

A rare case of synovial chondromatosis of the inferior TMJ compartment.

Diagnosis and treatment aspect

Davide Sozzi, MD¹
 Gabriele Bocchialini, MD¹
 Giorgio Novelli, MD¹
 Maria Gabriella Valente, MD²
 Francesca Moltrasio, MD²
 Alberto Bozzetti, MD¹

¹ Maxillofacial O.U.S.Gerardo Hospital Monza, Department of Medicine and Surgery, School of Medicine, University of Milano-Bicocca, Milan, Italy

² Pathology O.U. S.Gerardo Hospital Monza, Italy

Corresponding author:

Davide Sozzi
 Maxillofacial O.U.S.Gerardo Hospital Monza, Department of Medicine and Surgery, School of Medicine, University of Milano-Bicocca
 Via Cadore, 48
 20090 Monza, Italy
 E-mail: davide.sozzi@unimib.it

Summary

Aim. Synovial Chondromatosis (SC) is a rare, benign non neoplastic arthropathy characterized by the metaplastic development of cartilaginous nodules within the synovial membrane. In only 3% of all cases does it affect the temporomandibular joint (TMJ) and cases that arise from the lower compartment are rarely found in literature. The aim of this paper is to report a new case of SC of the inferior TMJ compartment with the description of the clinical, therapeutic and histopathological findings.

Case report. This article presents a 68-year-old woman with preauricular swelling on the right side, pain, crepitus and limited joint motion. This patient was evaluated by preoperative clinical manifestation, CT scan and MR images. Both showed multiple, calcified loose bodies in the inferior compartment. Based on these images as well as the patient's signs and symptoms, a surgical intervention was performed. A good functional recovery with no signs of recurrence at 36 months of follow up was obtained.

Conclusion. Among cases of synovial chondromatosis in literature, only twelve originating in the lower compartment have been reported, this one included.

In all the cases treated for SC in the lower compartment, both in literature and in our case report, surgical treatment led to healing.

Key words: inferior compartment, synovial chondromatosis, temporomandibular joint.

Introduction

Synovial chondromatosis (SC) is an uncommon, benign lesion characterized by formation of small, multiple, metaplastic nodules of cartilage generally within the joint space.

SC usually affects large synovial joints such as the elbow, knee, wrist and hip. 3% of cases occurs in the temporomandibular joint (TMJ) (1).

According to the literature more than 200 cases of SC of the TMJ have been described (2) and the majority of these arises in the upper compartment with various degrees of extension (3-7). This type of condition can be locally aggressive and cases with intracranial extension have been previously described (8-10).

International literature shows only eleven cases originating in the inferior compartment, before this case (11-17).

This disease is also known as sinoval osteochondromatosis, synovial chondrometaplasia, synovial chondrosis, synovial metaplasia, sinovaloma, and periarthritic tenosynovial chondrometaplasia (3).

In literature Milgram (18), Gerard (19) and Chen (17) have offered three different classifications.

Milgram, in 1977, categorized the disease process into 3 distinct phases where we can find the relationship between the cartilage foci and the synovium.

In phase I, metaplasia of the synovial intima occurs. Active synovitis and nodule formation is present, but no calcifications can be identified.

In phase II, nodular synovitis and loose bodies are present in the joint. The loose bodies are primarily still cartilaginous.

In phase III, the loose bodies remain but the synovitis has resolved. The loose bodies also have a tendency to unite and calcify.

The Gerard's classification was completed in 1993 and was based on the synovial activity of the disease and we can find 4 stages:

Stage 1: presence of cartilaginous or fibrocartilaginous nodules with plenty of ground substance in the synovium.

Stage 2: presence of a very thick synovium with numerous small calcification or ossification cartilaginous nodules.

Stage 3: presence of large and ossified nodules.
Stage 4: the synovium is nearly normal or atrophic without any signs of metaplasia.
Only in 2012 Chen et al. made an essential clinical-radiological classification according to the different structures involved; they classified SC in the inferior compartment of the TMJ into 3 stages.
Stage 1 involves multiple loose bodies that are noted with expansion of the inferior compartment with no bony erosion.
Stage 2 involves multiple calcified nodules that are conglutinated to the condyle, the condyle is enlarged by pressure erosion but the disc is intact.
Stage 3 involves multiple calcified nodules conglutinated to the condyle, the condyle is destroyed as a result of a pressure erosion or by direct bony invasion of the mass, the inferior surface of the disc is involved, and the lesion can't be detached from the disc.
The aim of this paper is to report a new case of synovial chondromatosis of the inferior TMJ compartment with the description of the clinical, therapeutic and histopathological findings.

