CASE REPORT

Tophaceous Pseudogout in the Temporomandibular Joint: A case report

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Synopsis
We report an unusual case of tophaceous pseudogout that appeared in the right temporomandibular joint (TMJ) accompanied with painless swelling at periauricular region of a Japanese elderly man. Radiographic examination including computed tomographic images revealed an exophytic radiopaque lesion with clear boundaries in the right peri-condylar region. Surgical resection was employed, and a microscopic examination of the surgical specimen showed the deposition of basophilic rhomboidal crystals around which foreign body reactions including multinucleated giant cells and chondrogenicity were localized. Aggregates of rhomboid crystalline deposits were exposed under polarized light. Further elemental analysis confirmed the involvements of Ca and P and positive birefringence under a polarizing microscope, which helped in the histopathological diagnosis of tophaceous pseudogout.

Key words: tophaceous pseudogout; calcium pyrophosphate dihydrate; pseudogout

Introduction
Calcium pyrophosphate dihydrate (CPPD) deposition disorder is a general term for arthritis characterized in the precipitation of CPPD crystals in joint tissues which causes various pathological symptoms. When there is deposition of sodium urate on the joints, gout develops, but pseudogout is not caused by sodium urate—CPPD crystal is deposited in the synovial fluid showing an acute gout joint-like seizure, which indicates similarity in its pathological condition to those of gout; thus, it is has been termed “pseudogout.” Moreover, pseudogout refers only to CPPD crystal deposits that have acute gout attack-like symptoms [1].

CPPD crystal deposition disorder may be nodular in form; however, there is a rare pathological condition in which crystals are deposited in lump form yet present as a mass or nodule—this is called tophaceous pseudogout, a subtype of CPPD crystal deposition disorder, which is distinguished from false gout [2, 3]. Tophaceous pseudogout is diagnosed when the joints involve a clinically detectable mass lesion showing radiographically evident tumor-like deposition of CPPD crystals [2, 4].

The most common sites of nodular pseudogout are the knee joints of the ankles and wrists, and this type of gout rarely occurs in the temporomandibular joint (TMJ) [5]. In the present study, we describe an unusual case of
tophaceous pseudogout in the TMJ of a Japanese elderly man, and we review previous cases of tophaceous pseudogout.

**Case Report**

A 66-year-old Japanese man was referred to our institution with a 10-year history of painless swelling in the right buccal region. The patient’s family reported no history of the complication; however, the patient had a medical history of hypertension, hyperlipidemia, and encephalorrhagia.

Clinical examination revealed one hard, hemispherical, elastic hard mass at the right temporomandibular joint (TMJ) area. The face was asymmetrical with swelling, but no trismus was observed. The patient had no history of trauma or systemic joint diseases. Additionally, radiographic examination revealed a well-defined exophytic calcified lesion with a clear boundary in the lateral side of the peri-condylar head. Axial and coronal sections of computed tomography (CT) images revealed the tumorous lesion appearing as a ground-glass matrix with the size of $4.2 \times 2.8 \times 3.0$ cm (Fig. 1). A clinical diagnosis of synovial chondromatosis was initially suspected as the above findings suggest, and surgical resection by the preauricular approach was administered under general anesthesia. During surgery, a significant amount of calcareous material, described as chalky or “grits-like,” was noted. Hematoxylin-eosin staining of the surgical specimen showed the deposition of basophilic rhomboidal crystals around the site where foreign body reactions including multinucleated giant cells and chondrogenicity were localized (Fig. 2). In addition, presence of Ca and P was confirmed by elemental analysis and positive birefringence was observed with a polarizing microscope; moreover, an analysis pattern corresponding to calcium pyrophosphate was obtained (Fig. 3). Finally, histologic diagnosis of tophaceous pseudogout (CPPD) was obtained. Postoperative healing was uneventful, and no locoregional recurrence had been detected 4 years post-surgery.

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*Figure 1  Preoperative radiographic findings*

A: A spherical opaque image was observed in the right temporomandibular joint. Panoramic radiograph revealed a well-defined calcified lesion with smooth outline on the right articulating space, which surrounded the condylar head. B: Axial, sagittal, and coronal of CT images demonstrated a continuous spherical region of high CT value on the right mandibular condyle.
Figure 2  Histochemical findings of tophaceous pseudogout.
Hematoxylin-eosin staining showed the deposition of basophilic rhomboidal crystals around which foreign body reactions including multinucleated giant cells and chondrogenicity were localized (A and B). Aggregates of rhomboid crystalline deposits were noted under polarized light (C and D). Original magnification: 10× (A), 40× (B–D).

