



Contents lists available at ScienceDirect

## Journal of Cranio-Maxillo-Facial Surgery

journal homepage: [www.jcmfs.com](http://www.jcmfs.com)

# Synovial chondromatosis of the temporomandibular joint with glenoid fossa erosion: Disk preservation for spontaneous anatomical recovery

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## ARTICLE INFO

### Article history:

Paper received 8 May 2019

Accepted 15 October 2019

Available online 23 October 2019

### Keywords:

Temporomandibular joint disorder

TMJ surgery

Synovial chondromatosis

Intracranial extension

Disk preservation

Pseudoneoplastic metaplasia

## ABSTRACT

Synovial chondromatosis (SC) of the temporomandibular joint is a pseudoneoplastic condition characterized by benign cartilaginous metaplasia of synovial tissue mesenchymal residues with intra-articular nodule formation. TMJ involvement is rare. Interposition of loose bodies in the articular space can generate pressure, leading to glenoid fossa erosion with intracranial extension.

The aim of this study was to present six SC cases with intracranial extension treated using a surgical procedure.

All the patients were treated with open surgery. The superior compartment of the TMJ was opened widely to carefully remove the metaplastic mass. Temporal synovectomy was then performed. Attention was paid to preserving the integrity of the articular disc. The exposed dura mater was also preserved. No material was used to reconstruct the gap in the glenoid fossa.

A 1-year follow-up showed no swelling or pain. Patients demonstrated good recovery of mouth opening, with improvement over previous mouth limitations. Morphological studies, performed using MRI and CT, showed complete anatomical recovery of the TMJ and total bone reconstruction of the glenoid fossa.

Simple removal of intra-articular nodules, with TMJ arthroplasty and articular disk preservation, represents an efficient treatment option for full anatomical and functional recovery in synovial chondromatosis of the temporomandibular joint with glenoid fossa erosion of less than 1 cm<sup>2</sup>.

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## 1. Introduction

Synovial chondromatosis (SC) is a chronic pseudoneoplastic endoarticular condition characterized by benign cartilaginous metaplasia of mesenchymal residues (Campbell et al., 2011). These residues originate from synovial tissue and usually form nodules that occupy the intra-articular space (Marín Fernández et al., 2013). Nodules are found both as floating loose bodies or attached to the synovial membrane, with different grades of calcification (Milgram, 1977). Temporomandibular joint (TMJ) involvement is rare — the most affected joints are the elbow, knee, shoulder, and hip (Khanna

and Ramaswami, 2017). SC etiology is still unknown, but various theories have been considered: primary chondromatosis is due to the permanence of multipotent, undifferentiated cells, which undergo a metaplastic process; secondary chondromatosis originates from traumas, articular dysfunctions, and degenerative arthritis (Lieger et al., 2007).

TMJ chondromatosis usually presents itself with a trio of symptoms: jaw movement limitation with episodic blocks, swelling, and pain (Guarda-Nardini et al., 2010). But there are other manifestations, such as malocclusion, crepitation, deviation to the affected side, mouth opening restriction, and neurological signs and symptoms due to facial and trigeminal nerve involvement (Lieger et al., 2007; Yokota et al., 2008; Shah et al., 2011; McCaffery et al., 2017). Since the clinical presentation shares characteristics with other TMJ diseases, diagnosis relies on imaging and

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histological examination. Surgery represents the treatment of choice for minimizing the possibility of relapse (Shah et al., 2011).

The interposition of loose bodies in the intra-articular space can lead to glenoid fossa erosion with possible intracranial extension; in particular, it can lead to the deterioration of the middle cranial fossa floor (Khanna and Ramaswami, 2017).

The aim of this study was to evaluate six patients with TMJ SC and intracranial extension treated in the Maxillo-facial Department of the 'Sapienza' University of Rome.

## 2. Case report

From 2009 to 2018, 16 patients were treated for TMJ SC. Data were collected from the Department database, taking into account name, sex, gender, previous medical history, signs and symptoms, clinical examination, and radiological findings. Inclusion criteria were: CT showing glenoid fossa erosion and deterioration (Fig. 1), MRI (Fig. 2), histological confirmation, and a follow-up of at least 1 year. Only six (37.5%) of the 16 cases fulfilled these criteria and were enrolled. 10 (62.5%) of the 16 were excluded because they did not have intracranial extension visible on CT.

Every patient underwent surgery by the same surgeon (P.C.), using the same technique.

Surgical operation consisted of a pre-auricular incision performed with a 45° inclination. The deep temporal fascia was reached by blunt dissection, visualizing the articular capsule below. The temporal vessels were then isolated and cut. The TMJ capsule was accessed and its lateral side widely exposed in order to surgically access the superior compartment.

