

A Case of Calcium Pyrophosphate Dihydrate Crystal Deposition Disease Presenting as an Acute Polyarthrititis

We report a case of calcium pyrophosphate dihydrate crystal deposition disease (CPDD) presenting as an acute polyarthrititis. A 66-yr-old woman was admitted with a 5-day history of fever and multiple joint pain including wrists, elbows, shoulders, knees, and ankles developed 5 days before admission. Her plain radiographs of wrists, elbows, shoulders, knees, and ankles showed chondrocalcinosis. The pubic symphysis, lumbar intervertebral discs, and both hip joints, which were asymptomatic, also had calcium deposits. The compensated polarized microscopic examination of the joint fluid, aspirated from the right knee revealed intracellular and extracellular weakly positive birefringent crystals, confirming the CPDD. This case showed that CPDD may manifest as an acute polyarthrititis mimicking acute onset rheumatoid arthritis.

Key Words : Arthritis; Calcium Pyrophosphate; Calcium Pyrophosphate Dihydrate Deposition Disease

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Received : 8 June 2001

Accepted : 26 July 2001

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INTRODUCTION

Calcium pyrophosphate dihydrate deposition disease (CPDD) includes arthritic syndromes associated with calcium pyrophosphate dihydrate (CPPD) crystal deposition in articular tissues (1). It has three common presentations consisting of acute synovitis that was also called pseudogout clinically, chronic arthritis, and asymptomatic chondrocalcinosis (2, 3). Acute pseudogout is the classic presentation and is the commonest cause of acute monoarthritis in the elderly. The knee joint is the most commonly affected site. Concurrent attacks in more than one joint are unusual. Moreover, polyarticular attacks are rare (4).

We report a case of CPDD presenting as an acute polyarthrititis with relevant literature review.

CASE REPORT

A 66-yr-old female was admitted via emergency room complaining of fever and polyarthralgia, including knees, wrists, shoulders, elbows, and ankles. She also complained of general weakness, myalgia, and anorexia. These manifestations suddenly developed 5 days before admission. Ten years earlier, she was diagnosed with osteoarthritis on both knees, and had analgesic medications intermittently. Family history was unremarkable. In the emergency room, her body temperature was 38.5°C. Blood pressure, pulse rate, and respiration rate were normal. Physical examination showed erythema, swelling, warmth, and tenderness on the knees, wrists, shoulders, el-

bows, and ankles.

Laboratory findings were as follows: peripheral blood leukocyte count 11,400/ μ L (neutrophils 72.3%), hemoglobin 12.4 g/dL, platelet 325,000/ μ L, Westergren erythrocyte sedimentation rate 114 mm/hr, C-reactive protein 18.8 mg/dL, and negative results for rheumatoid factor, antinuclear antibody, hepatitis B antigen, and anti-hepatitis C antibody. Blood cultures revealed no bacterial growth. Serum concentrations of calcium, phosphorus, magnesium, thyroid hormone, and parathyroid hormone were normal.

The plain radiographs of the wrists, elbows, shoulders, knees (Fig. 1), and ankles (Fig. 2) showed chondrocalcinosis. The pubic symphysis, lumbar intervertebral disc, and both hip joints, which were asymptomatic, also demonstrated calcium deposits (Fig. 2). Technetium 99m methylene diphosphonate scintigraphy showed increased joint uptake on the knees, wrists, shoulders, elbows, and ankles (Fig. 3).

By arthrocentesis on the right knee, 45 mL of turbid and yellow-whitish fluid was aspirated. In the joint fluid, the leukocyte count was 13,000/ μ L (polymorphonuclear leukocyte 86%). Gram stain and culture of the fluid revealed no evidences of bacteria. Under the compensated polarized microscope of the joint fluid, rod-shaped, weakly positive birefringent intracellular and extracellular CPPD crystals were demonstrated (Fig. 4).

