Idiopathic tophaceous calcium pyrophosphate dihydrate deposition disease of the temporomandibular joint in a young female

Calcium pyrophosphate dihydrate (CPPD) deposition disease is a metabolic disease that arises from the accumulation of CPPD crystals in the cartilage or soft tissue of joints. The major clinical manifestations are asymptomatic incidental finding (chondrocalcinosis), acute inflammatory arthritis (pseudogout), chronic degenerative arthritis, and the least common tophaceous nodule formation, such as gouty tophus. It commonly occurs in the knee, wrist, metacarpophalangeal, shoulder, hip, and ankle joints. However, most tophaceous CPPD nodules occur at the temporomandibular joint (TMJ). The clinical and imaging differential diagnoses include bone and cartilage tumors. CPPD deposition disease is strongly associated with patients of advanced age and is extremely rare in younger individuals. When it occurs in patients younger than 55 years, a comprehensive metabolic workup as well as clinical and family history should be taken to exclude secondary diseases. The possible secondary etiologies include rheumatoid arthritis, gouty arthritis, hemochromatosis, hypoparathyroidism, hypophosphatasia, hypomagnesemia, Wilson’s disease, hypothyroidism, acromegaly, X-linked hypophosphatemic rickets, diabetes mellitus, amyloidosis, and previous trauma or surgery of the joint.

We report a rare case of idiopathic tophaceous CPPD deposition disease of TMJ in a 29-year-old female patient without secondary etiology by comprehensive metabolic workup and history survey. The patient presented with gradual hearing impairment and tinnitus of the right ear for 9 months. She had no specific systemic or metabolic disease and had no family, trauma, or surgery history that could account for her condition. Her laboratory data consisting of serum parathyroid hormone, calcium, phosphate, transferrin, magnesium, uric acid, erythrocyte sedimentation rate, and blood glucose level were all within normal ranges, except for a mild anemia and low serum ferritin level. The computed tomography scan displayed a mass within the right TMJ with compression of the right external auditory canal, resulting in a threadlike narrowing of the auditory canal (Fig. 1A). The major clinical and imaging differential diagnoses included osteochondroma, chondroma, chondroblastoma, and synovial chondromatosis. The tumor was totally excised. Grossly, the tumor measured 2.7 cm × 1.5 cm × 0.7 cm in size. It contained numerous chalky white materials, resembling gouty tophus. Microscopically, the tumor showed rhomboid-shaped crystals with calcification and foreign body giant cell reaction to the crystals (Fig. 1B). By polarized light microscopy, the rhomboid-shaped crystals revealed weak positive birefringence, characteristic of CPPD and different from needle-shaped crystals with negative birefringence of gouty tophus (Fig. 1C). By scanning electron microscopy, the crystals were also rhomboid-shaped (Fig. 1D). Thus, CPPD deposition disease of the TMJ was diagnosed. After the surgery, the patient’s symptoms improved, and there was no recurrence of the disease after a follow-up period of 1 year.

CPPD appears to have a predilection for large joints and rarely occurs in the TMJ. In a recent review, there are 58 cases of tophaceous CPPD deposition disease of TMJ reported in the literature. Moreover, CPPD deposition disease usually occurs in elderly patients and is rarely found in younger patients. The average age of patients with CPPD deposition disease of the TMJ is 60.2 years. To the best of our knowledge, our patient is the youngest one with tophaceous CPPD deposition disease of the TMJ. We conclude that tophaceous CPPD deposition disease should be considered in the differential diagnoses of a TMJ mass lesion, even for patients in a young age group.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.
References


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Figure 1  Computed tomographic (CT), light microscopic, and scanning electron microscopic (SEM) photographs of our case. (A) The CT scan displayed an irregular amorphous mass (arrow) within the right temporomandibular joint (TMJ) with compression of the right external auditory canal, resulting in a threadlike narrowing of the auditory canal (inlet: gross picture of the surgical specimen). (B) Microscopically, the tumor shows a chronic inflammatory cell infiltrate and foreign body giant cell reaction (white arrows) to the aggregates of characteristic rhomboid-shaped crystals (black arrows; hematoxylin and eosin stain, ×200). (C) By polarized light microscopy, the rhomboid-shaped crystals reveal weak positive birefringence, which are characteristic of calcium pyrophosphate dihydrate (×400). (D) The SEM examination reveals rhomboid-shaped crystals of calcium pyrophosphate dihydrate (3000×).