Case report

Calcium pyrophosphate dihydrate arthropathy with condylar destruction of the temporomandibular joint

Hitoshi Osano, Koichi Matsumoto and Mikio Kusama

Department of Oral and Maxillofacial Surgery, Jichi Medical School, Tochigi 329-0498

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Abstract: We report a case of calcium pyrophosphate dihydrate (CPPD) arthropathy with condylar destruction of the temporomandibular joint. Diagnosis was made on the basis of clinical findings and x-ray evidence of severe destruction of the condyle. The most likely diagnosis was considered to be neoplasm and a secondary infection of the left TMJ. Review of sections of the condyle taken at operation confirmed the diagnosis of left TMJ pseudogout. The postoperative course was uneventful with improvement in the clinical symptoms. (J. Oral Sci. 45, 223-226, 2003)

Key words: calcium pyrophosphate dihydrate arthropathy; pseudogout; temporomandibular joint.

Introduction

Deposition of calcium pyrophosphate dihydrate crystals in articular cartilage is a common age-related phenomenon. Calcium pyrophosphate dihydrate crystals preferentially deposit within fibrocartilage and are the most common cause of cartilage calcification. A causal role for calcium pyrophosphate dihydrate crystals in acute inflammation is accepted, but their role in chronic arthropathy remains to be elucidated.

Pseudogout is a disease of calcium pyrophosphate dihydrate (CPPD) deposition. Involvement of the temporomandibular joint (TMJ) is rare. The main symptoms of CPPD deposition are acute attacks of arthritis, swelling, pain and limited joint mobility. Most attacks occur

Correspondence to Dr. Hitoshi Osano, Department of Oral and Maxillofacial Surgery, Jichi Medical School, 3311-1Yakushiji, Minamikawachi-machi, Tochigi 329-0498, Japan Tel: +81-285-58-7390 Fax: +81-285-44-8669 E-mail: osano@jichi.ac.jp spontaneously but provoking factors include intercurrent illness, surgery, and local trauma. Although any joint may be involved, the knee is by far the most common site, followed by the wrist, shoulder, and ankle.

Case report

A 40-year-old man was referred to our hospital with a chief complaint of pain and swelling of his left TMJ. The patient had experienced one episode of pain and swelling of his TMJ two years previously, and the symptoms were relieved by intravenous drip-infusion of antibiotics. The patient also had a history of peptic duodenal ulcer for which he had received medication. Physical examination showed diffuse swelling and pain in the left TMJ (Fig. 1). The interincisal opening was 16 mm with a distinct deviation to the left side. X-ray findings revealed anterior



Fig. 1 Extraoral appearance reveals bulging of the middle of the left side of the face.

and lateral expansion of the left condylar process. From the lateral view the coronoid space was filled with a walnut-sized mass that was radiopaque (Fig. 2). The appearance indicated a destructive lesion originating from the left condylar process and that had broken through its cortex. CT scan findings revealed a calcified mass in the joint space of the left TMJ. Magnetic resonance imaging (MRI) revealed lobulated masses in the left coronoid space that showed a higher signal intensity than muscle on T2weighted images (Fig. 3).

There were no abnormal findings on peripheral blood examination. Results of laboratory investigations revealed normal serum values of sodium, potassium, calcium, phosphorus, alkaline phosphatase, uric acid, and creatine. The C-reactive protein level was 5.68 mg/dl. Clinical and radiographic findings suggested an initial diagnosis of neoplasm and secondary infection of the left TMJ. The patient was treated with antibiotics and non-steroidal antiinflammatory drugs because of spontaneous pain prior to surgery, with symptomatic improvement. The left condylar process was exposed via a preauricular incision. Left condylectomy and synovectomy were performed for three lobulated masses. The surgically removed masses were solid whitish-gray in color, and chalky in appearance (Fig. 4). The resected left condyle showed extensive lysis and degenerative change, as noted on the CT scans.

Histopathological examination of the mass by hematoxylin and eosin staining revealed crystalline calcium deposits surrounded by fibroblasts, macrophages, and foreign body-type giant cells (Fig. 5). The granulomatous tissue of the hyperplastic synovial membrane contained



Fig. 2 X-ray findings reveal destruction of the left condylar process and the presence of a walnut-sized radiopaque mass in the coronoid space.



Fig. 4 Photograph showing three lobulated masses of the resected specimen.



Fig. 3 Magnetic resonance imaging reveals lobulated masses in the left coronoid space, showing higher signal intensities than muscle on T2-weighted images.



Fig. 5 Photomicrograph of the tumoral mass showing deposits of crystals surrounded by fibroblasts, macrophages, and foreign body-type giant cells.

crystal-like decalcified areas (Fig. 6). Examination by polarizing microscopy revealed rhomboid, rhomboidal and rod-like crystals that were weakly birefringent (Fig. 7). Consequently, the diagnosis was left TMJ pseudogout. In addition, postoperative x-ray findings revealed calcium deposition at the rotator cuff.

