

# Synovial Chondromatosis of the Temporomandibular Joint: An Imaging Perspective

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## Abstract

Synovial chondromatosis (SC) is a rare, benign, chronic, progressive and proliferative lesion that more often affects the large joints of the body. There is formation of cartilaginous nodules in the synovial joint space, which subsequently degrade, detach and form free-floating, calcified bodies within the joint space. The first case of SC affecting the TMJ was described by Axhausen in 1933. The etiopathogenesis still remains unclear but often associated with a history of trauma. Clinically patients present with symptoms of swelling, pain, crepitation, malocclusion and joint dysfunction. These signs and symptoms mimic other TMJ diseases thus establishing the importance of correct diagnosis of SC. Here we present a case of SC of the right TMJ highlighting the various imaging modalities which aid in the diagnosis of SC.

**Keywords:** Synovial chondromatosis, Temporomandibular joint, Calcified bodies

## Introduction

SC is a rare, benign, chronic, progressive and proliferative disease that usually affects large joints.<sup>[1]</sup> It is characterised by the formation of cartilaginous nodules within the articular space.<sup>[2]</sup> SC was first described by Ambroise Pare and later a detailed scientific description of the same was given by Axhausen.<sup>[3,4]</sup> SC is defined by the World Health Organization as a benign nodular cartilaginous proliferation arising from the joint synovium, bursae or tendon sheaths.<sup>[5]</sup>

The incidence of occurrence of SC is 1:100,000.<sup>[6]</sup> It affects the diarthroidal joints of axial skeleton like knee (35%), elbow (22%), wrist (11%), hip (4%) frequently and TMJ is affected very rarely.<sup>[7]</sup> It has been reported to affect the right TMJ more than left. Majority of the cases are monoarticular although bilateral involvement have also been reported.<sup>[3]</sup> The etiology of SC is unknown but it may be caused due to trauma and inflammation.<sup>[1]</sup> When involving the TMJ SC is more common in women in the ratio of 1.5:1 in the fourth and fifth decade of life.<sup>[2]</sup> The typical signs and symptoms of SC are preauricular swelling, pain, crepitation, clicking and and limitation of mandibular movement.<sup>[7]</sup> The objective of this article is to present a case of SC of the right TMJ, highlighting the various imaging modalities which aid in the diagnosis of SC.

## Case Report

A 38-year-old male patient came to our department with the history of pain in the right preauricular region on having food and on mouth opening since 7-8 years. The pain was insidious in onset, mild in intensity and aggravated on having food and on mouth opening. No history of trauma or any other joint problems. No significant medical and family history was recorded. On extra-oral examination mild, facial asymmetry

was noted on the right aspect of the face [Figure 1A] with a swelling in the preauricular region measuring 1 cm in greatest dimension [Figure 1B]. The surface over the swelling appeared normal.

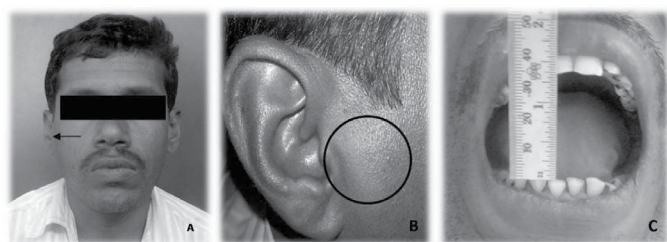
TMJ examination-On preauricular and intraauricular palpation no clicking or crepitus was noted. No deviation of mandible noted on mouth opening/closing. Maximum mouth opening noted was 35 mm [Figure 1C]. Masticatory muscles were non-tender on palpation. On intra-oral examination midline shift was noted.

Based on the history and clinical presentation the provisional diagnosis of right condylar hyperplasia was given. A differential diagnosis of condylar tumor was given taking into consideration the chronicity and severe increase in pain on mouth opening. Radiological investigations included OPG and CBCT were advised.

The OPG [Figure 2] of the right TMJ showed an increase in the joint space with mild erosive changes of the condylar head. As this was inconclusive a CBCT TMJ was advised. CBCT showed presence of faint opacities in the medial, anterior, posterior and superior aspect of condyle which probably represented extraarticular calcifications [Figure 3]. Since these calcifications were not clearly visible in the CBCT therefore a CT was advised.

