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Ossicular Erosion from a Posterior Pars Tensa Retraction Cholesteatoma

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A 14-year-old lass with Down syndrome presented with a 3-year history of recurrent purulent left otorrhea. The discharge had become more frequently blood-tinged. Otologic examination revealed a stenotic ear canal with polypoid granulation obstructing the view of the tympanic membrane. High resolution computerized tomographic (HRCT) imaging of the temporal bone was performed to assess the status of the middle ear and mastoid. Particular attention was given to assess for bony erosion associated with cholesteatoma formation.

Comparison of the scutum on coronal view *(Figure 1)* did not show a marked visual difference in the sharpness of the scutum edge. As erosion of the scutum edge is the hallmark radiological finding in the diagnosis of a pars flaccida or attic retraction-based acquired cholesteatoma, a confident radiologic diagnosis of cholesteatoma via this pathophysiologic mechanism could not be given.

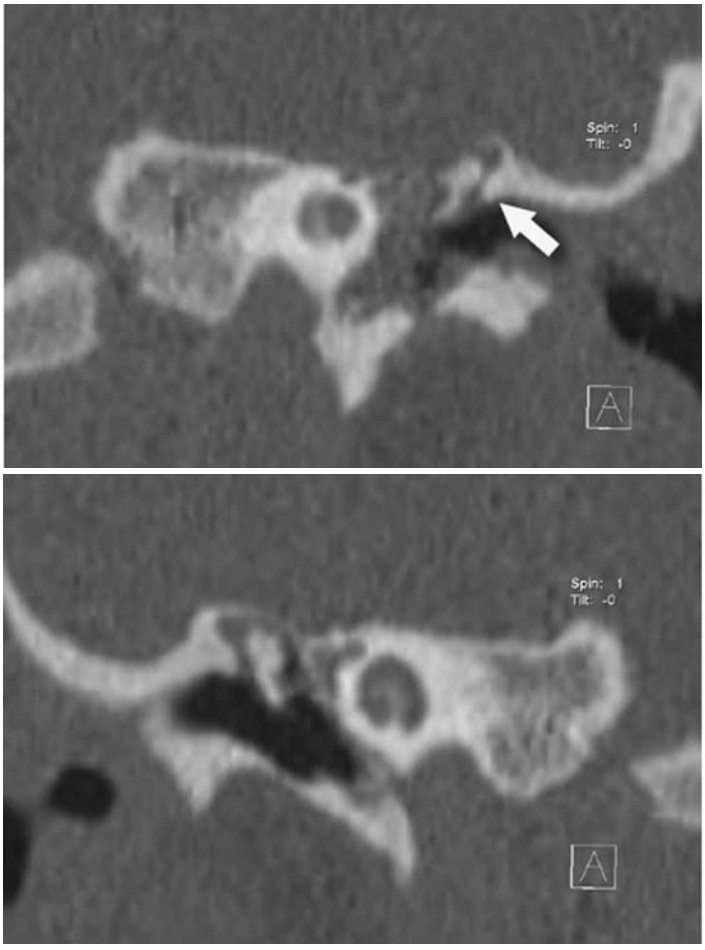

Figure 1. Coronal HRCT images of the scutum. The top scutum (white arrow) does not appear to be blunted when compared to the bottom scutum, which is known to be intact. The left middle ear space is entirely filled with a soft tissue lesion, whilst there is only soft tissue in the right epitympanum lateral and superior to the ossicles.

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Meticulous examination of the ossicles on axial view *(Figure 2)* showed a subtle thinning of the short process of the incus by sharply marginated, seemingly expansile soft tissue medially located between the incus and the tympanic segment of the facial nerve.