Intra-articular methylprednisolone injection into the right knee joint was performed. Then, she was treated with nonsteroidal anti-inflammatory drug (370 mg of talniflumate three times a day), colchicine (0.6 mg once a day), and acetaminophen (1,300 mg three times a day). Five days

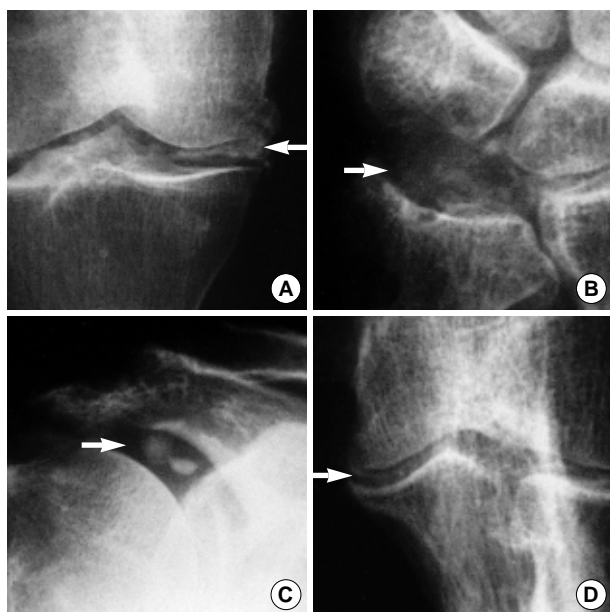


Fig. 1. The plain radiographs of the patient demonstrate chondrocalcinosis on knee (A), wrist (B), shoulder (C), and elbow (D).

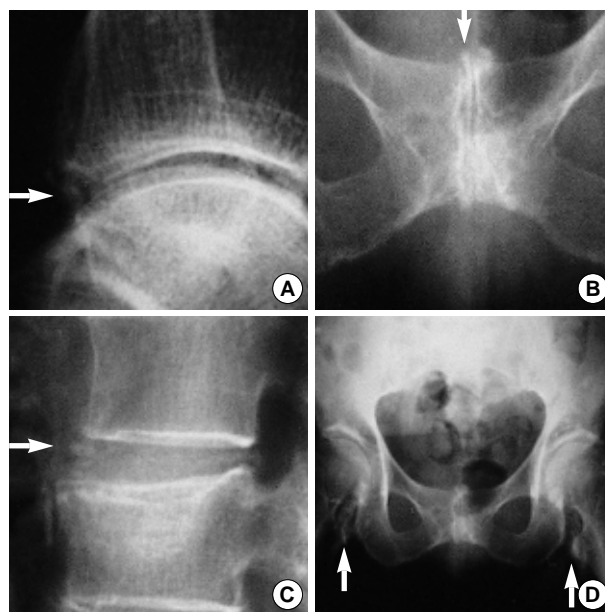


Fig. 2. The plain radiographs of the patient demonstrate chondrocalcinosis on ankle (A), pubic symphysis (B), lumbar intervertebral disc (C), and both hip joints (D).

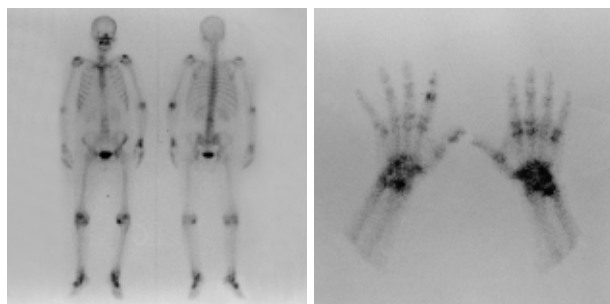


Fig. 3. Technetium 99m methylene diphosphonate scintigraphy shows increased joint uptake on the knees, wrists, shoulders, elbows, and ankles.

later, her symptoms including fever, multiple arthralgia, anorexia, and general weakness gradually subsided. The patient was discharged and her regular follow-up for 2 months found no evidence of arthritic recurrence.