Once access to the superior compartment was obtained, the lateral ligament was identified, and masses were removed whilst carefully preserving the articular disk. Temporal synovectomy was performed and the medial region was investigated and cleaned with arthroscopy. The dura mater was examined for possible infiltrations. All the patients treated had a dura mater exposure of about 1 cm<sup>2</sup>. Nodules were sent for histological examination (Fig. 3).

Age, sex, gender, and affected side distribution were analyzed, as well as the onset of clinical manifestations and the evolution of

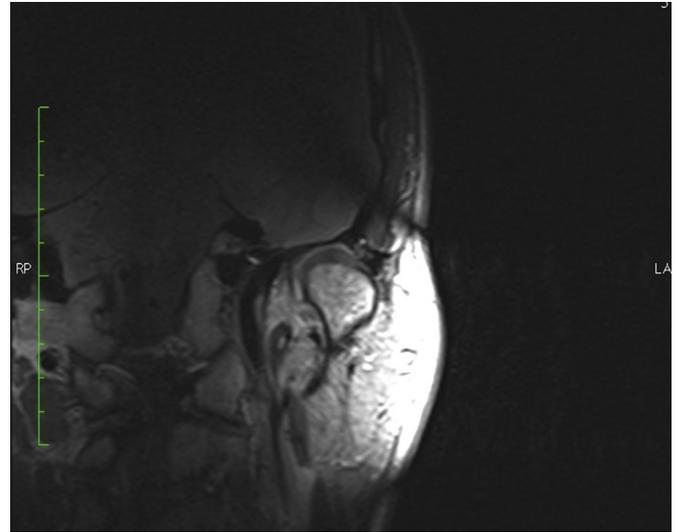


Fig. 2. MRI of the left TMJ with antero-medial disk dislocation and presence of a hyperintense nodule laterally.

signs (malocclusion and asymmetry of the lower third of the face), pre- and postoperatively. A comparison between our data and those in the literature was then performed.

The mean age of patients was 48.6 years old, with a range from 30 to 61. Females were more affected than males, with a ratio of 4/6 (67%). The most frequent side of nodule formation was the left (4/6; 67%). Clinically, our patients reported pain (6/6; 100%), swelling (5/6; 83%), limitation of jaw movements (5/6; 83%), and crepitation (3/6 50%). Deviation to the opposite side from the disease was seen in three patients (3/6; 50%). Half of the patients experienced intermittent lock (3/6; 50%).

Five of the cases reported the onset of symptoms, on average, 4 years (range 3–5) before the first visit. Patient 3 had a short medical history, with only 1 month of symptoms before the first visit (Table 1).

Patients were evaluated 1 year after the surgery using clinical, functional, and instrumental examination. Morphological studies, performed using MRI and CT, showed complete anatomical recovery of the TMJ and total bone regeneration of the glenoid fossa (Fig. 4).



Fig. 1. Erosion of the right glenoid fossa in coronal view.



Fig. 3. SC nodules after tumor enucleation.

**Table 1**  
Characteristics of the study population.

PN	Sex	TMJ	Age	Years after onset of symptoms	Pain	JML	SWE	CRE	Lock	Associated muscular pain
1	M	L	30	3	x		x	x		
2	M	L	52	5	x	x	x			
3	F	R	50	1 month	x	x			x	x (neck and arms)
4	F	R	61	5	x	x	x		x	
5	F	L	49	2.5	x	x	x	x		
6	F	L	50	3	x	x	x	x	x	

Abbreviations: PN patient number, TMJ temporomandibular joint, JML jaw movement limitation, SWE swelling, CRE crepitus.

Clinical follow-ups showed the absence of swelling, pain, and jaw blocking. Patients demonstrated good recovery of mouth opening and improvements to previous limitations in jaw movement.

### 3. Discussion

The first reported case of TMJ SC was described by Baron Albrecht von Haller in 1764 (Khanna and Ramaswami, 2017). In 1933 Axhausen first described the SC histological presentation (Lieger et al., 2007). Loose bodies originate from synovial residues, which gradually grow into nodules and detach from the articular surface. This leads to friction between the nodules and the glenoid fossa. The process continues, with consequent mechanical bone erosion. When this happens, loose bodies come into contact with the dura mater, leading to 'intracranial extension' (Pau et al., 2014).

SC of the TMJ is a predominantly unilateral condition, with a female prevalence and an age range from 39 to 55 at the point of diagnosis. This trend was confirmed in our patients — all cases were unilateral, the percentage of females was higher, and the ages ranged from 30 to 61 (McCaffery et al., 2017). Only one bilateral chondromatosis case was found in the literature (Guarda-Nardini et al., 2010) (Pau et al., 2014), while only 16 (6%) of 260 cases of TMJ SC described in the literature were intracranial.

