Synovial chondromatosis of the middle ear: A case report

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Abstract

Synovial chondromatosis is a rare pathology of unknown etiology characterized by multinodular cartilaginous proliferation in the synovium. When it does occur, it usually develops in the large synovial joints (e.g., the knee, hip, elbow, and shoulder). Only rarely does it occur in the head and neck (most often in the temporomandibular joint). We report a case of synovial chondromatosis of the middle ear (i.e., the incudomalleolar joint) in a 64-year-old woman, and we describe the clinical, radiologic, and intraoperative findings in this case. To the best of our knowledge, this is the first reported case of synovial chondromatosis in the middle ear.

Introduction

Synovial chondromatosis, or synovial osteochondromatosis, is a rare pathology of the synovial joints. It most commonly affects the knee, hip, elbow, and shoulder. It is rare in the smaller joints and especially rare in the head and neck; several cases of temporomandibular joint (TMJ) involvement have been reported,[1-5] some with invasion into the middle cranial fossa.[1,2]

Synovial chondromatosis is a monoarticular disease that has a benign course, although rare cases of malignant transformation have been reported.[6] It is characterized by nodular proliferation of the synovial lining of a joint with detachment of fragments that may grow, calcify, and ossify.

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Previous presentation: The information in this article was originally presented as a poster at the XVIIIth International Federation of Oto-Rhino-Laryngological Societies (IFOS) World Congress; June 25-30, 2005; Rome.

The etiology of the disease is unknown; possible causes are trauma and infection. It is commonly accepted that synovial chondromatosis develops as a result of a metaplastic chondroid transformation of the connective tissue of the synovial membrane.[7] Some authors make a distinction between a primary form, which represents an active cartilaginous metaplasia, and a secondary form that is characterized by less cellular atypia, which is a passive process that involves intra-articular loss of fragments secondary to trauma, arthropathies, or arthritis.[4,7] While patients of both sexes and all ages can be affected, there is a predilection for males and a peak incidence in the fifth decade of life, probably because of the chronic and slowly progressive nature of the disease. The one exception with regard to the sex predilection is that synovial chondromatosis of the TMJ is four times more common in women than in men.[3]

Histologic examination reveals foci of chondrometaplasia in the synovium formed by disorganized cartilage with cytologic atypia, and a differential diagnosis with synovial chondrosarcoma should be performed. Milgram[8] classified synovial chondromatosis into three histologic stages:

- The first stage is characterized by the presence of active synovial chondrometaplasia without intrasynovial free bodies.
- The second stage is transitional, characterized by active intrasynovial metaplastic proliferation with free bodies that are partially covered with a synovial membrane that contains active chondrocytes.
- The third stage is synovial osteochondromatosis without synovial metaplasia and with free osteochondral bodies.

Surgery is considered the treatment of choice. The primary prognostic consideration is the possibility of a late recurrence. Some studies have shown that a partial