DISCUSSION

CPPD crystalline deposits occur almost in and around the joints and are characterized by the presence of calcification of the articular cartilage, menisci, synovium, and periarticular tissues (5). These deposits may be asymptomatic or associated with chronic articular manifestations or acute episodic arthritis, as well as a wide variety of other musculoskeletal manifestations. The various clinical patterns of CPPD include pseudogout (acute synovitis), pseudorheumatoid arthritis,

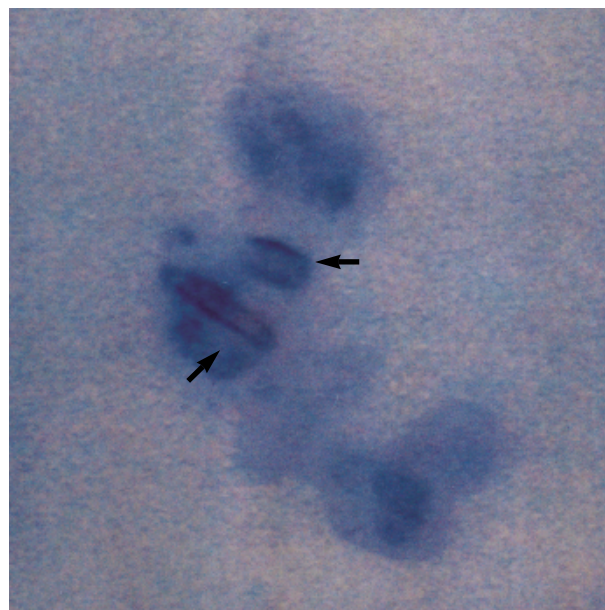


Fig. 4. Polarizing microscopic examination of the right knee joint fluid shows rod-shaped, weakly positive birefringent intracellular crystals of calcium pyrophosphate dihydrate ($\times 400$).

chronic pyrophosphate arthropathy (pseudo-osteoarthritis), asymptomatic CPPD deposition, pseudoneuropathic joints, hemarthrosis, monoarthropathy, solitary tophaceous deposit, traumatic arthritis, and pseudo-ankylosing spondylitis (6). Three common presentations are pseudogout, chronic pyrophosphate arthropathy, and asymptomatic CPPD deposition

(2, 3). Among them, asymptomatic CPPD crystal deposition is the most common clinical presentation. The pseudogout presentation is a frequent cause of acute monoarthritis in the elderly. This attack may be precipitated by trauma, surgery, serious medical illness, such as myocardial infarction, stroke, and pneumonia, and arthroscopy (7). The knee is the most commonly involved joint, followed by the wrist, shoulder, ankle, and elbow. The pseudogout should be differentiated from gout (4) and septic arthritis (8, 9). Like pseudogout, chronic pyrophosphate arthropathy principally affects large and medium-sized joints, the knees being the most commonly involved joints. The disease may manifest as chronic pain, early morning and inactivity stiffness, limitation of motion, and functional impairment.

Approximately 5% of the CPDD patients present as pseudorheumatoid arthritis with multiple joint involvements with subacute or chronic attacks lasting 4 weeks to several months. In these cases, the differential diagnosis between CPDD and rheumatoid arthritis is difficult (10, 11). Such a diagnostic problem can be solved by radiologic evaluations. The patients with CPPD usually show chondrocalcinosis and bony erosions are not apparent in uncomplicated CPDD, while the patients with rheumatoid arthritis show bony erosions and destructive joint changes (12).

In our case, the patient with presented as acute onset of the disease and polyarticular involvement including knees, wrists, shoulders, elbows, and ankles. Radiography revealed chondrocalcinosis even in asymptomatic joint areas, such as pubic symphysis, lumbar vertebra, and both hips as well as symptomatic joints. This pattern of presentation mimicking acute onset rheumatoid arthritis is rare.

CPDD is associated with age, trauma, osteoarthritis, and some metabolic diseases, such as hyperparathyroidism, hypomagnesemia, gout, hemochromatosis, hypothyroidism, and hypophosphatasia (13, 14). Our patient was old and had knee osteoarthritis, but had no evidence of metabolic disorders. In addition, we can not find the evident precipitating factor which provoke the attack.

Diagnosis of CPDD can be confirmed by the demonstration of rhomboid or rod-shaped, weakly positive birefringent CPPD crystals in synovial fluid or articular tissues and the presence of characteristic intraarticular calcified deposits in synovium, articular cartilage, or menisci (14, 15).

This is the first reported case of CPDD presenting as an acute polyarthritis in Korea, which should be differentiated from acute onset rheumatoid arthritis.

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