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## CASE REPORT

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# Calcium Pyrophosphate Dihydrate Crystal Deposition Disease of the Temporomandibular Joint

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### ABSTRACT

*Calcium pyrophosphate dihydrate crystal deposition disease of the temporomandibular joint is a rare entity that has a tendency to mimic bone tumour, leading to unnecessary surgery. This report describes a patient with calcium pyrophosphate dihydrate crystal deposition disease in the temporomandibular joint with characteristic computed tomography and histological features.*

*Key Words: Calcium pyrophosphate; Chondrocalcinosis; Temporomandibular joint; Tomography, X-ray computed*

## 中文摘要

### 顳下頷關節的二水焦磷酸鈣晶體沉積病

陳慧儀、陳澤宗、黃發基

顳下頷關節的二水焦磷酸鈣晶體沉積病很罕見，由於症狀與骨腫瘤相似，往往會導致不必要的手術。本文報告一宗顳下頷關節的二水焦磷酸鈣晶體沉積病，病人有典型的電腦斷層掃描及組織學特點。

### INTRODUCTION

Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease is the most common crystal deposition arthropathy. CPPD crystal deposition was first described by Zitnan and Sitaj in 1958.<sup>1</sup> The condition is characterised by accumulation of CPPD crystals in intra-articular and juxta-articular tissues. CPPD crystal deposition of the temporomandibular joint (TMJ) is an uncommon entity that has a tendency to mimic bone tumour, which may significantly alter the management.

### CASE REPORT

A 73-year-old woman presented to the North District Hospital, Hong Kong, in February 2010 with a mass over the left TMJ for 4 months. The mass had gradually enlarged over time until it became static in size. The mass was painful initially, but the pain later subsided spontaneously. The patient was otherwise well, with no symptoms involving any other joints.

On physical examination, a 2-cm bony hard mass was felt over the left TMJ. There was no facial nerve palsy

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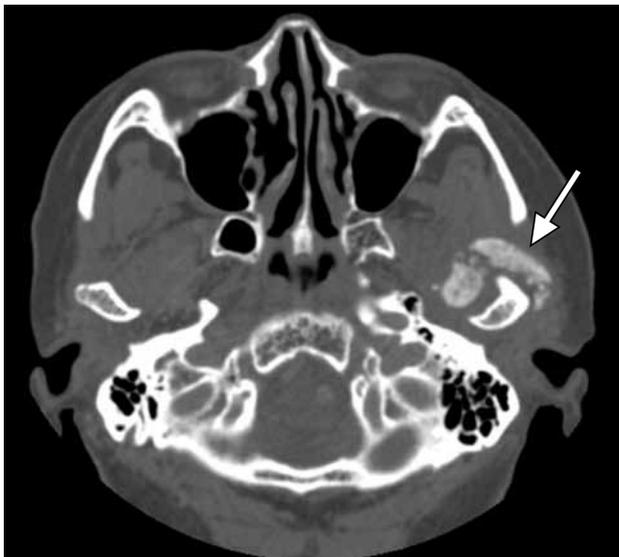
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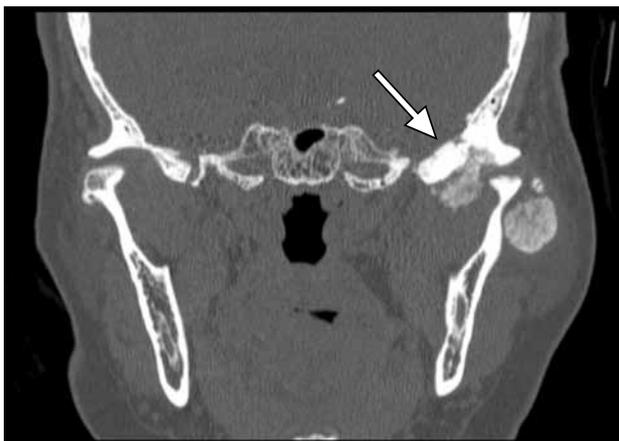
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or cervical lymphadenopathy. The remainder of the physical examination was unremarkable, and the results of laboratory studies were within normal range.

Non-enhanced computed tomography (CT) revealed a calcified mass around the left TMJ (Figure 1). A few bulky amorphous calcifications were present within the mass that were associated with minimal non-calcified soft tissue component. There was erosion of the mandibular fossa of the left TMJ (Figure 2). The TMJ space was preserved. There was no definite marginal osteophytosis or subchondral cystic change. A few foci of calcifications with similar characteristics were present



**Figure 1.** An axial computed tomography scan demonstrating a calcified mass at the left temporomandibular joint with regional involvement to muscles of mastication (arrow).



**Figure 2.** A coronal computed tomography scan demonstrating a calcified mass at the left temporomandibular joint with erosion of the temporal bone (arrow).

within the adjacent muscle of mastication, suggestive of cartilaginous matrix. In view of imaging features of presence of cartilaginous matrix and bony erosion noted on CT, as well as the presence of pain initially, chondrosarcoma could not be excluded. Although CPPD crystal deposition rarely occurs in the TMJ, it also presents as a calcified mass with pressure erosion of the adjacent bone, so was considered as a differential diagnosis. After obtaining the patient's consent, CT-guided biopsy of the mass (Figure 3) was performed with Temno soft tissue biopsy needle, coaxial 18G x 15 cm (Carefusion, McGaw Park [IL], USA). Frozen section and routine histology showed sclerotic fibrous tissue with focal deposition of birefringent crystals in polarised light, which is diagnostic of CPPD crystal deposition disease.