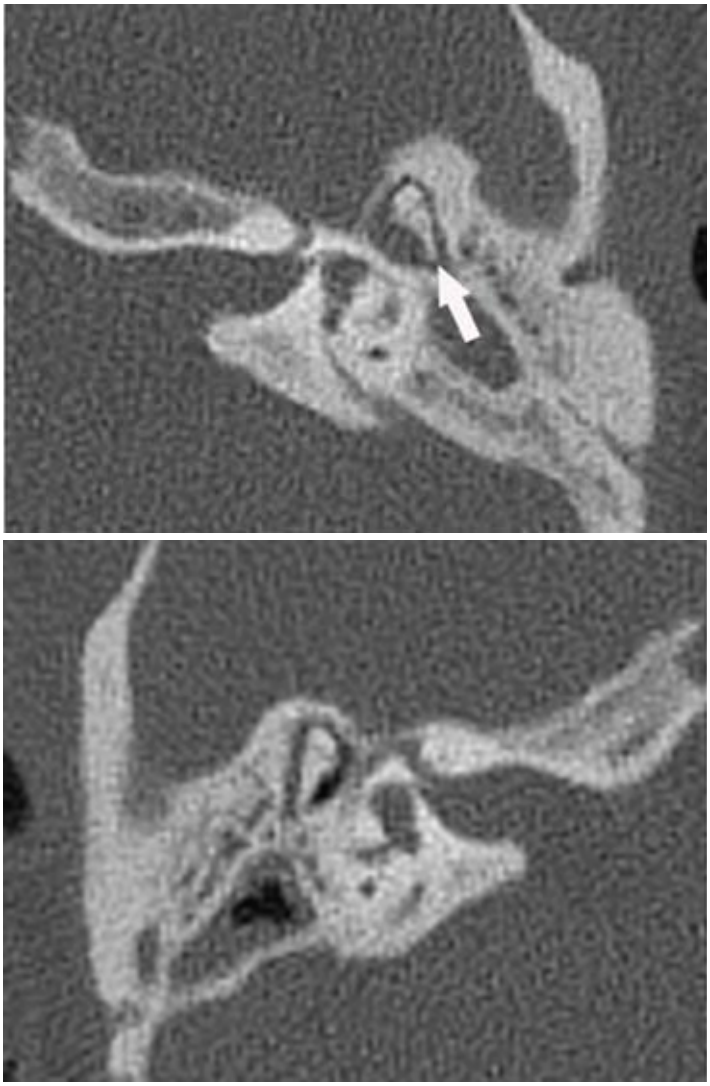

Figure 2. Axial HRCT images at the level of the epitympanum showing the 'ice-cream cone' appearance of the malleus head and incus body. The white arrow is pointing to the short process of the top incus, which appears to be subtly thinned from the medial side with increased concavity when compared to the bottom incus.

Exploratory surgery via an initial external end-aural approach revealed the presence of a posterior pars tensa retraction cholesteatoma underneath the granulation polyp. The cholesteatoma had extended medial to the ossicular chain causing erosion of the long process and medial portion of the short process of the incus, as well as the entire stapes superstructure. Definitive surgery consisting of a canal-wall down mastoidectomy via a post-auricular approach, cartilage graft tympanoplasty without ossicular chain reconstruction, meatoplasty and partial mastoid obliteration was performed.

This case is particularly instructive regarding two issues: otologic disease in Down syndrome and radiological evidence of erosive middle ear disease. An increased incidence of otologic conditions in Down syndrome is well established in the medical literature. These include external auditory canal stenosis, ossicular chain abnormalities and otitis media with effusion (OME).[1,2] Cholesteatoma as a sequelae of undiagnosed or untreated OME has to be suspected in children with Down syndrome, especially in those with recurrent otorrhea and persistent hearing loss. Unfortunately, the identification of a cholesteatoma may be difficult due to stenosis of the external auditory canal or a sub-optimal otologic examination due to behavioral problems in children with Down syndrome.[2] These factors were both present in this particular case, as the cholesteatoma remained undiagnosed for several years despite regular consultations with an otolaryngologist.

Radiologic evaluation with high-resolution computerized tomographic (HRCT) imaging is extremely important in these situations. As described by Barath *et al.*, the "typical findings associated with cholesteatoma include a sharply marginated expansile soft-tissue lesion, retraction of the tympanic membrane, scutum blunting, and erosion of the tympanic tegmen and ossicles. Holotympanic absence of bony changes is suggestive of otitis media without cholesteatoma formation, whereas presence of bony erosions (along with clinical suspicion) indicates cholesteatoma."[3] In this particular case, the presence of soft tissue within the epitympanum and antrum accompanied by the subtle evidence of ossicular erosion were crucial in the decision to advise and perform surgery. Although it may be argued that a high clinical suspicion based on the suggestive otological history in a child with Down syndrome may be enough to warrant surgical exploration, it cannot be disputed that the radiological findings help in advising patients pre-operatively about the indications for and expected outcomes of surgical management. In this case, it also impacted on the surgical approach – a transmeatal procedure appropriate for a limited middle ear exploration was initially performed with conversion to a standard post-auricular approach appropriate for more extensive mastoid surgery once the presence and extent of the cholesteatomatous disease was confirmed intra-operatively.

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