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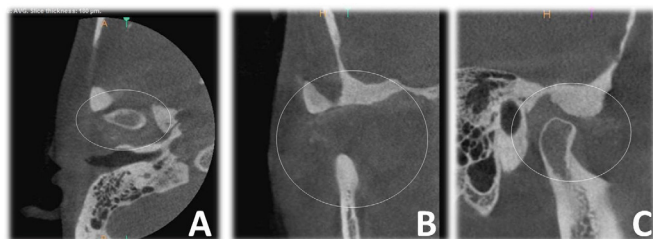
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**Figure 1:** A. Frontal view showing mild facial asymmetry. B. Swelling in the preauricular region. C. Maximum mouth opening (35 mm).



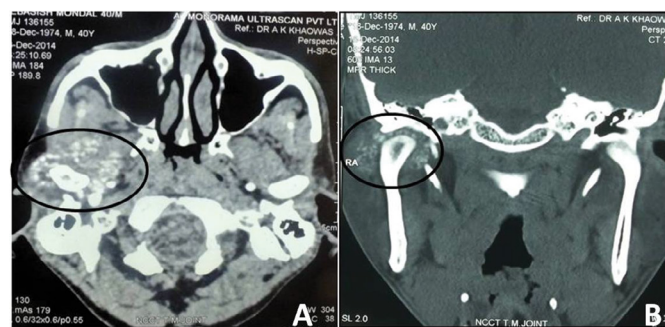
**Figure 2:** OPG-showing increased joint space in the right TMJ.



**Figure 3:** CBCT-A. Axial view. B. Coronal view. C. Sagittal view.

Axial CT of the right TMJ shows multiple calcified loose bodies in the medial, lateral, anterior and posterolateral aspect of the condyle within a low attenuated soft tissue mass [Figure 4A]. In the coronal section, there was an increase in the joint space in the right TMJ along with calcified loose bodies in the medial, lateral and superior aspect of the condyle [Figure 4B]. After CT evaluation, a radiographic diagnosis of SC of the right TMJ was given.

Radiographic differential diagnosis of osteoarthritis, chondrocalcinosis and osteochondritis dissecans were considered. Osteoarthritis is an age-related degenerative disease, common in women which shows osteophyte formation, erosion, flattening, sclerosis of mandibular fossa and reduction in joint space. Although osteoarthritis may affect the joint space, the calcifications (joint mice) are smaller as compared to those in our case. Calcium pyrophosphate dihydrate disease (CPDD), or pseudogout, is a type of arthritis caused by the precipitation of calcium pyrophosphate crystals. It is considered a disease of older patients. The radiographic appearance of CPDD is called chondrocalcinosis, which resembles SC histologically and radiographically. However, CPPD of the TMJ is rare and usually associated with predisposing factors. There were no predisposing factors evident in our patient, nor did he show any sign of a metabolic disease (gout). Osteochondritis dissecans, a reactive joint condition can cause focal calcifications.



**Figure 4:** CT-A. Axial view. B. Coronal view.

Excision of tumor under general anaesthesia with oro-endotracheal intubation was planned. Following Alkayat Bramley incision, layered dissection was done to expose the tumor mass which was well encapsulated and situated in the capsule. Tumor mass composed of cartilaginous pebbles like structure, was removed from the anterior, superior and lateral aspect of the condyle. Gap titanium plate with 2mm hole was adapted on the lateral surface of the condyle. Subsequently, osteotomy of the neck of the condyle was performed. Then the condyle was swung to gain approach to the medial aspect of the condyle and calcified bodies were removed. Osteotomy was necessary for the approach to the medial aspect of the condyle, regardless of a normal mouth opening. Later the articular disc was repositioned and sutured. The condylar neck was fixed with miniplates and screws.

The gross pathologic appearance showed hyperplastic synovium covering white multilobulated nodular projections of hyaline cartilage [Figure 5]. Histopathology of the same revealed globular masses of cartilage tissue which exhibits areas of calcification. Chondrocytes are showing mild atypia in the lacunae. The synovial connective tissue in between the cartilage is delicate and fibrillar. The peripheries of the globular masses are cellular with numerous blood vessels [Figure 6].