## DISCUSSION

CPPD crystal deposition disease is a metabolic arthropathy associated with calcium pyrophosphate crystal deposition in cartilage and other soft tissue structures. CPPD crystal deposition predominantly affects fibrocartilage, with relatively rare involvement of hyaline cartilage. Frequently, involved sites include the shoulder, hip, elbow, wrist, and knee.<sup>2</sup> Acute and chronic forms have been described.<sup>3</sup> The more common acute form usually manifests as acute knee arthritis with joint effusion. The less common chronic forms are often indistinguishable from osteoarthritis. Tophaceous



**Figure 3.** A computed tomography scan showing guided needle biopsy of the calcified mass at the left temporomandibular joint.

pseudogout, also known as massive CPPD crystal deposition or destructive CPPD crystal deposition arthropathy,<sup>4</sup> is a chronic manifestations characterised by massive crystal deposition in a single joint. Unlike gout, in which massive sodium urate crystals or tophi deposition in soft tissues or joints commonly occurs, massive deposition of crystals in soft tissue is rare in CPPD crystal deposition disease.<sup>5</sup> Only 36 cases of CPPD crystal deposition disease affecting the TMJ have been reported in the literature. The disease develops more commonly in women (n = 24; 66%) than in men, and in patients aged 40 to 85 years (mean age, 58 years). Pain (n = 23; 64%) and periauricular swelling (n = 29; 81%) are the main clinical symptoms.<sup>3-34</sup>

Non-enhanced CT is the imaging study of choice.<sup>29</sup> CT can demonstrate homogeneous distribution of fine granular calcifications and lobulated configuration of the calcific mass near a joint. Radiolucent septi within the mass and erosion and disruption of the adjacent bony cortex, as seen in this patient, had also been described. These features are considered specific for CPPD crystal deposition disease.<sup>35</sup> Initially, there is no invasion into the joint space. With disease progression, involvement of the joint space with degenerative changes of the surrounding bones, including articular space narrowing, osteophytosis and subchondral cyst formation, can also be seen.<sup>29</sup>

Magnetic resonance imaging features of CPPD crystal deposition are rarely described. The calcified masses have intermediate-to-low signal intensity on T2-weighted images<sup>36</sup> and inhomogeneous enhancement on postcontrast T1-weighted images, which is probably caused by granulomatous inflammation due to periarticular crystal deposits. However, T1 and T2 low-signal-intensity periarticular masses are also noted in other cartilaginous diseases such as amyloid, gout, and synovial chondromatosis, along with post-traumatic sequelae, making this modality less helpful in the diagnosis of CPPD crystal deposition disease.<sup>29</sup>

Histologically, the presence of calcium pyrophosphate crystals in biopsy tissue, demonstrated as weakly birefringent with polarised light, is diagnostic. Electron probe microanalysis, scanning electron microscopy, and X-ray diffraction are also helpful in identifying the CPPD crystals.<sup>37</sup>

Diagnosis of CPPD crystal deposition disease in the TMJ is sometimes difficult as the condition has

a tendency to mimic a malignant condition, which leads to unnecessary surgery. Calcified masses may be mistaken for chondroid tumours or other soft tissue tumours.<sup>38</sup> Differentiation from chondrosarcoma is particularly difficult when extensive destruction of the temporal bone is present. Ischida et al<sup>5</sup> reported CPPD crystal deposition at different sites, in which three out of seven cases involved the TMJ, with two of them (66%) misdiagnosed as chondrosarcoma and chondroblastoma. When the diagnosis is doubtful, conventional radiographs or CT of the wrist or knee may contribute to the diagnosis by demonstrating calcium deposition in the menisci (knee) or triangular cartilages (wrist).<sup>29</sup> Histologically, chondroid metaplasia, which is often associated with crystals, can mimic malignancy if it predominates in a biopsy.<sup>33</sup> The use of decalcified sections, from which calcium pyrophosphate crystals are lost, can also lead to a misdiagnosis of chondrosarcoma.<sup>39,40</sup> Frozen sections, which possess more crystals than formalin-fixed sections, should therefore be obtained if possible.<sup>12</sup>

Management of CPPD crystal deposition disease depends on the clinical manifestations. Asymptomatic CPPD crystal deposition disease should not be treated unless it is associated with secondary causes. Treatment is based on prevention of crystal formation, dissolution of crystals, and decreasing the biological consequences of crystal cell interactions. Surgical removal of calcifications from the joint may improve joint mobility, but this is considered an experimental procedure.<sup>29</sup>

In conclusion, CPPD crystal deposition disease should be considered as differential diagnosis of a calcified TMJ mass. Diagnosis of CPPD crystal deposition disease is challenging as it has a tendency to mimic other conditions, particularly chondrosarcoma. Non-enhanced CT is the imaging modality of choice. Frozen sections rather than formalin fixed sections should be obtained, if possible, to improve the accuracy of diagnosis.

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