Case Report

Tophaceous Calcium Pyrophosphate Dihydrate Deposition Disease of the Temporomandibular Joint

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ABSTRACT. Tophaceous pseudogout is a rare manifestation of calcium pyrophosphate dihydrate (CPPD) deposition disease that particularly affects the temporomandibular joint (TMJ). We describe a case of tophaceous pseudogout and review the literature. Thirty-four cases of chronic CPPD deposition disease affecting the TMJ are described. Symptoms usually included pain and swelling. Most patients required surgery because of extensive crystal deposits, usually localized to the joint and adjacent structures but occasionally invasive. For many patients, malignancy was the preoperative diagnosis. Although patients with acute pseudogout of the TMJ may have involvement of other joints, tophaceous pseudogout was predominantly isolated to the TMJ. (J Rheumatol 2008;35:717–21)

Key Indexing Terms:
PSEUDOGOUT
CALCIUM PYROPHOSPHATE DIHYDRATE DEPOSITION
CHONDROCALCINOSIS
TEMPOROMANDIBULAR JOINT

Calcium pyrophosphate dihydrate (CPPD) deposition is common in elderly patients with a radiographic prevalence of 30%–40% in patients over 84 years of age[1]. However, CPPD deposition disease can also be associated with osteoarthritis, prior joint trauma, or metabolic conditions such as hyperparathyroidism, dialysis-dependent renal failure, hemochromatosis, and hypomagnesemia. The crystals may be deposited in a joint asymptomatically, or they may cause acute inflammatory arthritis (pseudogout), chronic degenerative arthritis (pseudoosteoarthritis), or chronic symmetric inflammatory polyarthritis (pseudo-rheumatoid arthritis). Rarely, pseudotophaceous or tumoral deposits of CPPD can cause isolated joint destruction, particularly in the temperomandibular joint (TMJ). This has been termed "tophaceous pseudogout." We describe a case of tophaceous pseudogout limited to a single TMJ and associated with destruction of the joint and part of the mandible. We also review the published cases.

CASE REPORT

Our patient was a 52-year-old woman with longstanding and gradually worsening left TMJ pain. The pain was episodic, occurring about twice per year and lasting for 2 weeks. There were no clear precipitating factors. She had some superimposed regular pain associated with prolonged chewing on that side. The right TMJ was asymptomatic. She had a remote history of trauma with a baseball bat to the left side of her face and several molars removed on that side secondary to caries. There were no other systemic symptoms, and no other joint symptoms.

On examination, she had some facial asymmetry with the right mandible being more prominent. Her left TMJ was tender with some swelling. Mouth opening was limited and painful. The remainder of the examination was unremarkable.

Blood testing showed a slight elevation in erythrocyte sedimentation rate to 22 (upper limit of normal 21), normal complete blood count, and negative rheumatoid factor and antinuclear antibody. Thyroid-stimulating hormone was normal, as were electrolytes and serum creatinine. Uric acid level was not done. Magnetic resonance imaging showed a normal right TMJ. The left TMJ had soft tissue material filling and distending the joint space and eroding the articular cortex of the condylar fossa (Figure 1). The erosions extended quite close to the inner table of the skull. There was minimal anterior translation of the left mandibular condyle with mouth opening.

Subsequently, she underwent an open exploration of the left TMJ to biopsy the soft tissue. At the same time she underwent a condelectomy of the TMJ to improve access to the diseased tissue and to reduce the chances of a maxillary artery bleed. Pathology report of the excised mandibular head was normal. The excised soft tissue (Figure 2A) from the joint showed fragments of synovial tissue with abundant calcium pyrophosphate crystal deposition. The crystals evoked a histiocytic inflammatory reaction, as well as prominent cartilaginous metaplasia (Figure 2B). There were some fragments of sclerotic bone within the tissue mass. This was diagnostic of tophaceous or tumoral pseudogout.