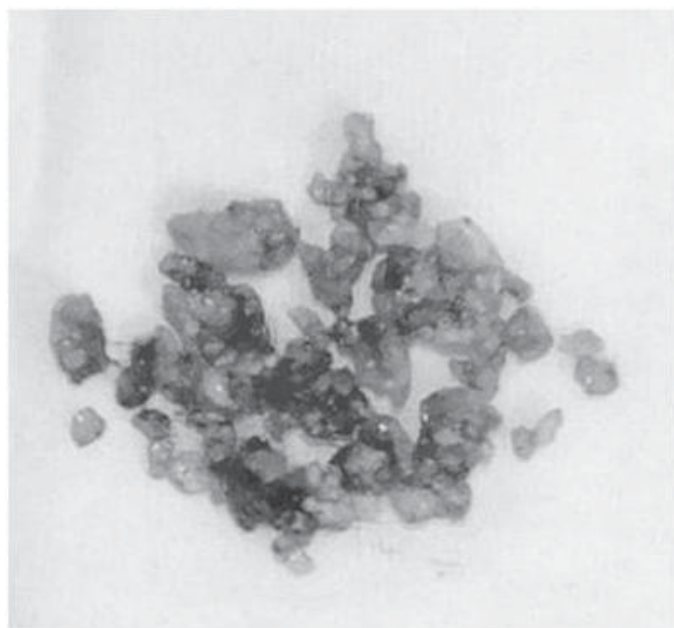
Based on the clinical, radiological and histopathological findings the diagnosis of SC of right TMJ was given. Patient was kept under regular follow up. Figure 7 shows the postoperative OPG of the patient.

## Discussion

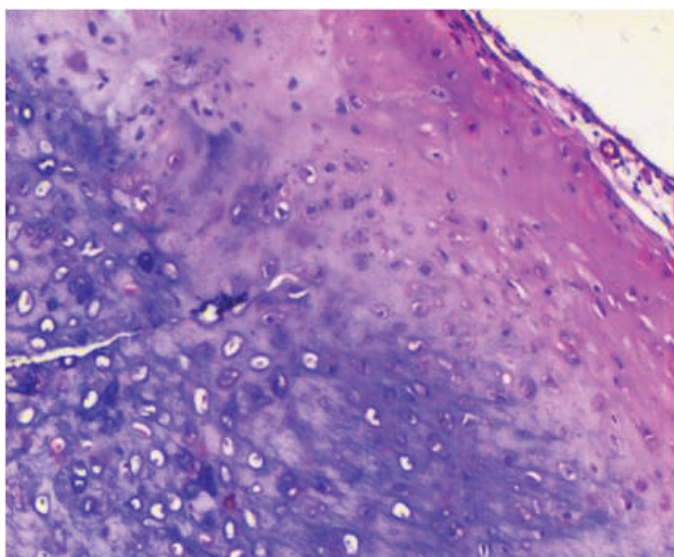
SC TMJ is a rare chronic arthropathic condition which is generally monoarticular. Trauma, inflammatory conditions, degenerative diseases and infections can act as contributing factors.<sup>[4]</sup>

Pathogenesis of SC presents with the chondrometaplasia of the synovial membrane, through the fibroblastic growth factor receptor 3 (FGFR3). Also seen are immunoreactions against the anti-transformer of growth factor beta (TGF- $\beta$ ) on the surface of the synovial membrane and to tenascin (TN) in the extracellular matrix. TGF  $\beta$  is known to cause an increase in the production of proteoglycans and formation of chondroblasts by differentiation of mesenchymal cells. Since fibroblastic growth factor 9 (FGF9) in synovial fluid of patients with SC is shown to be high when compared with synovial fluid of normal individual, it can be





**Figure 5:** Gross specimen showing surgically removed calcified bodies.



**Figure 6:** Histopathology showing the features of SC.

concluded that FGFR 3 causes proliferation of mesenchymal cells and chondrogenesis in the extracellular matrix.<sup>[3]</sup>

The signs and symptoms of SC include pain, swelling, limitation of movements of the jaw, clicking and creptation on opening the mouth. In advanced stage of SC there may be presence of cranial nerve dysfunction.<sup>[4]</sup>

Based on the cause, SC can be divided into 2 forms: primary and secondary form. The primary form, doesn't have clear etiology, and may be associated with embryological disorder wherein metaplastic changes in the synovium cause foci of calcification to later degrade, detach and to lie free in the synovial cavity.<sup>[4]</sup> In the primary form fibroblast undergo metaplasia beneath the synovial membrane and there will be deposition chondromucin and this loose bodies enlarge and detach from the synovial membrane.<sup>[7]</sup> The secondary form which is associated with trauma and other arthropathies, has a slower and more chronic presentation. The two forms are difficult to differentiate from



**Figure 7:** Post-operative OPG.

each other, especially the long standing primary form which may predispose to degenerative joint disease conditions.<sup>[4]</sup> However in primary form the mineralised bodies are usually over five in number and equal sized where as in secondary form there are fewer bodies of variable size.<sup>[6]</sup>

Pathologically SC has been described in three stages by Milgram, in the first stage chondromas are attached to the synovium. In the second stage the chondromas are breached out of the synovium and are released as free bodies into the joint capsule. The third stage is reached when the synovial disease becomes inactive but the intraarticular nodules remain which vary in size from less than 1 mm to greater than 10 mm.<sup>[6,8,9]</sup>

SC is a benign condition, nevertheless it has shown the potential for destroying the middle cranial fossa, invading the intracranial structures and in rare instances, a malignant transformation with a prevalence rate of 5%.<sup>[4,10]</sup> Perry, *et al.* have reported malignant transformation of the cartilage with development of chondrosarcoma.<sup>2</sup> Hence an early diagnosis is essential for an appropriate management and better prognosis.<sup>[4,10]</sup> This is possible through radiological evaluation which are usually considered pathognomonic for SC.

