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Calcium pyrophosphate crystal deposition disease of the malleoincudal joint

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Abstract. This case report describes the clinical and imaging features of biopsy-proven calcium pyrophosphate crystal deposition disease of the malleoincudal joint in a 73-year-old woman. We discuss imaging clues that helped differentiate this very rare presentation of crystal-induced arthropathy from more common cholesteatoma in the diagnosis of conductive hearing loss.

Introduction

Calcium pyrophosphate dihydrate crystal deposition diseases (CPPDs) are characterized by the precipitation of crystals of calcium pyrophosphate dihydrate in connective tissue such as cartilage, ligaments, tendons, bursae, and joint capsules. Crystal deposition in articular and periarticular soft tissue can lead to acute and chronic inflammatory arthropathy of the involved joint. CPPD is also called pseudogout, chondrocalcinosis, and pyrophosphate arthropathy. In the head and neck region, CPPDs in and around the spine, the skull base, and the temporomandibular joint are well documented. Only one case of CPPD in the middle ear is published in the literature.[1] This case report discusses the clinical and imaging features of a biopsy-proven case of CPPD of the malleoincudal joint with an emphasis on the imaging findings that should prompt the physician to consider this rare disease presentation in the differential diagnosis of conductive hearing loss.

Case report

A 73-year-old woman consulted the otolaryngologist because of hearing loss and a feeling of pressure in her right ear. She had no relevant medical history. Her medications included acetylsalicylic acid and angiotensin II receptor blockers for supraventricular extrasystoles and hypertension. Micro-otoscopy of the right ear showed a retraction of the middle part of the anterior tympanic membrane along the malleus handle with a white discoloration towards the anterior epitympanum. The retraction of the tympanic membrane suggested a pathology underneath the eardrum rather than in the membrane itself, e.g. myringosclerosis. There was medialization of the malleus handle at the affected right ear. Pure tone audiometry showed hearing loss on the right side with an average air-bone gap of 40 dB at 250, 500, 1000, 2000, and 4000 Hz. Tympanometry showed a type C curve on the affected side and a normal type A curve on the other side. Tubomanometry showed normal bilateral tubal function.

Based on the retraction pocket, the whitish discoloration, and the conductive hearing loss pattern, the tentative diagnosis was cholesteatoma in the right middle ear. The anterior location of the retraction pocket was rather atypical. High-resolution computed tomography (CT) of the temporal bone showed a partially ossified mass in the right middle ear adjacent to the tympanic membrane and the malleus with a slight retraction of the tympanic membrane superiorly (Figure 1). The ossicular chain appeared intact. CT was inconclusive in differentiating the calcified mass, but it excluded the presence of a typical cholesteatoma.

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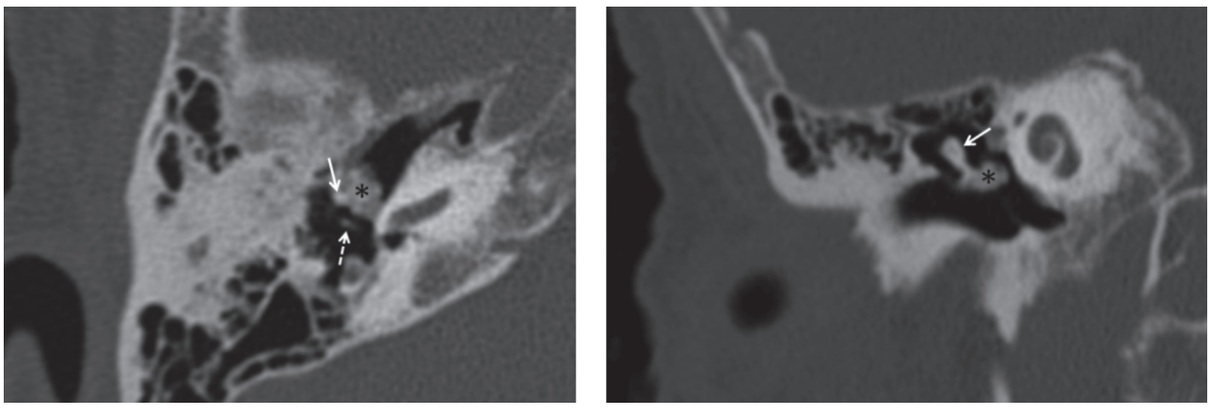

Figure 1

Computed tomography of the right temporal bone showing a partially calcified mass in the middle ear (asterisk; 5 mm x 6 mm x 2 mm). The axial plane is on the left, and the coronal plane is on the right. A solid white arrow indicates the malleus, and the dashed white arrow indicates the incus.

Oto-endoscopic surgery showed an empty retraction pocket that was not filled with epithelium, as would be expected for a cholesteatoma. A tympanomeatal flap was created, and inspection of the middle ear showed a calcified mass originating from the malleoincudal joint that extended along the malleus handle and retracted the tympanic membrane towards the middle ear cavity with medialization of the malleus. The mass was not surrounded by a typical sac, which is found in most cases of cholesteatoma. The mass easily fell apart during manipulation and consisted of multiple tiny white particles. After removal of the mass, palpation of the ossicular chain showed a significant gain in mobility. The retraction pocket was resected, and the defect was restored using Tutopatch®, a collagenous membrane derived from solvent-preserved bovine pericardium, using an underlay technique. The neotympanum was kept in place by Gelfoam® (absorbable gelatin) soaked in ciprofloxacin.