DISCUSSION

We found 3 reports of a total of 5 cases of acute pseudogout involving the TMJ[3-5]. All 5 patients had CPPD deposits in additional joints including wrists, knees, or both. Three patients were treated with nonsteroidal antiinflammatory drugs and 2 had no treatment. Symptoms resolved in all

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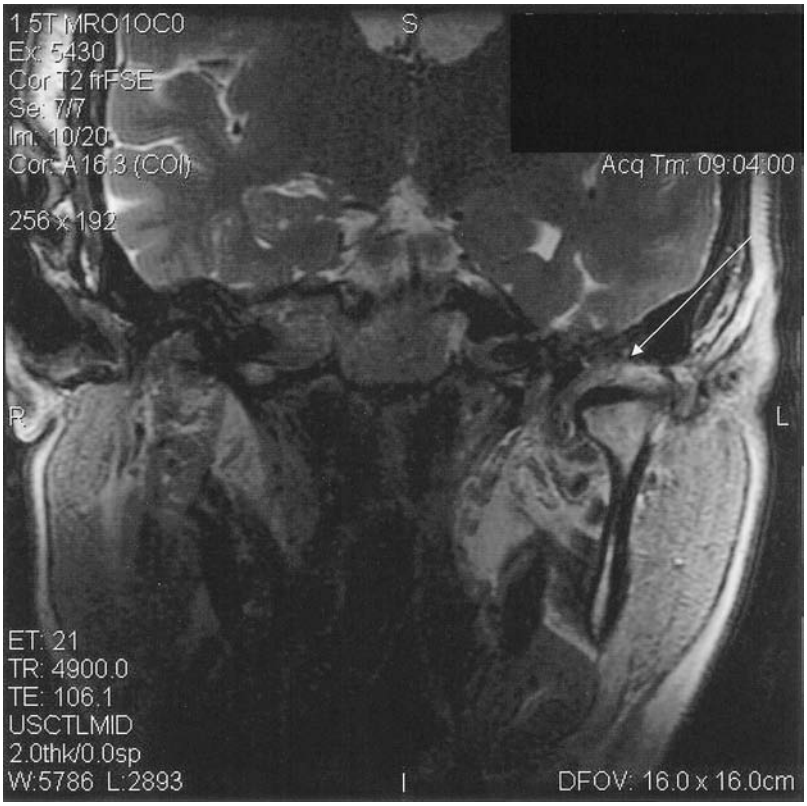

Figure 1. T2 weighted magnetic resonance imaging showing soft tissue mass surrounding left mandibular condyle, distending the temperomandibular joint (white arrow).

patients, and only 1 recurrence was described. One patient had symptoms associated with bruxism, and one patient received prednisone treatment for previously diagnosed polymyalgia rheumatica.

There have been 34 published case reports of chronic tophaceous pseudogout, including ours. Patients have a mean age of 58 years, and the ratio of women to men is about 2:1. Symptom duration prior to surgery ranged from 2 months to 20 years. Pain was present in 65% of cases, predominantly in the TMJ itself, though occasionally affecting the ear and pre-auricular area. Swelling was present in 79% of patients. Jaw opening was decreased in 53% of patients, though it ranged from subtle to severe. In contrast to acute TMJ pseudogout cases, all of which involved other joints, only 12% of the chronic cases had CPPD deposits documented in other joints. Four patients had associated decreased hearing, 3 had associated diabetes mellitus, 2 had remote preceding trauma, and 1 had coexistent gout. One patient was found to have hypercalcemia, hypophosphatemia, and increased parathyroid hormone, and on investigations was diagnosed with papillary thyroid cancer and a parathyroid adenoma. The published cases are summarized in Table 1, including the clinical symptoms and any followup data available.

As our data review shows, almost all patients required surgery due to the large size of deposits, joint destruction, and, occasionally, invasion of the crystal deposits into adjacent structures. One patient was unable to have complete resection due to the extent of tumor invasion; however, the patient had decreased pain and swelling up to 1 year postoperatively[29]. Only 2 patients were managed conservatively after diagnosis was confirmed by aspiration of synovial fluid or biopsy. Recurrence was described in 8.8% of patients, although length of followup was variable and was not described in all cases.

CPPD deposition in cartilage can be an incidental radiographic finding or may cause either acute or chronic arthritis. There is predilection for fibrocartilage, and the most frequently involved sites are the knee menisci, the triangular ligament of the wrist, and the symphysis pubis. Although the TMJ also contains fibrocartilage, symptomatic disease in this area is quite rare.

