Clinical Case Reports: Open Access

Yumed Text
OPEN ACCESS JOURNALS

Case Report | Vol 5 Iss 2
ISSN: 2582-5038
https://dx.doi.org/10.46527/2582-5038.207

Calcium Pyrophosphate Deposition Disease Simulating (or Coexisting) with Remitting Seronegative Symmetrical Synovitis with Pitting Edema (RS3PE Syndrome)

Attout H[*] and Amichi S

Internal Medicine Unit, General Hospital les Cévennes, 30100 Ales, France

[*]**Corresponding author:** Hassene Attout, Internal Medicine Unit, General Hospital les Cévennes, 30100 Ales, France, Tel: +33 4 66 78 33 33; E-mail: dr.attout@ch-ales.fr

Received: March 16, 2022; **Accepted:** March 24, 2022; **Published:** April 02, 2022

Abstract

Calcium pyrophosphate deposition (CPPD) disease is arthritis caused by calcium pyrophosphate (CPP) crystals. Patients typically present with the acute onset of monoarticular or polyarticular arthritis. The polyarticular form can simulate systemic disease as rheumatoid arthritis, polymyalgia rheumatica or gout.

We present the case of a 77-year-old man with a 6-week history of a full spectrum of signs and symptoms compatible with RS3PE. He was treated by prednisolone 15 mg/day. Because persisting swelling and pain, he was admitted for further investigations. Radiographs of the knees revealed significant chondrocalcinosis. Analysis of synovial fluid revealed presence of calcium pyrophosphate crystals.

This case demonstrated the importance of considering the possibility of crystal-induced arthritis such as CPPD, as well as a malignant disease when diagnosing the primary disease responsible for RS3PE syndrome. Conservative therapy with steroids rapidly resolved all symptoms.

Keywords: ***Calcium pyrophosphate deposition disease; chondrocalcinosis; Remitting seronegative symmetrical synovitis with pitting edema***

1. Introduction

Calcium pyrophosphate deposition (CPPD) disease is arthritis caused by calcium pyrophosphate (CPP) crystals. Until recently, CPPD was also named chondrocalcinosis / pseudogout. Patients typically present with the acute onset of mono-oligo articular or polyarticular arthritis. So, it can mimic rheumatoid arthritis (RA), polymyalgia rheumatica (PMR), osteoarthritis and gout [1].

Remitting seronegative symmetrical synovitis with pitting edema (RS3PE) syndrome is an uncommon disorder characterized by an acute-onset seronegative inflammation with pitting edema in the distal extremities. It is a rare disease, and the incidence rate is 0.09% in patients aged 50 and over. Most cases of RS3PE syndrome are idiopathic, although the syndrome has been associated with malignancy, PMR, RA, Sjögren's syndrome, and neurodegenerative diseases. Patients show excellent response to low dose steroids with complete and sustained remissions [2,3].

In this case report we discuss a case of CPDD syndrome where the initial presentation was bilateral pitting edema of the extremities simulating RS3PE syndrome. Patient showed dramatic response to low dose steroids.

2. Case Presentation

We present the case of a 77-year-old man with a full spectrum of signs and symptoms compatible with RS3PE. There was a positive past medical history of Parkinson's disease.

The patient had been diagnosed with RS3PE syndrome 1 month before admission in our unit. He was treated by prednisolone 15 mg per day but showed no significant response. He was admitted to our hospital for further examination.

On admission, her blood pressure was 140/86 mmHg, heart rate was 80/min, respiratory rate was 16/min, and body temperature was 37.5°C.

Examination revealed bilateral pitting edema of dorsum of hands and feet. He also had synovitis at wrists and effusion of knees. His left knee joint was particularly tender and painful. No superficial lymph nodes were noted. The Parkinson's symptoms were well controlled by L. dopamine and entacapone.

Initial blood test showed hemoglobin of 12 g/l, raised inflammatory markers (ESR 70, CRP 251 mg/l) and normal WBC. Serum uric acid calcium ionized, and ferritin were within the normal range. Autoantibody screen and rheumatoid factor were negative. Serum level MMP-3 (matrix metalloproteinase 3) assays was not done because not available in our hospital.

Analysis of synovial fluid of the left knee revealed slightly yellowish synovial fluid with WBC count of 15000 cells/mm^3, RBC count of 1200 cells/mm^3, and CPP crystals.

Radiographs of the knees revealed significant chondrocalcinosis.

In view of low-grade pyrexia, possibility of infective focus was ruled out by repeated blood and urine cultures. Infective endocarditis was ruled out by an echocardiogram. Enhanced computed tomography and laboratory data showed no evidence of malignancy.

A diagnosis of CPDD simulating RS3PE was suggested, and patient responded extremely well to prednisolone at 30 mg daily dosage. After 7 days, pain, swelling and CRP decreased dramatically. The patient was later discharged on a prednislone taper and with an outpatient rheumatology follow-up.