The pathology report described numerous multi-nucleated giant (foreign-body) cells and amorphous deposits with multiple chondrocyte-like cells within fibrotic connective tissue surrounded by an ossified border (Figure 2). Chondrometaplasia was also present. Crystals could no longer be seen with polarized light microscopy; this was attributed to decalcification/formalin-fixation. Taken together, the findings suggested a diagnosis of CPPD.

Postoperative endo-otoscopy performed 1 month after surgery showed normal healing of the tympanic membrane, and pure tone audiometry showed an average conductive hearing gain of 20 dB at the operated side at 250, 500, 1000, 2000 and 4000 Hz. Tonal audiogram showed a Fletcher index (FI) of 63 dB on the operated side and a FI of 53 dB on the other side. Follow-up was uneventful at 10 months and will be continued since there are no available data regarding the re-occurrence of this type of lesion.

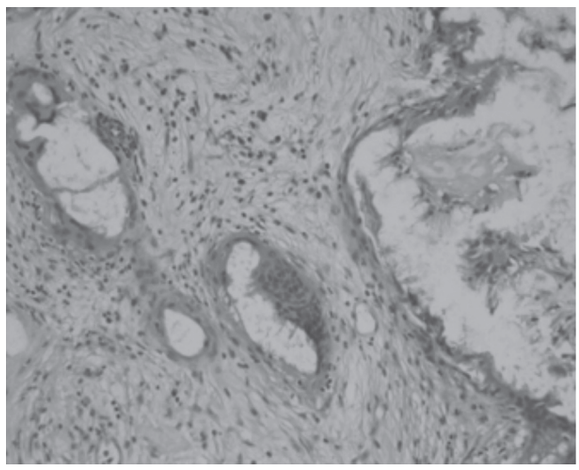

Figure 2

A biopsy of the middle ear mass. Multi-nucleated giant cells and amorphous deposits with multiple chondrocyte-like cells within fibrotic connective tissue are surrounded by an ossified border.

Discussion

CPPD is frequently asymptomatic and is often an incidental radiographic finding in elderly subjects[5].

Larger joints, such as the knee, wrist, elbow, shoulder, and hip joints, are most frequently involved in CPPD disease. CPPD may also be associated with symptoms such as acute arthritis symptoms – in fact, CPPD is the most common cause of acute monoarthritis in the geriatric population. There is a strong association between CPPD and age, with the prevalence increasing from 3% in those aged 55–59 years to 18% in those aged 80-84 years according to estimates based on radiographic studies.[5] The average age at diagnosis ranges between 65 and 75 years. Detailed prevalence data are not available for younger patients, and there is no major sex predominance.

The incudomalleolar joint is a saddle-shaped synovial joint that that contains a wedge-shaped meniscus; the joint is surrounded by an articular capsule. Because of this anatomy, the incudomalleolar joint is prone to crystal-induced arthropathies.[1] Signs of chondrocalcinosis in the incudomalleolar joint can highlight CPPD disease, reflecting the presence of granulation tissue, hypertrophied synovium, foreign-body granulomas, and calcified cartilage[1]. Gout, focal tympanosclerosis, and primary synovial chondromatosis can also lead to a non-traumatic calcified mass in the middle ear, although just one case involving gout and one case involving primary synovial chondromatosis are reported in the literature.[2,3]

During surgery, a white discoloration can mimic a cholesteatoma, so careful oto-endoscopic inspection of the tympanic membrane and retraction pocket is important. Periarticular masses/calcifications associated with gout or pseudogout are not located inside the retraction pocket. Endoscopic surgery was preferred as a minimally invasive approach to explore the epitympanum in this 73-year-old woman, avoiding an atticotomy or transmastoid approach. Endoscopy gives a more detailed view of the lesion/extension and also allows visualization of the content of the retraction pocket, allowing CPPD to be differentiated from myringosclerosis or cholesteatoma.

In imaging studies, crystal-induced arthropathies can reveal signs of chronic destructive arthropathy in combination with juxta-articular calcifications. Thus, the presence of bone erosion only around the incudomalleolar joint plus the presence of a periarticular calcified mass (i.e. calcium deposition) are important clues for differentiating this entity from a cholesteatoma on CT. These clues may influence the subsequent surgical approach, i.e. minimally invasive endoscopic surgery versus a more classic surgical approach.[4] In the two cases of gout and pseudogout reported previously, biopsy was necessary to establish the final diagnosis.[1,2]

Conclusion

Crystal-induced arthropathy of the incudomalleolar joint should be included in the differential diagnosis of conductive hearing loss, particularly when CT reveals bone erosion around this joint in combination with calcium deposition.

References

1. Shamil E, Willems S, Grolman W, Topsakal V. Pseudogout in the middle ear. *Otol Neurotol.* 2014;35(5):e202203.
2. Reineke U, Ebmeyer J, Schutte F, Upile T, Sudhoff HH. Tophaceous gout of the middle ear. *Otol Neurotol.* 2008;30(1):127-128.
3. Saggese D, Fernandez IJ, Salfi NM, Rinaldi Ceroni A. Synovial chondromatosis of the middle ear: a case report. *Ear Nose Throat J.* 2009;88(7):996-1000.
4. Deniz M, Uslu C, Koldaş C, Deniz B. Which technique is better for cholesteatoma surgery? *B-ENT.* 2015;11(2):109-115.
5. Richette P, Bardin T, Doherty M. An update on the epidemiology of calcium pyrophosphate dehydrate crystal deposition disease. *Rheumatology.* 2009;48(7):711-715.

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