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Unmasking the Mimic - A Case Report of Calcium Pyrophosphate Deposition Disease Presenting As Atypical Osteoarthritis

Priyanka Deshmukh[1] and Prashant Kaushik[2]*

[1]PGY2 resident, Internal Medicine Residency Program, TMG/OMECO at the NHS, Tahlequah, Oklahoma.

[2]Chief, Division of Rheumatology, Northeastern Health System (NHS); Clinical Professor, Department of Internal Medicine, Oklahoma State University Center for Health Sciences; Associate Program Director, Internal Medicine Residency Program, TMG/OMECO at the NHS, Tahlequah, Oklahoma.

***Correspondence:**
Prashant Kaushik, Chief, Division of Rheumatology, Northeastern Health System (NHS), Clinical Professor, Department of Internal Medicine, Oklahoma State University Center for Health Sciences, Associate Program Director, Internal Medicine Residency Program, TMG/OMECO at the NHS, Tahlequah, Oklahoma, Phone: 918 207 0025, Fax: 918 207 0026.

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ABSTRACT

The objective of this case report is to describe the importance of radiological evaluation in a case where calcium pyrophosphate deposition disease [CPPD] presented as osteoarthritis (OA). The dilemma with the overlapping presentation can be a challenging scenario to evaluate and treat. A comprehensive review of both radiographic and ultrasonographic imaging findings can be the key to connect the missing dots to address the complex clinical presentation as in the following case-report. We present a case of a 64-year-old male with symptoms of joint pain and history of being diagnosed as osteoarthritis while there was underlying CPPD, leading to perplexity and delay in diagnosis and management of CPPD.

Keywords

Calcium pyrophosphate disease, Pseudo-osteoarthritis.

Introduction

CPPD is microcrystalline arthritis caused by calcium pyrophosphate (CPP) crystals. Acute CPP crystal arthritis which has typical presentation with acute onset of monoarticular or oligoarticular arthritis is the most likely recognized form of this disease. It is the chronic CPP form which creates an intricate clinical pattern, presenting as a polyarticular form of arthritis that resembles OA or even seronegative rheumatoid arthritis. This creates uncertainty in diagnosis leading to potential challenges with treatment options especially when the joints involved are not the ones resembling OA. Involvement of the wrist joint, glenohumeral joint, elbows and spine are most commonly affected joints in this form of CPPD which has been historically referred to as "pseudo-OA" because of its resemblance to OA.

OA with CPPD is not an uncommon and yet often unrecognized form of symptomatic CPPD; for example, 20 percent of unselected patients examined at total knee joint replacement for OA can have CPP crystals in synovial fluid samples [1]. The recently approved 2023 ACR/EULAR classification criteria for CPPD include demographic, clinical and radiologic characteristics along with synovial fluid crystal analysis [2]. CPPD is often underdiagnosed; around 20% of unselected patients who are examined at the time of total joint replacement for OA of the knee have CPP crystals in their synovial fluid [3]. The expanding use of musculoskeletal ultrasound in the outpatient clinic will aid the diagnosis of CPPD. With ultrasonography both sensitivity and specificity are high for detecting CPP deposits criteria for CPPD suggested that OA pattern was observed in 55.5% of individuals fulfilling the criteria [5].

Case Report

A 64-year-old male presented to the rheumatology clinic for the first time with chronic joint pain. He mentioned experiencing pain for two years primarily in his left knee and wrist, with some involvement in the left ankle and right shoulder. Previously diagnosed with degenerative arthritis in his spine, elbow, knee, and

wrist, he had grown accustomed to the discomfort. However, he emphasized the cumulative impact on his daily activities and well-being. Notably, he expressed concern about a gradual decrease in strength in his left upper extremity, particularly the wrist. While initially attributing this weakness to a possible stroke (never formally diagnosed), he now believed the joint pain to be the major contributing factor.

The patient reported experiencing persistent joint pain that was often inadequately controlled by his current pain management regimen. Examination of the musculoskeletal system revealed left wrist swelling with mild tenderness to palpation. Additionally the range of motion in the left wrist was limited and he demonstrated decreased hand grip strength.