Case report

A 68-year-old woman was referred to the O.U. of Maxillo-Facial Surgery, Monza S. Gerardo Hospital, University of Milano-Bicocca School of Medicine with right sided preauricular swelling lasting 4 months, a limitation of mouth opening and pain aggravated by joint palpation and mastication. There was no history of trauma or other event contributing to the onset of symptoms.
The patient was edentulous and the maximal mouth opening was 20 mm with a deviation on the right side. The physical examination revealed that no facial nerve dysfunction, hearing or facial sensation disturbances were detected.
A computer tomographic scan (CTs) revealed the normal contour of the right condyle, no destruction of the glenoid fossa or temporal bone on the same side, no widening of the joint space, no bony changes of the skull base. It did reveal, however, several calcifying lesions in the right TMJ region (Fig. 1 a-b).
Magnetic resonance (MRI) showed multiple, calcified loose bodies in the inferior compartment; the disc was irregular and patchy and its position was anterior to the condyle (Fig. 2).
Because the CT and MRI imaging powerfully indicated a benign lesion, no exploration biopsy was done. According to Chen's classification (Stage I) a surgical intervention was performed under general anaesthesia with a right preauricular approach. The incision was followed by the exposure of the right temporomandibular region and cartilaginous nodules of varying size were visible floating in the lower compartment (Fig. 3).
All these cartilaginous fragments were removed together with the affected synovium (Fig. 4).
Removed the nodules, the articular disc resumed its proper position.

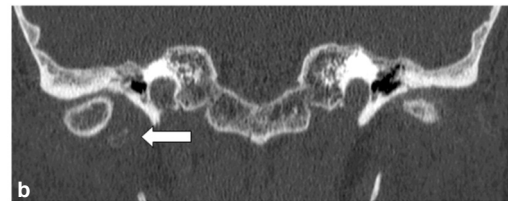
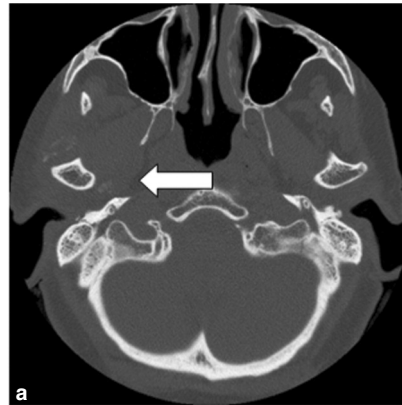


Figure 1 a-b. CT axial and coronal view showing calcifying lesion and the normal contour of the right condyle (arrows).

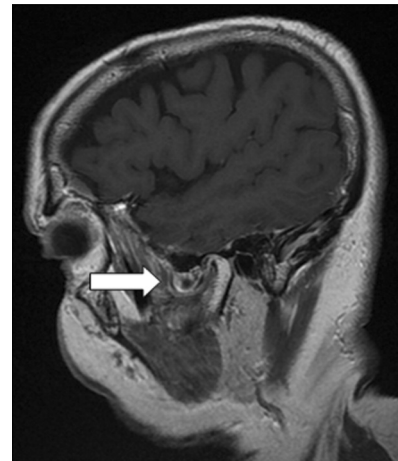


Figure 2. MRI sagittal view showing nodular images in the inferior compartment with a little involvement in the superior compartment even (arrow).

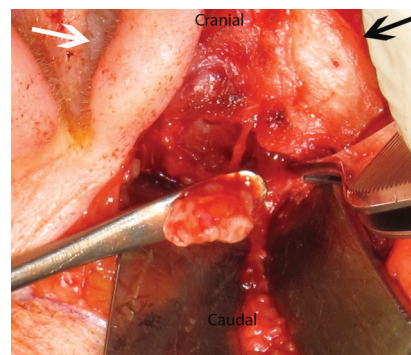


Figure 3. Intraoperative view of the preauricular approach of the TMJ region during the removal of the loose bodies (White arrow – external meatus, Black arrow – Zygomatic arch).