Figure 3  Elemental analysis of rhomboid crystalline deposits.
A: A scanning electron microscope image showing quadrangular prism crystals. Scale bar represents 5µm. B and C: Ca and P were precipitated, and an X-ray analysis pattern corresponding to CPPD is shown.
Discussion
We describe CPPD deposition disorder as a general term for arthritis which characterizes precipitated CPPD crystals in joint tissues which lead to various pathological conditions. The deposition of CPPD crystals in the synovial fluid resulted in acute gout joint-like seizures, not sodium urate; this was later known as “pseudogout” mainly due to the periodic attacks of acute synovitis associated with the condition. CPPD crystals are also deposited in joint tissues and surrounding soft tissues, but pseudogout refers only to CPPD crystal deposits that cause acute arthritis and gout attack-like symptoms [1]. Moreover, a previous study noted a condition in which acute joint symptoms were absent due to calcification of the articular cartilage and intervertebral discs [6]. Chondrocalcinosis occurs in the knee joint, and because most of these are CPPD crystals, chondrocalcinosis is used almost synonymously with CPPD deposit disorder [7, 8].

CPPD crystal deposition disorder tends to manifest in nodular form; on the other hand, a rare pathological condition in which crystals are deposited in lump form and present as a mass or nodule is called nodular pseudogout—a subtype of CPPD crystal deposition disorder. When massive, clinically detectable deposition of CPPD crystals is observed around joints, tophaceous pseudogout is a common diagnosis [2, 4].

The lower limbs, especially the toes, are the most common sites of gout, but pseudogout tends to occur in the knees of larger joints, ankles, wrists, elbows, and hips. Moreover, the most common site of nodular pseudogout is the knees, but the TMJ has the least known cases [5]. In the case of the TMJ, there is a higher chance for gout to occur unilaterally rather than developing systemically [5]. Synovial chondromatosis and osteosarcoma are the commonly known forms of differential diseases of tophaceous pseudogout. Radiographic examination is importantly viable in diagnosing tophaceous pseudogout, although CPPD crystal deposition in the TMJ is not accompanied by clear calcification [9-11]. Therefore, a reliable diagnosis is recommended to fully confirm the presence of CPPD crystal deposition. One approach involves collecting CPPD crystals floating in joint synovial fluid and performing pathological diagnosis. In addition, polarization microscopy, X-ray analysis, and electronic analysis are useful methods in identifying CPPD crystals [1]. One also has to consider that CPPD crystal deposits may also be associated with systemic disorders such as hyperparathyroidism and hemochromatosis [12].

In most common cases, swelling and local pain help confirm tophaceous pseudogout. In our case, however, a tumor mass in the TMJ area had been left untreated for more than 10 years due to painless swelling. The patient finally presented to our hospital with a complaint of unfavorable aesthetics. Like other benign tumors, indolent clinical symptoms of tophaceous pseudogout sometimes lead to a delay of diagnosis, similar to our case. The etiology is thought to be due to hereditary, idiopathic, metabolic disease or trauma, but in our case, no obvious etiology was observed; hence, it was considered to be idiopathic.

Since CPPD crystal deposition disease often manifests symptoms very similar to those of temporomandibular disorders such as trismus and joint pain, CPPD crystal deposition disorder is latent in some cases, leading to treatment employed for the latter. Although symptomatic treatment through the administration of analgesics is effectively used as one of the treatment options, surgical treatment is essential for radical treatment and more importantly in improving the aesthetic disorders and articular dysfunctions. Since it is not a neoplastic lesion, it is common to perform an excision procedure confined to the lesion in order to preserve the oral function of the TMJ [12]. Few recurrences have been reported. When the site of tophaceous pseudogout increases, there is a possible spread from the TMJ to the peripheral region including the adjacent skull—so it should be highly advisable to plan surgical treatment early when it is accurately diagnosed.

In the present study, hematoxylin-eosin staining showed the deposition of basophilic rhomboidal crystals around which foreign body reactions including multinucleated giant cells and chondrogenicity were localized. Aggregates of rhomboid crystalline deposits were noted under polarized light. Additionally, Ca and P were confirmed by elemental analysis and positive birefringence was observed by a polarizing microscope, and an analysis pattern corresponding to calcium pyrophosphate was obtained, which finally led to the final histopathological
diagnosis of the patient as having tophaceous pseudogout (CPPD).

Conservative resection was ultimately employed in our case, which resulted in an uneventful recovery without locoregional recurrence over a long-term period of six years. In addition, the patient obtained a favorable outcome without any unfavorable aesthetics or functional impairment. However, due to the conservative resection method, close periodical long-term follow-up might be highly necessary.

**Patient consent**
Written consent was obtained from the patient.

**References**

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