3. Discussion

Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease is characterized by the deposition of CPPD crystals in the articular cartilage visualized as intra-articular calcifications [chondrocalcinosis (CC)], with identification of the crystals in the synovial fluid, and an acute arthropathy called pseudogout. Knee is the most commonly involved joint, followed by the wrist, metacarpophalangeal, hips, shoulder, and ankle joints. It is polyarticular in approximately two-thirds of patients, and often occurs in an asymmetric pattern. Rarely, there may be deposition of CPPD crystals in other parts of the skeleton, e.g. in the spine, symphysis pubis or temporomandibular joint. CPPD can also be associated with tendinitis, tenosynovitis and bursitis.

If young people, CPPD may be associated with metabolic diseases such as hemochromatosis, hyperparathyroidism, hypophosphatasia, hypomagnesemia, Wilson's disease, hypothyroidism, gout, acromegaly, and X-linked hypophosphatemic rickets.

The gold standard for a definitive diagnosis of CPPD disease is the identification of CPPD crystals in synovial fluid [1].

The prevalence estimates of CPPD are usually based on radiographically detected CC and do not represent the whole spectrum of clinical CPPD disease. However, it has been reported that calcium pyrophosphate (CPP) crystal arthritis in an Italian population survey of the elderly is the third most common inflammatory rheumatic disease with the prevalence of 0.42% [5].

CC is frequently asymptomatic, and often identified as an incidental radiographic finding in elderly subjects...

However, CPPD deposits may associate with symptoms and present a variety of clinical phenotypes including an acute mono- or oligo-arthritis (pseudogout), as a generalized OA, sometimes referred to as chronic pyrophosphate arthropathy, and rarely as a form of polyarthritis that superficially resembles RA, PMR or gout [4]. For example, Paalanen et al. found that the prevalence of possible CPDD in their seronegative RA patients was 3.9% among those >60 years old [6]. In a large serie of 118 patients, Pego-Reigosa et al. found that 82 patients (69%) were diagnosed as having pure PMR and 36 (30%) met the diagnosis criteria for both PMR and CPDD [7].

The present case fulfilled the original criteria of diagnosis for RS3PE advocated by McCarty et al. [1]. Our patient had a full spectrum of signs and symptoms compatible with RS3PE: sudden onset of symmetrical distal synovitis and pitting oedema over the dorsum of the hands and feet and seronegative rheumatoid factors.

To the best of our knowledge, CPPD simulating RS3PE was never reported. Our patient was originally diagnosed with RS3PE and treated with low dose of prednisolone 15 mg/day.

The coexistence of acute CPPD and RS3PE in the same patient is theorically possible but until now no case was reported. Even so, rare cases of association of gout and RS3PE were published [8-10].

This case demonstrated the importance of considering the possibility of crystal-induced arthritis such as CPPD, as well as a malignant disease when diagnosing the primary disease responsible for RS3PE syndrome.

4. Learning Points

- The polyarticular form of CPDD can have symptoms that mimic Rheumatoid arthritis, gout, osteoarthritis or polymyalgia rheumatica.
- Remitting seronegative symmetrical synovitis with pitting edema (RS3PE) syndrome is an uncommon disorder characterized by an acute-onset seronegative inflammation with pitting edema in the distal extremities.
- CPDD can simulate RS3PE.

REFERENCES

1. Rosenthal AK, Ryan LM. Calcium pyrophosphate deposition disease. N Engl J Med. 2016;374(26): 2575-84.
2. McCarty DJ, O'Duffy JD, Pearson L, et al. Remitting seronegative symmetrical synovitis with pitting edema. RS3PE syndrome. JAMA. 1985;254(19):2763-7.
3. Olivio D, D'Amore M, Lacava R, et al. Benign edematous polysynovitis in the elderly (RS3PE syndrome). Clin Exp Rheumatol. 1994;12(6):669-73.
4. Gabay C, Van Linthoudt D, Ott H. The polyarticular form of chondrocalcinosis. Rev Med Suisse. 1989;109(11):901-06.
5. Ramonda R, Musacchio E, Perissinotto E, et al. Prevalence of chondrocalcinosis in Italian subjects from northeastern Italy. The Pro.V.A. (PROgetto Veneto Anziani) Study. Clin Exp Rheumatol. 2009;27(6):981-4.
6. Paalanen K, Rannio T, Asikainen J, et al. Prevalence of calcium pyrophosphate deposition disease in a cohort of patients with seronegative rheumatoid arthritis. Clin Exp Rheumatol. 2020;38(1):99-106.
7. Pego-Reigosa JM, Rodriguez-Rodriguez M, Hurtado-Hernandez Z, et al. Calcium pyrophosphate deposition disease mimicking polymyalgia rheumatica: a prospective follow-up study of predictive factors for this condition in patients presenting with polymyalgia symptoms. Arthritis Rheum. 2005;53(6):931-8.
8. Sugisaki K, Hirose T. Remitting seronegative symmetrical synovitis with pitting edema (RS3PE) syndrome following rupture of a gouty tophus. Mod Rheumatol. 2008;18(6):630-3.
9. Palazzi C, Olivieri I, D'Amico E, et al. Symmetrical pitting edema resembling RS3PE in gout. Clin Rheumatol. 2003;22(6):506-07.
10. Hakozaki M, Fukuda H, Tajino T, et al. Remitting Seronegative Symmetrical Synovitis with Pitting Edema Syndrome caused by crystal-induced arthritis of the wrist. Med Princ Pract. 2013;22(3):307-10.