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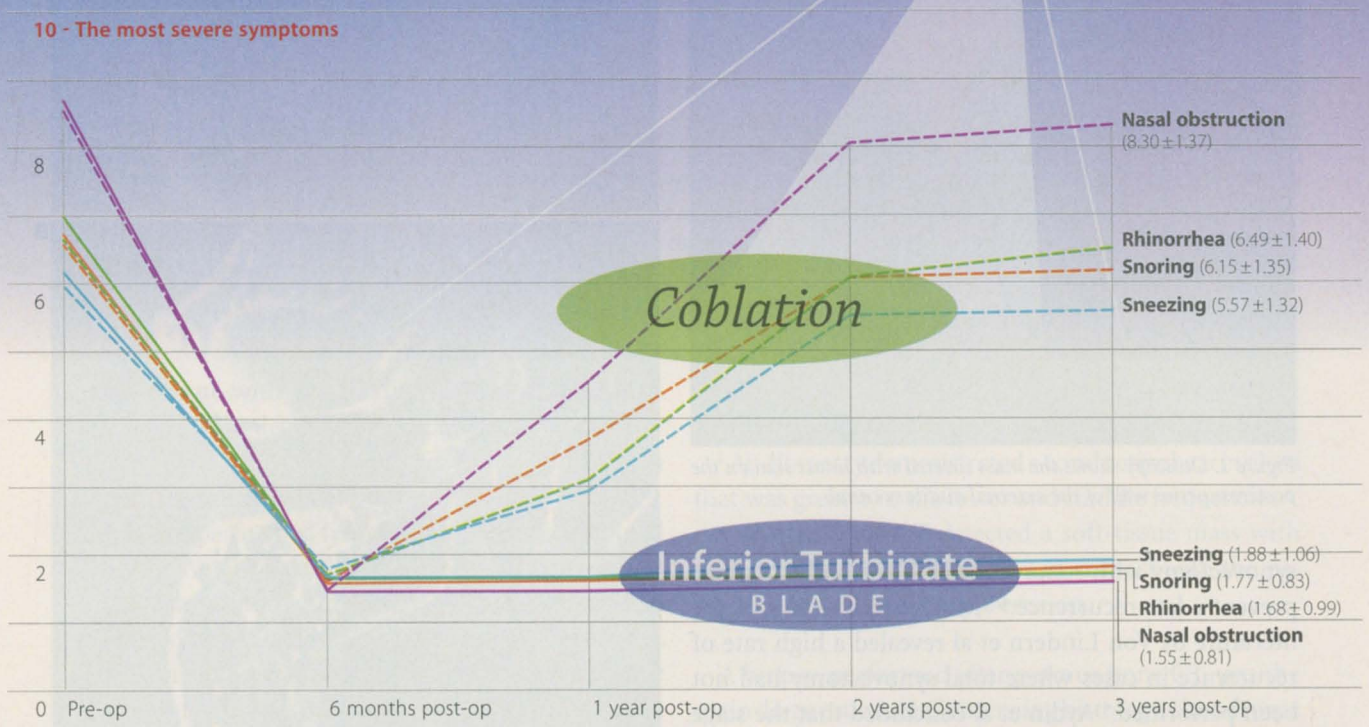
Visual Analog Scale (VAS) Scores



Key

Radiofrequency-Assisted Inferior Turbinoplasty - - - - - - - -

Microdebrider-Assisted Inferior Turbinoplasty ————

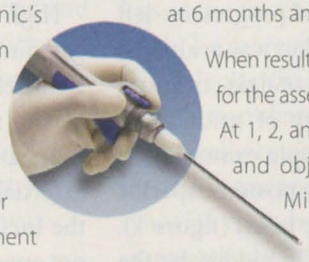
On the VAS Scale: 0 = No symptoms 10 = The most severe symptoms

Visual Analog Scale (VAS)

A subjective patient questionnaire that evaluates their perception of his or her health; in this case, pertaining to nasal obstruction, sneezing, rhinorrhea, and snoring. Answers usually range from zero (no symptoms) to 10 (the most severe symptoms).[1]

In the first three-year study directly comparing long-term efficacy of the two techniques, patients treated with Medtronic's microdebrider experienced significantly better long-term outcomes than those who received treatment with Coblation® radiofrequency.[1]

The study found that, compared with preoperative findings, subjective and objective measurements in the microdebrider group had improved significantly at all postoperative assessment times. In the Coblation group, significant improvements were observed at 6 months and 1 year, but not at 2 and 3 years.



When results in the two groups were compared, none of the values for the assessed variables were significantly different at 6 months. At 1, 2, and 3 years, however, all results for both the subjective and objective variables were significantly better in the Microdebrider group. Neither group experienced significant complications from surgery.

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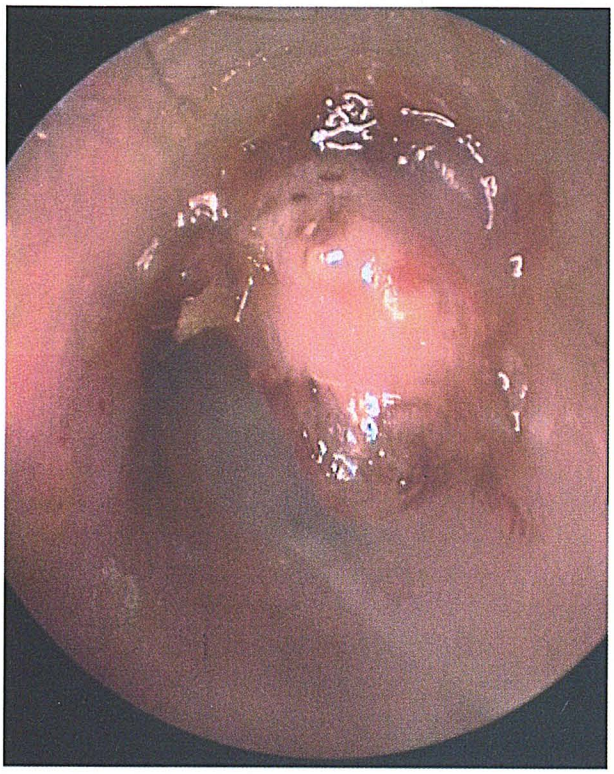