## Imaging and Management

Radiographic evaluation is responsible for detecting multiple intraarticular calcification in 70% to 95% cases of SC.<sup>[11]</sup> Noyek *et al.* pointed out radiologic features of SC in the TMJ, namely 1) widening of the joint space, 2) limitation of motion, 3) irregularity of the joint surface, 4) presence of calcified loose bodies (cartilage), and 5) sclerosis or hyperostosis (overgrowth) of the glenoid fossa and mandibular condyle.<sup>[10]</sup> Although many conditions present with loose bodies within the TMJ one notable difference is that the patients with SC have more than 10 loose bodies which are packed in the soft tissue mass giving a typical appearance of *grains of rice in sushi*.<sup>[4]</sup>

In ultrasonography SC is depicted as a heterogenous, avascular mass surrounded by fluid. It appears has hyperechoic foci with acoustic shadowing. Ultrasound is the additional tool rather than genuine diagnostic aid.<sup>[6]</sup> In arthrography it is seen as numerous rounded filling defect which are attached or separated from the synovium.<sup>[6]</sup>

In CBCT, SC is seen as widening of the joint space with irregular or sclerotic glenoid fossa and the presence of multiple, calcified, loose bodies in the joint space. CBCT further facilitates examination of TMJ anatomy thoroughly without

superimposition and distortions, thus permitting analysis of bone morphology and joint space in all three dimensions. However, in our case, these calcifications were not clearly visible in CBCT. Hence to analyze the characteristics of the calcifications, CT was advised for further radiological confirmation. On CT, calcified cartilaginous bodies of SC are easily identified.<sup>[7]</sup> In cases such as ours, where radiographic features on conventional radiographs and CBCT were normal or equivocal due to complex osseous anatomy, CT particularly helped for identifying characteristic ring and arc or punctuate mineralization and multiplicity of nodules.<sup>[6,11]</sup> CT is also helpful in the evaluation of extrinsic erosion of bone due to its cross sectional imaging capabilities.<sup>[11]</sup> The loose bodies usually appear packed within the soft tissues which are more than 10 in number and seen as “rice grains in sushi”.<sup>[4]</sup>

MRI shows a distinctive appearance of joint effusion and intra articular soft tissue mass with internal nodules. This mass is characterised as hypo/isointense to muscle on T1 weighted image and hyperintense on T2 weighted images.<sup>[6,10]</sup> Meng *et al.* described the presence of spherical cartilaginous bodies in a “ring-like” form as a sign of SC TMJ on MRI.<sup>[5]</sup> SC is more aggressive when it is extending till the skull base and intruding into the intracranial structures therefore, MRI is vital in accurately identifying boundaries and extent; and in assessing internal derangement and detecting intracranial extension.<sup>[4]</sup>

SC is a benign disease which doesn't respond to non-surgical treatment and doesn't show spontaneous resolution. Surgical treatment of SC involves either the removal of calcified bodies with or without synovectomy and arthroscopy. Although less invasive, arthroscopy is not optimal for retrieving the intraarticular calcified loose bodies. Therefore, surgical removal of the calcified bodies is the treatment of choice, with regular follow up examination to detect any signs of recurrence or malignant transformation.<sup>[2,4,7]</sup>

## Conclusion

SC of TMJ is a rare benign pathological condition showing chondrometaplasia of the synovial membrane. It should be included in the differential diagnosis in patients with a preauricular pain as well as radiographic presence of heterogeneous mass affecting the TMJ. Clinical symptoms of pain, joint swelling and dysfunction seen in SC overlap

with that of other TMJ diseases. Hence appropriate imaging modality can play a pivotal role in the diagnosis of SC. In the present case, a precise diagnosis couldn't be arrived at using conventional imaging as well as CBCT. Ultimately, CT imaging proved to be a valuable tool in the diagnosis of SC as CT can identify radiologic features which are difficult to evaluate due to complex osseous anatomy of TMJ SC.

## Financial Disclosure Statement:

All the authors have no financial disclosures relevant to current submission.

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