The postoperative course was uneventful. During followup over a 2-year period, the patient attained an interincisal opening of 38 mm without pain; however there was still a slight deviation of the mandible to the left side during mouth opening.

Discussion

Common presentations of pyrophosphate arthropathy are acute synovitis (pseudogout), chronic arthritis, or as an incidental finding. Other presentations are rare. Pseudogout is the most common cause of acute monoarthritis in the



Fig. 6 Photomicrograph of the hyperplastic synovial membrane. The granulomatous tissue contains crystal-like decalcified areas.



Fig. 7. Examination by polarizing microscopy reveals rhomboid, rhomboidal and rod-like crystals that are weakly birefringent.

elderly. Most attacks occur spontaneously but provoking factors include intercurrent illness, surgery, and local trauma. Although any joint may be involved, the knee is by far the most common site, followed by the wrist, shoulder, and ankle. Involvement of the TMJ is rare, and the first report of calcium pyrophosphate dihydrate arthropathy of the TMJ was made by Pritzker et al (1). Only 20 cases of this condition affecting the TMJ have been reported (1-16). In the literature, this disease develops more frequently in women (16 cases, 80%), particularly those aged 50-60 years (average age, 57.4 years). Pain (11 cases) and swelling (13 cases) of the temporomandibular joint and limited mouth opening are the main clinical symptoms.

Critical investigations include synovial fluid analysis and plain radiographs. Needle aspiration of an affected joint may provide sufficient crystal samples to make a histological diagnosis (7), although it may not even be considered if a neoplastic process is suspected. In pseudogout, aspirated fluid is often turbid or bloodstained with an elevated cell count, while fluid from chronic pyrophosphate arthropathy shows variable characteristics.

A number of systemic conditions have been reported to be associated with CPPD, including hyperparathyroidism, hemochromatosis, hypomagnesemia, and aging (2). Other associated conditions or diseases include osteoarthritis, hypothyroidism, amyloidosis, trauma, and surgical intervention. In the literature, coexistent systemic diseases considered to be closely associated with CPPD of the TMJ, including diabetes mellitus, CPPD in the wrist and knee, generalized arthritis, polymyalgia rheumatica, hypothyroidism, hypertension and hiatal hernia, have been described in 8 patients out of 20 reported cases (1-16). CPPD deposition disease of the TMJ usually involves the condyle and fibrocartilage (11 out of 20 cases) (1-16).

Radiographic aspects relate both to calcification and arthropathy. However, these radiographic findings are nonspecific (1). The radiographic features of CPPD of the TMJ are quite consistent with the presence of a calcified mass anterior to the condyle, erosion and sclerosis of the fossa, and misshapen condyles. Many of these features are evident on plain X-ray films, but they are more conclusive on computed tomography scans and magnetic resonance imaging. Since TMJ involvement is rare, and laboratory and radiographic studies are nonspecific for CPPD, this entity is rarely suspected preoperatively (6). In this case, based on the clinical and radiographic findings, an initial diagnosis of neoplasm and secondary infection of the left TMJ was made.

Histological features of CPPD include nodular, friable, gritty granulomatous tissue within the joint space and

sometimes within the periarticular soft tissues (8), and condylar and fossa erosion is often seen. In this case, areas of CPPD crystals, a foreign body type of chondromatosis reaction containing histiocytes and giant cells, and chondroid metaplasia in and around the areas of crystal deposition (8) were observed.

The principal differential diagnosis for pseudogout is sepsis, gout or tumor. In our case CPPD deposition disease was not considered because radiographic examination and CT scans indicated that the severe destruction of the condyle was due to a malignant tumor.

Gram staining and culture of joint fluid should be undertaken even when calcium pyrophosphate dihydrate crystals are identified. Marked bloodstaining may lead to the consideration of other causes of hemarthrosis.

The preferred treatment for pseudogout is local therapy, since it usually affects only one or a few joints in elderly patients. Treatment of CPPD arthropathy is usually conservative. Aspiration alone often relieves symptoms, but may be combined with intra-articular steroids in severe cases. Simple analgesics and NSAIDs provide additional benefit but should be used cautiously in the elderly. Surgical treatment is indicated if masses consisting of calcareous material are deposited in the TMJ (3). Mogi et al. reported that surgical treatment is an effective way to remove the crystalline deposits, particularly for uniarticular involvement (4). The literature cites 16 cases in which masses were excised by surgical operations, including 4 cases of condylectomy.

Fibrocartilage injury is considered to be an initiating factor in CPPD, but the mechanism of CPPD deposition is imperfectly understood. Good et al. reported a case of CPPD arthropathy of the TMJ associated with bruxism (5). Patients with CPPD deposition in the TMJ can also develop CPPD deposition in organs, and long-term follow-up is necessary.

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