Figure 1. Otoscopy shows the mass covered with intact skin on the posterosuperior wall of the external auditory canal.

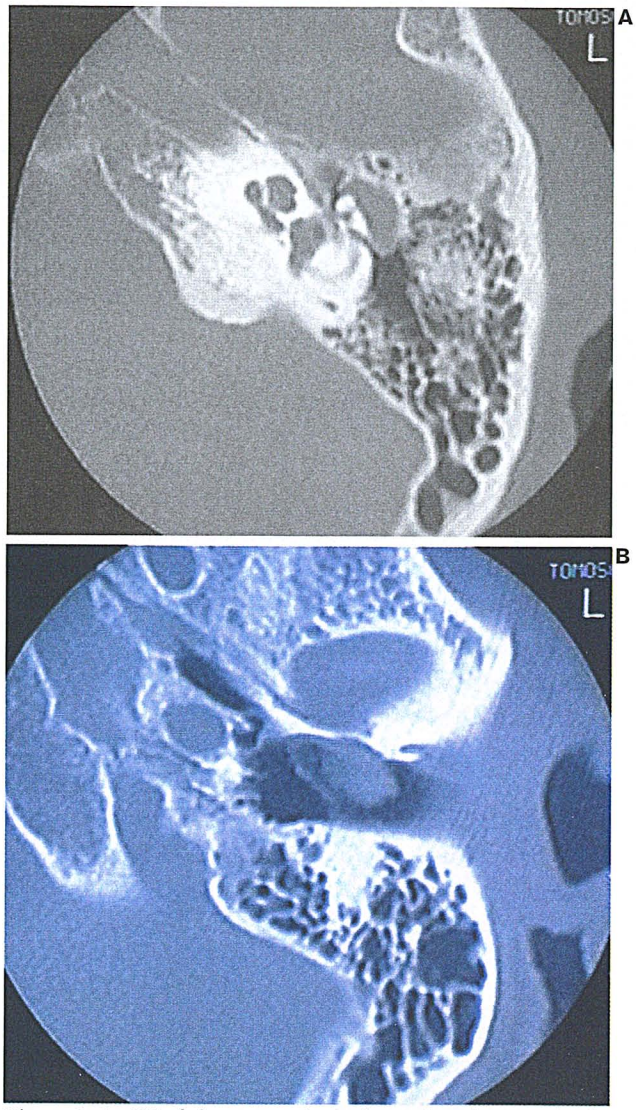

Figure 2. ***A:*** *CT of the petrous bone shows that the mass in the epitympanum is in contact with the incudomalleolar joint. The incus and malleus do not appear to be eroded.* ***B:*** *The mass extends from the aditus ad antrum to the external auditory canal. The pathologic tissue appears as a low-density mass that resembles a soft-tissue lesion.*

synovectomy with removal of all loose bodies cannot prevent a late recurrence.[4,9] Moreover, a review of the literature by Von Lindern et al revealed a high rate of recurrence in cases where total synovectomy had not been performed.[4] Aydin et al concluded that the stage of the disease is associated with the risk of recurrence, and they proposed a treatment based on stage.[5]

In this article, we report a case of synovial chondromatosis of the middle ear. As far as we know, no other case of middle ear involvement has been previously reported in the literature.

Case report

A 64-year-old woman complaining of progressive left hearing loss of several months' duration was evaluated in our outpatient clinic. She denied otalgia, tinnitus, and vertigo. She reported no history of middle ear otitis, trauma, or ear surgery. Otoscopic examination detected a firm mass bulging from the posterosuperior wall of the left bony external auditory canal (figure 1). The tympanic membrane was partially hidden by the mass. Audiometry revealed a severe conductive hearing loss in the lower and middle frequencies of the left ear; the bone threshold on the left was similar to that on the right.