Unlike gout, in which large masses of sodium urate crystals or tophi commonly can deposit in soft tissues or joints, massive deposition of crystals is rare in CPPD disease. However, the literature on tophaceous pseudogout suggests that the most common site for this disease is the TMJ[2]. Other sites reported include the finger, toe, cervical spine, wrist, hip, anterior cruciate ligament, elbow, hand, and knee[2].

Although the first description of tophaceous pseudogout

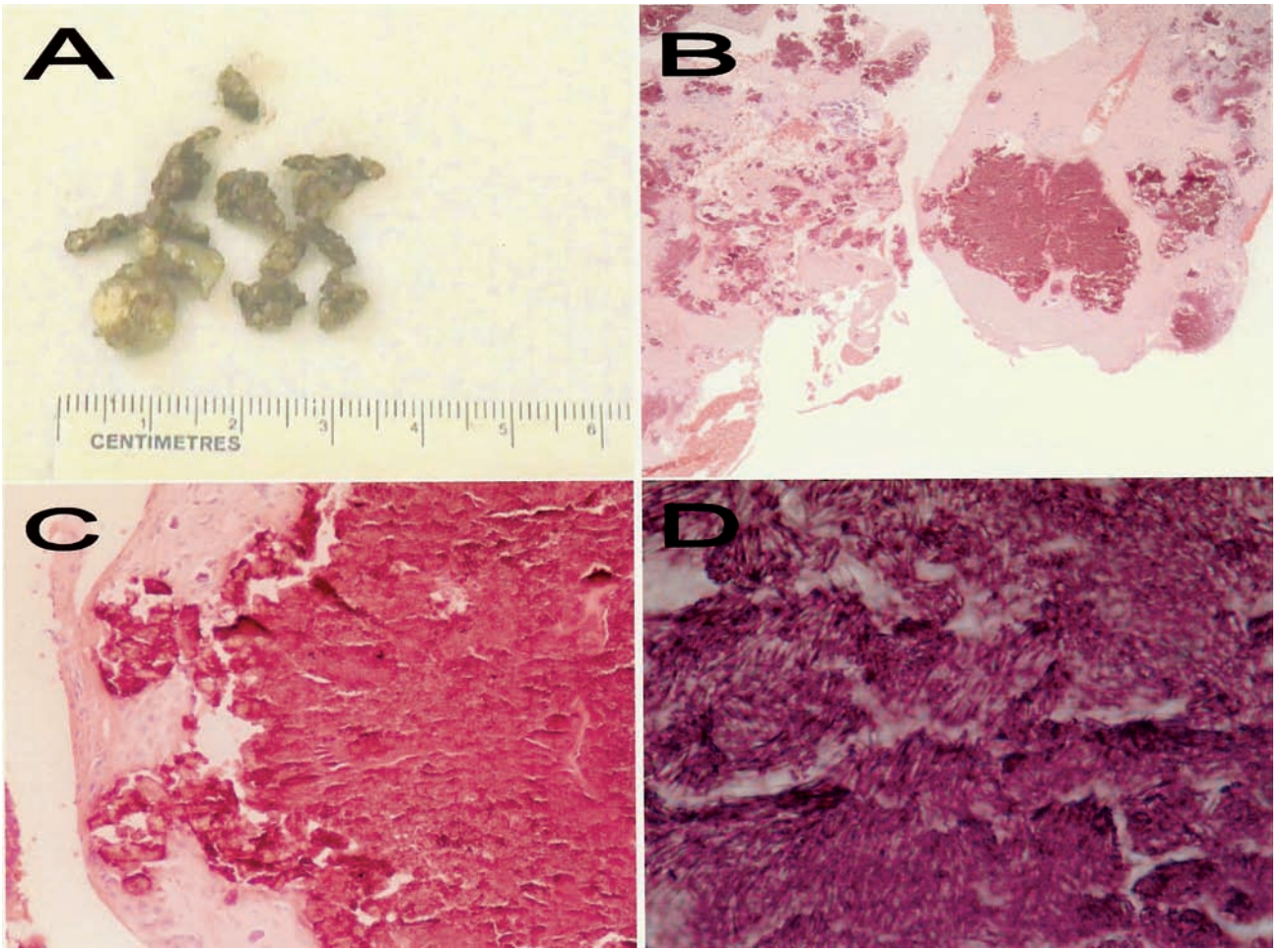

Figure 2. Pathology of resected mass from left TMJ. A. Gross specimen of irregular yellow-brown fragments of tissue, including several bone fragments. B-D. Histopathology showing fragments of synovial tissue with abundant calcium pyrophosphate crystal deposition. The crystals have evoked a histiocytic inflammatory reaction, as well as prominent cartilaginous metaplasia. Magnification: B: 20×, C: 100×, D: 500× (oil).