It is possible to distinguish two forms of chondromatosis, primary and secondary: primary chondromatosis is based on metaplasia of synovial membrane cells; secondary chondromatosis originates from temporomandibular disorders. Primary SC shows a more aggressive behavior (Lieger et al., 2007).

The primary process originates in the synovial membrane, with outbreaks caused by multipotent, undifferentiated cells undergoing cartilaginous metaplasia (Campbell et al., 2011). The nodules formed by these groups of cells then become detached to form loose bodies. The number of nodules present at the moment of enucleation varies greatly because of their different dimensions and their tendency to fuse with each other. Cell populations involved in this condition are predominantly chondroblasts and chondrocytes, but they can often develop as osteoblasts, as seen in some calcified loose bodies (Yoshitake et al., 2016). This aspect helps to enable image-based diagnosis using CT.

When the SC has a secondary origin, the metaplasia originates from pieces of synovial tissue detached from the membrane as a result of primitive organic processes. Causes include traumas of the TMJ, articular dysfunctions, degenerative arthritis, and inflammatory diseases (Shah et al., 2011).

Typical microscopic findings are represented by hard hyaline cartilaginous tissue that is focally and densely cellulated, with chondrocytes at different stages of metaplasia (Holmlund et al., 2003). It is possible to find cells with multiple nuclei and other atypical features such as pleomorphism and nuclear enlargement (McCaffery et al., 2017). Malignant transformation of the disease is very rare, but cases of SC of the knee joint developing into

chondrosarcoma have been found in the literature (Pau et al., 2014; McCaffery et al., 2017).

In 1977 Milgram proposed a disease activity pathological classification comprising three stages. The first, early stage is characterized by the formation of nodules, with only intrasynovial metaplasia and protruded — but not detached — bodies. In the second, transitional stage metaplasia begin to produce loose bodies, which occupy the joint space. The final stage represents the phase in which metaplasia become inactive, and multiple, loose bodies fill the articular space (Milgram, 1977). In this stage it is possible to find secondary calcification known as Henderson–Jones syndrome (Khanna and Ramaswami, 2017). Intraoperative or postoperative histopathological examination, performed using an open or arthroscopic approach, provides confirmation of the synovial chondromatosis diagnosis. Microscopic observation and immunohistochemical tests allow the diagnosis of benign metaplasia. The most used antigens are the proliferating cell nuclear antigen (PCNA) and Ki-67 (Campbell et al., 2011; Pau et al., 2014; Yoshitake et al., 2016). Both are important for detecting the activity of the disease at the moment of the diagnosis: PCNA is usually high in the proliferation stages (Milgram stages 1 and 2); Ki-67 is commonly found low (Yoshitake et al., 2016). For these reasons histology allows a differential diagnosis from other pathologies, such as osteochondromatosis, chondroblastoma, pigmented villonodular synovitis,



**Fig. 4.** CT of the previous patient at 1-year follow-up.

chondrocalcinosis, and rheumatoid arthritis, all of which could be misdiagnosed as SC (Lieber et al., 2007; Shah et al., 2011).

TMJ synovial chondromatosis manifests itself in a non-specific way — there are no pathognomonic signs. Symptoms are usually represented by jaw movement limitation with episodic blocks, swelling, and pain (Guarda-Nardini et al., 2010). Other manifestations include secondary malocclusion, crepitation, deviation to the affected side, asymmetry of the lower third of the face, mouth opening restriction, and neurological signs and symptoms (pain and paralysis) due to facial and/or trigeminal nerve involvement (Lieber et al., 2007; Campbell et al., 2011; Shah et al., 2011; Pau et al., 2014; Khanna and Ramaswami, 2017; McCaffery et al., 2017). Occasionally, there are ipsilateral hearing disturbances caused by Eustachian tube invasion (Yokota et al., 2008). An absence of clinical evidence can lead to a delay in diagnosis or a misdiagnosis of SC (Lieber et al., 2007; Pau et al., 2014; McCaffery et al., 2017).

A relapse-remitting trend is often reported by patients with SC. A long delay from the beginning of the metaplastic process to a diagnosis is supported by the literature, with 80% of the published cases being at Milgram stage 3 (Shah et al., 2011).

The lack of clinical specificity is addressed using CT and MRI imaging. In the intracranial extension, high-resolution CT allows examination of glenoid fossa erosion, and involvement of the articular joint and middle cranial fossa (Khanna and Ramaswami, 2017; McCaffery et al., 2017). MRI allows evaluation of articular fluid collection and dura mater involvement, in order to complete the morphological examination (Shah et al., 2011). Histological examination then provides certainty of SC, clarifies the benign behavior of the disease, and so defines the surgical treatment (Holmlund et al., 2003). The arthroscopic approach is useful in the early stages with small, loose bodies, but can be limited by different-sized nodules (McCain and de la Rua, 1989).