High-resolution computed tomography (CT) of the petrous bone demonstrated a low-density mass with well-defined margins that resembled a soft-tissue neoplasm (figure 2). The mass had arisen in the epitympanum, and it extended from the aditus ad antrum to the external auditory canal. The lesion was in contact with the incudomalleolar joint. The incus and malleus did not appear to be eroded, but they were displaced toward the promontory. Phlogistic-appearing tissue occupied the mastoid cells. No abnormalities of the ipsilateral TMJ were found.

On biopsy analysis, the lesion featured cartilaginous

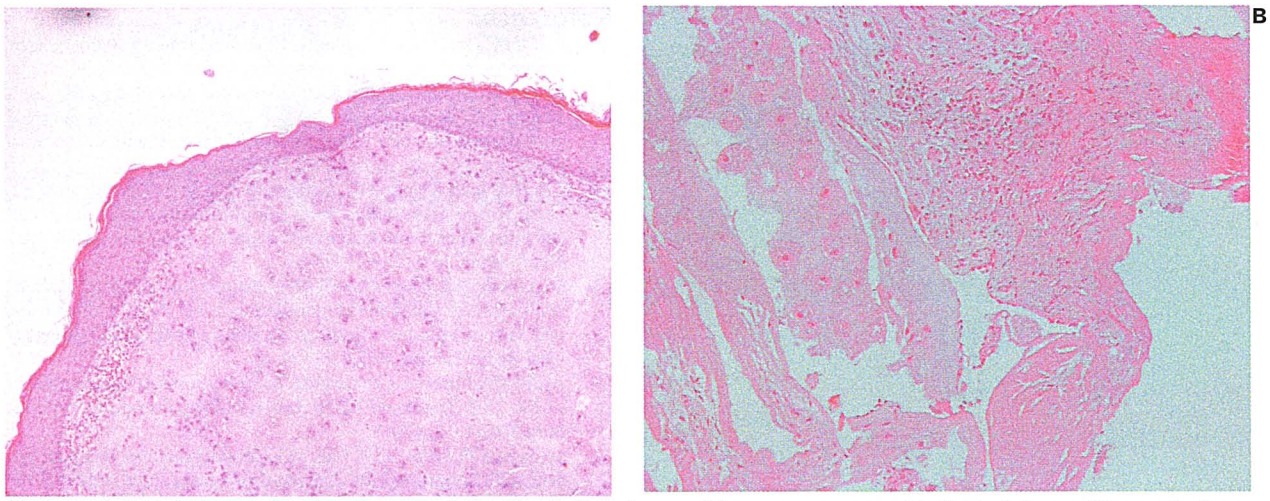

Figure 3. ***A:*** *Histologic examination shows a cellular island of cartilage pushing on squamous epithelium of the ear. The cartilaginous tissue exhibits a mild degree of cytologic atypia. The chondrocytes are arranged in small nests (H&E, original magnification ×50).* ***B:*** *Higher-power view shows the atypical cartilaginous tissue beneath the synovium but not involving it. Aspecific necrosis and calcifications are present (H&E, original magnification ×100).*

necrotic tissue with calcifications. Cartilaginous cells exhibited a mild degree of cytologic atypia, and they were arranged in small nests that were in contact with the synovial lining (figure 3).

Our team performed tympanoplasty via a combined approach. We exposed the posterior limit of the mass with a mastoidectomy and posterior tympanotomy and the superior face of the mass with an epitympanotomy. Intraoperatively, the lesion was as a lumpy mass (figure 4). Exploration confirmed that the mass extended from the aditus ad antrum to the external auditory canal and involved the incudomalleolar joint; the mobility of the joint was extremely limited. The incudostapedial joint, the footplate, and the malleus handle did not appear to be involved. To prevent a late recurrence, we removed the pathologic tissue with the head of the malleus and the incus as a whole. In the reconstructive step, we performed an overlay myringoplasty with autologous temporal fascia, a tragal cartilage scutumplasty, and an ossiculoplasty with placement of a total ossicular replacement prosthesis (TORP) between the footplate and the eardrum with cartilage interposition.

At 1 year of follow-up, otomicroscopy did not detect any tumor recurrence, and audiometry demonstrated a complete closure of the preoperative air-bone gap.

