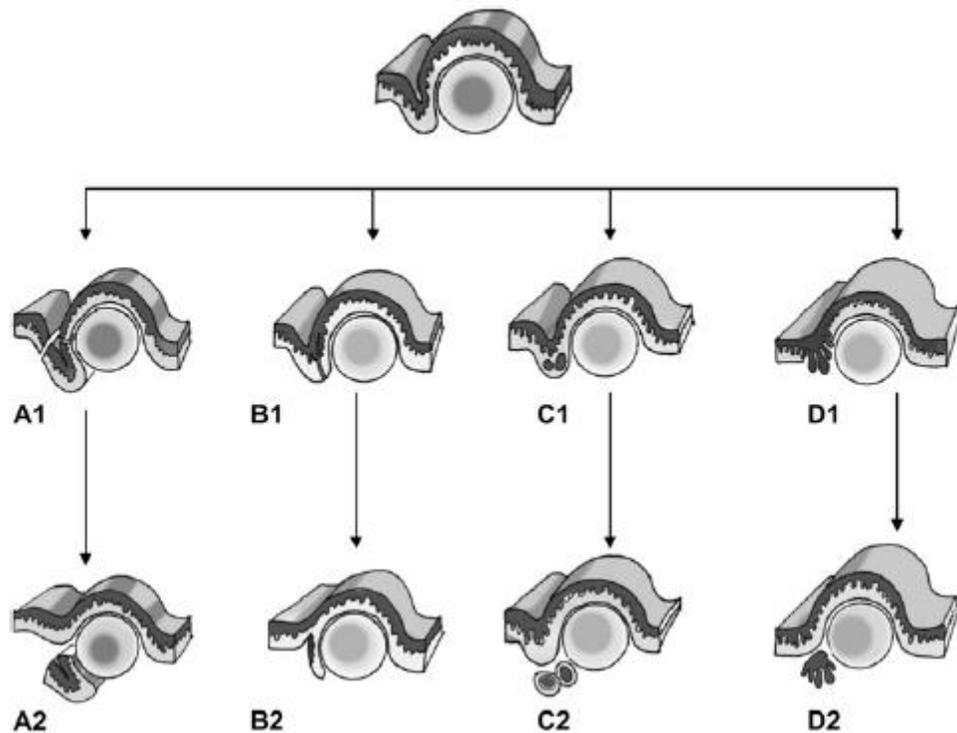
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Congenital Cholesteatoma

- CC of the middle ear is now thought to be on the rise, and accounts for 2% to 5% of all cholesteatomas
- Derlacki and Clemis described six cases of CC and established the clinical criteria for the diagnosis. These include
 - a pearly white mass medial to an intact tympanic membrane
 - a normal pars tensa and flaccida
 - no history of otorrhea, perforation, or previous otologic procedure.
- Levenson revised the criteria by adding that previous bouts of otitis media or effusion should not be exclusion criteria.
- The most common location of CCs is the anterior superior quadrant of the tympanic membrane, followed by the posterior–superior quadrant
- Two staging systems for CC of the middle ear have been suggested. The first, by Potsic [14], suggested the following stages:
 - ✓ Stage 1: Single quadrant with no ossicular or mastoid involvement
 - ✓ Stage 2: Multiple quadrants with no ossicular or mastoid involvement
 - ✓ Stage 3: Ossicular involvement but no mastoid involvement
 - ✓ Stage 4: Mastoid extension
- Nelson suggested classification of CC into three categories [15]:
 - ✓ Type 1: Mesotympanum with no incus or stapes erosion
 - ✓ Type 2: Mesotympanum or attic with ossicular erosion but no mastoid extension
 - ✓ Type 3: Mesotympanum with mastoid extension
- Theories of congenital cholesteatoma
 - Teed-Michael’s epidermal rest theory
 - Persistence of epithelial rests in lateral wall of eustachian tube

in proximity of the tympanic ring in anterosuperior quadrant of the middle ear

- Ruedi (OCNA 2006)
 - Inflammatory injury to an intact tympanic membrane results in microperforations in the basal layer that lead to invasion of the squamous epithelium by proliferating epithelial cones through a macroscopically intact but microscopically injured tympanic membrane
- Toss “Acquired” inclusion theory (OCNA 2006)
 - Observed that antero-superior cholesteatoma had a frequent attachment to the anterior aspect of the malleus handle or neck and that postero-superior cholesteatoma had an attachment to the posterior aspect of the malleus handle and to the incudostapedial joint
 - This location was far from the anterior tympanic annulus and the lateral wall of the eustachian tube where epithelial rests are usually found.
 - Furthermore, he speculated that if the site of origin was the lateral eustachian tube wall and the area anterior to the tympanic annulus, cholesteatoma would block the eustachian tube before extending into the tympanic cavity and the area of the malleus handle, a finding that has not been described previously
 - Therefore, he argued against the epithelial rest theory and explained the pathogenesis of congenital cholesteatoma by the acquired inclusion theory (Fig. 1)



- (A1, 2) The tympanic membrane retracted and adherent to the malleus handle, malleus neck, or long process of the incus is loosened and torn leaving a small cuff of viable keratinized epithelium adherent to the ossicles with a small residual tear in the tympanic membrane. As the tear heals, the included epithelium leads to formation of an inclusion cholesteatoma
- (B1, 2) A tangential tear is created as the retracted and adherent tympanic membrane is loosened from the underlying structure resulting in a remnant of epithelial cells without a perforation of the tympanic membrane that results in an inclusion cholesteatoma
- (C1, 2) Microperforations of the traumatized retracted tympanic membrane result in invasion of the basal membrane by

epithelial cones. As the ear drum is suddenly loosened, these cones are left behind and included in the tympanic cavity

- (D1, 2) Similar to the previous mechanism, repeated inflammation of the tympanic membrane result in proliferating epithelial cones that penetrate the basal membrane and proliferate into the subepithelial space. These cones are included in the tympanic cavity as the drum is loosened and detached from the underlying bony structures

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