It is common for SC diagnosis to occur at an advanced stage, characterized by large, loose bodies, when an arthroscopic approach would be less effective (Lee et al., 2019). For this reason, open surgery is necessary for the removal of large nodules, and is reported for all cases with intracranial extension (Pau et al., 2014).

Various surgical techniques are reported in the literature for treatment of SC of the TMJ. Access is usually achieved via pre-auricular incision, with possible variation — such as the extension of the cut — to better expose the structures involved. The Al-Kayat and Bramley incision with a retrotragal approach (Pau et al., 2014) was used by Shishir and Pau. In five of 16 cases, exposition of the TMJ was performed with osteotomy of the zygomatic arch (Yokota et al., 2008; Campbell et al., 2011; Pau et al., 2014; Khanna and Ramaswami, 2017; McCaffery et al., 2017). The procedure of choice involves joint cleaning with tumor enucleation and associated synovectomy, as proposed by most authors (Lieber et al., 2007; Mercuri, 2008; Yokota et al., 2008; Guarda-Nardini et al., 2010; Campbell et al., 2011; Shah et al., 2011; Marín Fernández et al., 2013; Pau et al., 2014; Milne et al., 2016). Synovectomy has a central role in the surgical approach for TMJ SC and should always be performed to reduce the possibility of recurrence (McCaffery et al., 2017). Milgram et al. propose not performing synovectomy in the third stage because of the low disease activity and recurrence rate (Milgram, 1977).

Depending on the patient's joint stability, the amount of erosion, and the Milgram stage, additional procedures like condylectomy and/or discectomy can be performed to achieve disease resolution and recovery of joint functionality (Mercuri, 2008).

The most common techniques used for glenoid fossa reconstruction are stabilization with a titanium mesh (Pau et al., 2014) and a temporal muscle flap to prevent articular ankylosis (Shah et al., 2011). Interposition of the muscle flap has been proposed

to avoid bone-to-bone contact that does not allow regeneration of the involved surface. In six of 16 cases, a temporal muscle flap was used (Campbell et al., 2011; Shah et al., 2011; Pau et al., 2014; Khanna and Ramaswami, 2017; McCaffery et al., 2017); in only one case (Milne et al., 2016), the glenoid fossa defect was corrected using bone picked from craniotomy.

The surgeon in our study decided to perform an arthroplasty with synovectomy to achieve a double goal — removal of the metaplastic mass and a reduction in the possibility of recurrence. When glenoid fossa erosion is no more than 1 cm<sup>2</sup> reconstruction may not be necessary — rather, just tumor enucleation, due to its frictional action. After the removal of free bodies and the consequent reduction in 'roof' pressure, the glenoid fossa bone can regenerate spontaneously. The removal of pathogenic noxa stops the erosion phenomenon and promotes new bone formation. In all six cases presented, due to its benign nature, metaplastic lesion never infiltrated the dura mater. In all the intracranial cases reported in the literature the mass always remained extradural (Pau et al., 2014). The absence of macroscopic infiltration during TMJ surgery underlines the importance of the preservation of all the anatomical structures affected. Once the tumor has been removed, the balance of the joint returns to normality and the bone of the glenoid fossa regenerates without the interposition of material inside its intracranial extension. Our experience leads us to believe that respecting TMJ anatomy and physiology leads to a faster functional recovery and allows for the complete ossification of the missing bone.

#### 4. Conclusion

SC is an uncommon TMJ disease and its intracranial extension is rare. This statement is supported by the presence of glenoid fossa erosion in only 16 cases in the literature and in six patients treated over 9 years of clinical practice in our unit.

This study proposes a more conservative approach, underlying the importance of disk preservation. Approaching the glenoid fossa erosion without the use of natural or artificial materials to fill it, represents a valid method for achieving complete glenoid fossa regeneration: removing the cause of the friction is the key to enabling the bone's self-reconstruction. Preserving the disk allows the joint to recover its function earlier.

All treated patients regained normal articular mobility, glenoid fossa erosion was repaired with new bone tissue, and there was no recurrence at 1-year follow-up. The removal of pathogenic noxa, whilst preserving articular structures, represents a valid surgical treatment for TMJ SC with intracranial extension.

#### Ethical approval

Not required.

#### Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

#### Declaration of Competing Interest

None of the authors has any possible conflict of interest.

#### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.jcms.2019.10.005>.

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