Discussion

Several clinical, radiologic, and histologic findings were of particular interest in our diagnosis of synovial chondromatosis of the middle ear:

- Otoscopy revealed a well delimited mass covered with intact skin.
- Audiometry demonstrated a conductive hearing loss that was greater in the lower and middle frequencies.
- Petrous bone CT detected a soft-tissue mass with well-defined margins that was in contact with the ossicular chain without bone erosion.

A biopsy, performed after petrous bone CT, was the main diagnostic tool. Concerning histology, chondroma and chondrosarcoma of the middle ear were the main differential diagnoses.

On the basis of our own surgical and histologic findings, we hypothesize that the pathologic process started in the incudomalleolar joint, which is a synovial joint like the other articulations that are more frequently involved by synovial chondromatosis. Our hypothesis is supported by the fact that our patient had a conductive hearing loss at the lower and middle frequencies. This finding might be more attributable to the increased rigidity of the ossicular chain secondary to incudomalleolar joint dysfunction than to a mass effect.

The cell atypia and the absence of a history of ear trauma in our patient supported a diagnosis of primary synovial chondromatosis. We excluded tenosynovial extra-articular chondromatosis—another entity that has been reported as synovial chondromatosis originating in the tenosynovial membranes[10]—in view of the absence of any involvement of the stapedius and tensor tympani tendons.

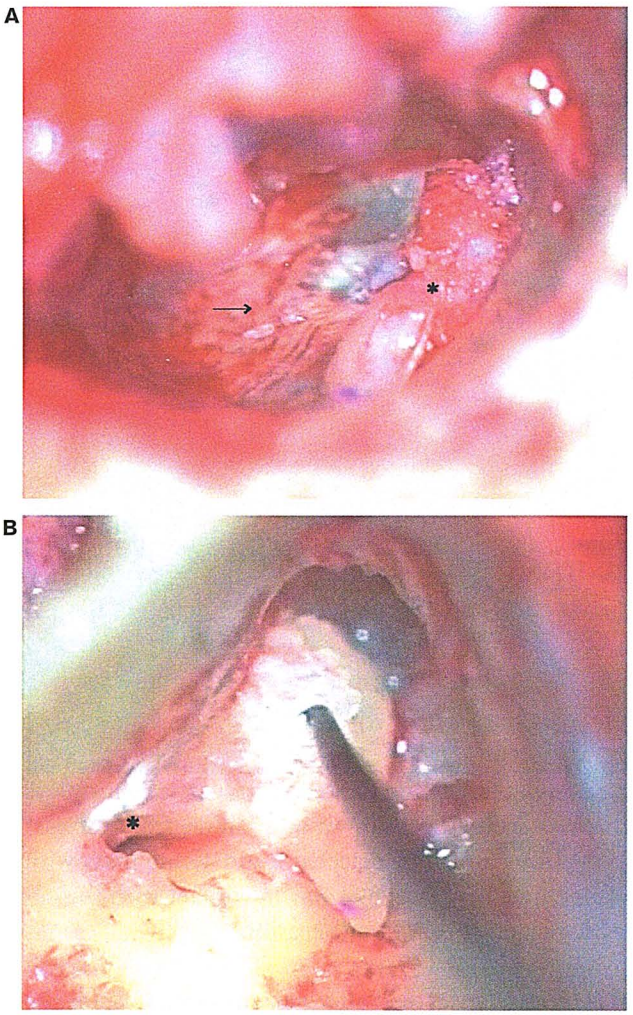

Figure 4. ***A:*** *Intraoperative endomeatal view shows the mass (*) under the posterosuperior quadrant of the eardrum (arrow) after preparation of the tympanomeatal flap.* ***B:*** *In this transmastoid view, an instrument points to the pathologic tissue on the articular surface of the incudomalleolar joint during removal of the incus through a posterior tympanotomy (*).*

Regarding our surgical strategy—and the management of the ossicular chain in particular—we decided to remove the incus and the malleus instead of taking a more conservative approach because of the risk of recurrence. This decision was made easier by the small size of the articulation involved.

In conclusion, synovial chondromatosis typically involves the larger synovial joints, but we found a case that involved one of the smallest joints in the body. To the best of our knowledge, this is the first case of synovial chondromatosis of the middle ear to be described in the literature. Therefore, synovial chondromatosis may be taken into consideration in the differential diagnosis of middle and external ear masses.

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