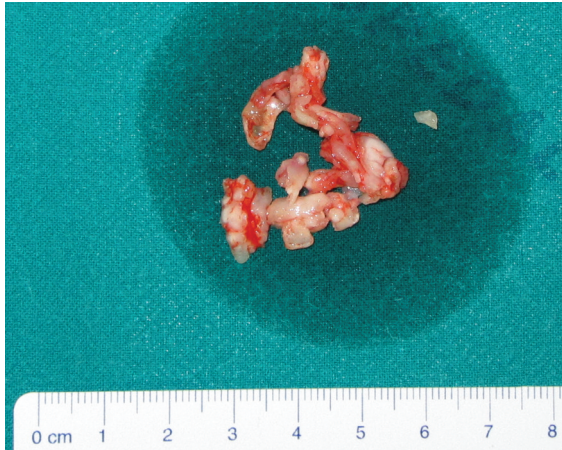


Figure 4. Macroscopic image of nodules of variable size from the lower compartment.

After careful analysis and macroscopic description, the surgical specimens were fixed in 4% buffered formalin and paraffin-embedded.

Subsequently, four-micron hematoxylin and eosin-stained sections of tumor were examined microscopically.

Diagnostic criteria for SC included lobulated chondroid proliferation with foci of hypercellularity and calcification within synovium and no underlying arthritis. Microscopically, our case presents nodules of hyaline cartilage lining by flattened synovial epithelium.

These nodules were composed by clusters of chondrocytes and separated by a cartilaginous matrix. The chondrocytes showed nuclear hyperchromasia, enlargement and binuclear forms (Fig 5a-b).

According to the radiological and histopathological aspects, the SC in this patient was classified as a phase III in Milgram's classification, as a stage II in Gerard's classification and as a stage I in Chen's classification.

One week after surgery the patient started physiotherapy for 45 days in order to improve and increase mouth opening and movement capability.

The CT was repeated at 6, 12 months and 24 months after the operation and confirmed the total removal of the tumor without any recurrence during the follow up period (Fig. 6 a-b). The patient was able to open her mouth to 35 mm with no deviation even though she reported slight pain during mandibular movement.

The follow up period is currently 36 months.

Discussion

Synovial chondromatosis (SC) is a rare disease characterized by the formation of nodules of metaplastic cartilage under the surface of synovial membranes, joints, tendons and bursae.

It was first described according to Ginaldi in 1558 by Ambrosio Pare (20); in 1764 Albrecht von Haller showed the first report in *Elemental physiologiae corporis humani* (21) and Axhausen in 1933 composed the first detailed scientific work (22).

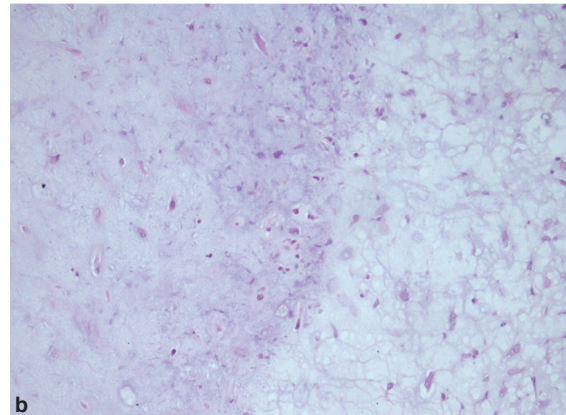
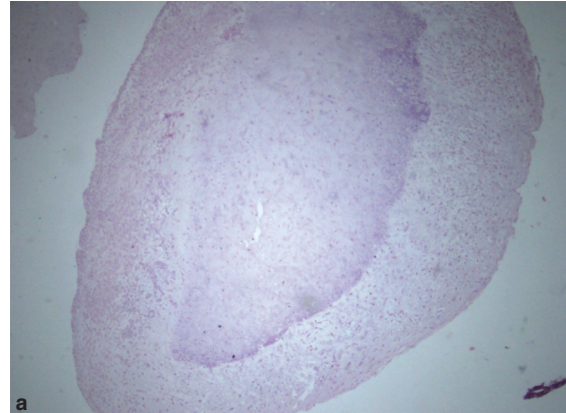


Figure 5. (a) Photomicrograph of a typical loose body of hypercellular cartilage covered with a layer of synovium (EE, 40x). (b) High power view of lesion shows variable areas of atypia and hypercellularity (EE, 200x).