affecting the TMJ was published in *The Journal of Rheumatology* in 1976[6], the majority of further cases have been described in nonrheumatologic journals.

This condition has a tendency to mimic more aggressive and malignant conditions, particularly chondrosarcoma. Other pre-operative differential diagnoses included synovial chondromatosis, tumoral calcinosis, and benign tumors like chondroma and osteochondroma. Early identification of CPPD crystals reduces the extent of surgery and improves the prognosis.

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Table 1. Summary of published cases of chronic tophaceous pseudogout of the TMJ.

Reference	Age, Sex	Pain, Swelling	Duration	CPPD in Other Joints	Recurrence (time)
Pritzer[6]	55 M	No, yes	8 mos	NA	NA
de Vos[7]	51 F	Yes, no	14 mos	Yes*	No (1 yr)
Zemplenyi[8]	51 F	Yes, yes	2 yrs	No	Yes (2 yrs)
Kamatani[9]	57 M	No, yes	2 yrs	NA	NA
Mogi[10]	54 F	Yes, yes	8 yrs	No	No (20 mo)
Gross[11]	59 F	Yes, yes	chronic	Yes*	No (10 mo)
Lambert[12]	41 M	No, yes	chronic	No	No (NA)
Magno[13]	53 F	Yes, no	NA	NA	No (2 yrs)
Dijkgraaf[14]	53 F	Yes, yes	18 mos	Yes†	Yes (10 mo)**
Ishida[2]	47 F	No, yes	8 yrs	No	NA
Ishida[2]	50 F	No, yes	20 yrs	No	Yes[5]
Ishida[2]	55 F	No, no	None	No	NA
Pynn[15]	58 M	No, yes	18 mos	No	No (5 mo)
Chuong[16]	65 F	Yes, yes	8 yrs	No	No (2 yrs)
Onodera[17]	48 F	Yes, yes	3 yrs	No	No (18 mo)
Kurihara[18]	85 M	Yes, yes	2 yrs	NA	No (6 mo)
Vargas[19]	66 F	Yes, yes	2 mos	No	No (6 mo)
Jordan[20]	80 M	No, no	11 mos	NA	NA
Strobl[21]	51 F	Yes, no	18 mos	NA	NA
Goudot[22]	63 F	Yes, yes	10 yrs	Yes‡	No (1 yr)
Nakagawa[23]	60 F	Yes, yes	15 yrs	NA	No (3 yrs)
Nakagawa[23]	45 F	Yes, no	yrs	NA	NA
Aoyama[24]	45 F	Yes, yes	8 yrs	No	No (7 mo)
Mostafapour[25]	65 F	Yes, yes	3 mos	No	No (18 mo)
Eriksson[26]	72 M	Yes, yes	yrs	NA	No (5 yrs)
Olin[27]	51 F	No, yes	2 mos	NA	No (18 mo)
Appel[28]	69 M	No, yes	1 yr	NA	No (18 mo)
Koitschev[29]	66 F	No, yes	18 mos	No	Not resectable
Osano[30]	40 M	Yes, yes	16 mos	No	No (2 yrs)
Marsot-Dupuch[31]	70 F	Yes, yes	10 yrs	No	NA
Marsot-Dupuch[31]	53 M	No, no	1 yr	NA	NA
Meul[32]	54 M	Yes, yes	3 yrs	No	NA
Smolka[33]	74 F	Yes, yes	NA	No	No (1 yr)
Reynolds	52 F	Yes, yes	yrs	NA	No (2 yrs)

* Not specified; † knees, left elbow, right proximal interphalangeal joint; ‡ knees, hips; ** second resection and No recurrence after 10 yrs; # 3 surgeries over the years. TMJ: temporomandibular joint.

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