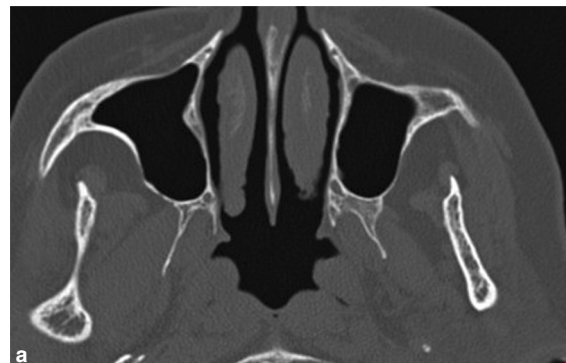


Figure 6 a-b. CT axial and coronal view confirm the total removal of the tumor with no recurrence (24 months).

Afterwards, descriptions of this pathology have been characterized by discrepancies of nomenclature.

The aetiology of SC is not clearly recognized but the presence of clonal chromosomal aberrations suggests the neoplastic nature of this condition (23). Cases are divided into two categories: primary SC, without specific aetiological factors that represent an active cartilaginous metaplasia originating in the synovial membrane, and secondary SC related to previous trauma, repetitive microtrauma and degenerative arthritis or other arthropathies where a less cellular atypia was found (24).

Primary SC is deemed more aggressive (3).

The most common signs and symptoms that we found in the SC of the inferior compartment of the TMJ were: pain, swelling, limited mouth opening, joint crepitus and malocclusion. These symptoms were nonspecific.

The differential diagnosis includes chondrosarcoma, osteoarthritis, rheumatoid arthritis, avascular necrosis (25), osteochondritis, intracapsular condylar fractures (24).

The type of imaging techniques used for diagnosis and surgical planning were CT scan and MRI.

CT scan findings consist in: multiple, calcified loose bodies in the joint space, widening of the joint space, erosion of the glenoid fossa, bony changes of the skull base and condyle head (26, 27).

The MRI found: intra-articular loose bodies present as small areas of low signal intensity, massive expansion of the joint capsule, fluid accumulation within the joint space and an irregular articular disc (27).

The histopathological aspects are represented macroscopically by cartilaginous bodies that remain confined to the synovial membrane that may present as diffusely thickened with a cobblestone appearance or may break off and protrude into the joint cavity as loose bodies. They may be large or small, cartilaginous, osseous or both, ranging in number from a few to hundreds. Microscopically there are several histopathological differences between primary and secondary forms of SC (28, 29).

Primary SC presents alterations such as crowding and irregularity of the cell nests, varying degrees of atypia of chondrocytes with plumped and binucleated elements, which do not necessarily indicate a malignant transformation.

Although the same increased cellularity occurs in secondary SC, it is uniformly distributed with more organized cellular growth; chondrocytes also appear regular and homogeneous.

In addition, a further difference lies in the histological pattern of calcification: the primary form is patchy and diffuse whereas the secondary form is zonal with "ringlike" aspects.

A distinctive trait of secondary SC is the presence of normal articular cartilage fragments with subchondral bone. Areas of endochondral ossification are visible in both metaplastic forms, in some cases complicating their distinction.

In 1977, Milgram (18) categorized the disease process into 3 distinct phases where the relationship

between the cartilage foci and the synovium is placed, while Gerard's classification (19) is based on the synovial activity of the disease where 4 stages can be recognized.

An aggressive surgical treatment in the early stages is recommended both in Milgram's and Gerard's classification.

The most recent clinical-radiological classification was made by Chen et al. according to the different structures involved. They classified SC in the inferior compartment of the TMJ into 3 stages (17).

As above mentioned, different stages point out that different structure are engaged; consequently different stages was supposed to have different principles of management (5).

Guidelines for surgical treatment in these patients now derive from this latest and highly important classification; so that in the first stage we can only remove both the loose bodies and the affected synovium, in the second stage we could perform condilectomy, in the third stage we could perform a condilectomy and discectomy. The reconstruction in the stage two and three could be made with costochondral graft or/and a pedicled deep temporal fascia fat flap (17).

As previously revealed in our case report we found multiple, calcified loose bodies in the inferior compartment with a normal contour of the right condyle. Thus the removal of the loose bodies and affected synovium is generally enough, as Chen's classification state; according to the radiological and clinical findings we perform this type of treatment in our patient with a preauricular approach.

No recurrence has been observed in the current 36-month-follow up period.

Open surgery has always been proposed as the therapy of choice even though some Authors support a less invasive procedure such as arthroscopy. This latter option is very difficult to carry out due to the position of the disease (2).

Conclusion

The case that we reported is the twelfth described in the literature. The treatment performed by us, after a thorough evaluation of the literature, has proved effective with low morbidity.

In all the cases treated for SC in the lower compartment, both in literature and in our case report, no recurrence has occurred so far.

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