The patient's reported history of multiple emergency department visits for joint pain and multiple falls warranted a more comprehensive review of his medical record. It was noted that during his evaluation for joint pain 2 years ago, a wrist radiograph performed revealed well circumscribed lucencies within the scaphoid bone consistent with geodes (subchondral cysts). Osteopenia with chondrocalcinosis of triangular fibrocartilage complex (TFCC) and calcification of scapholunate ligament was also seen. The knee radiograph was suggestive of patellofemoral joint space narrowing and preservation of tibiofemoral joint space along with chondrocalcinosis. Patellar enthesophytes were seen (Figure 1).

A subsequent computerized tomography (CT) scan of the wrist demonstrated subchondral cystic changes, predominantly affecting the distal ulnar metaphysis and lunate bone. Additionally the CT identified intercarpal and triangular fibrocartilage complex chondrocalcinosis and chronic cortical disruption. Degenerative joint changes in multiple locations including elbow and knees were noted. Rheumatologic workup with autoimmune panel was pan-negative.

In light of the prior imaging findings, an office-based ultrasound examination of the left wrist was performed to further evaluate the suspected cystic changes and cortical disruption. It revealed multiple punctate calcification in the TFCC. Joint effusion between the radiolunate and scapholunate joints was also recognized (Figure 2).

These findings, in conjunction with the patient's history of multiple joint pain and the previously noted degenerative changes on past imaging studies, suggested a diagnosis of CPPD affecting the wrist joint and the left knee joint simulating OA. The degenerative changes observed were consistent with features of OA that can sometimes accompany CPPD and present as what

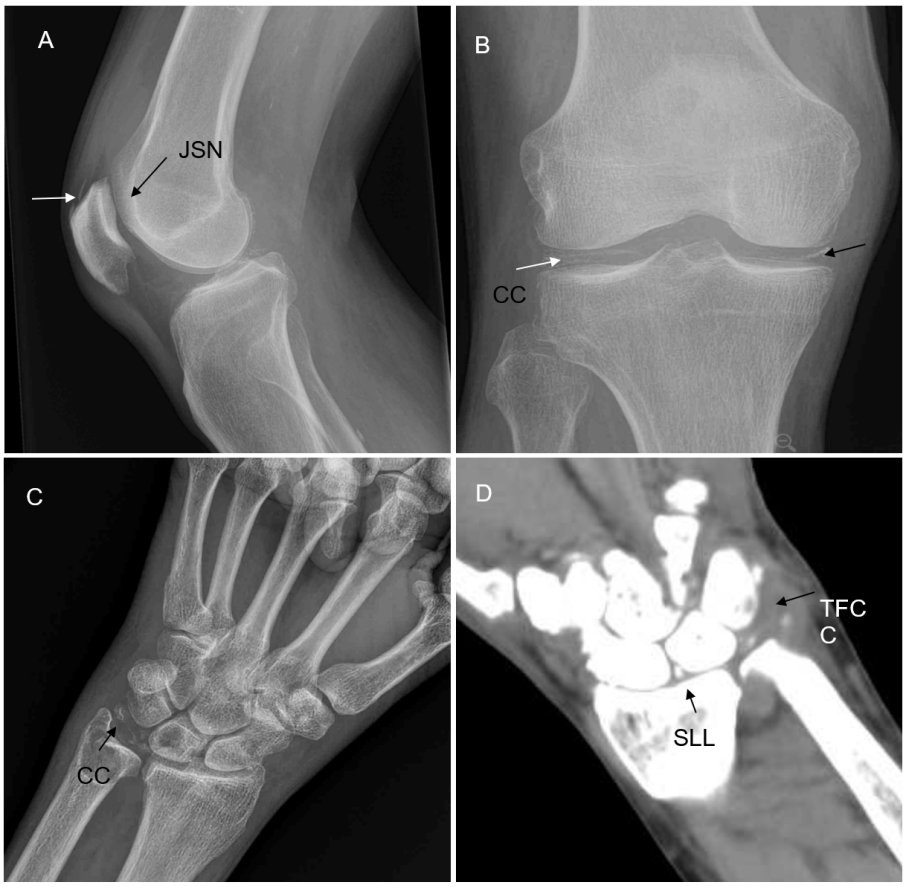

Figure 1: [A] Radiograph of left knee lateral view: Patellofemoral joint space narrowing [JSN] as shown by black arrow, enthesophyte (white arrow). [B] Radiograph AP view knee: Chondrocalcinosis (CC) and linear calcification in the tibiofemoral joint (black arrow). [C] Radiograph left wrist: chondrocalcinosis (CC). [D] CT left wrist: scapholunate ligament calcification (SLL) and triangular fibrocartilage complex (TFCC) calcification.

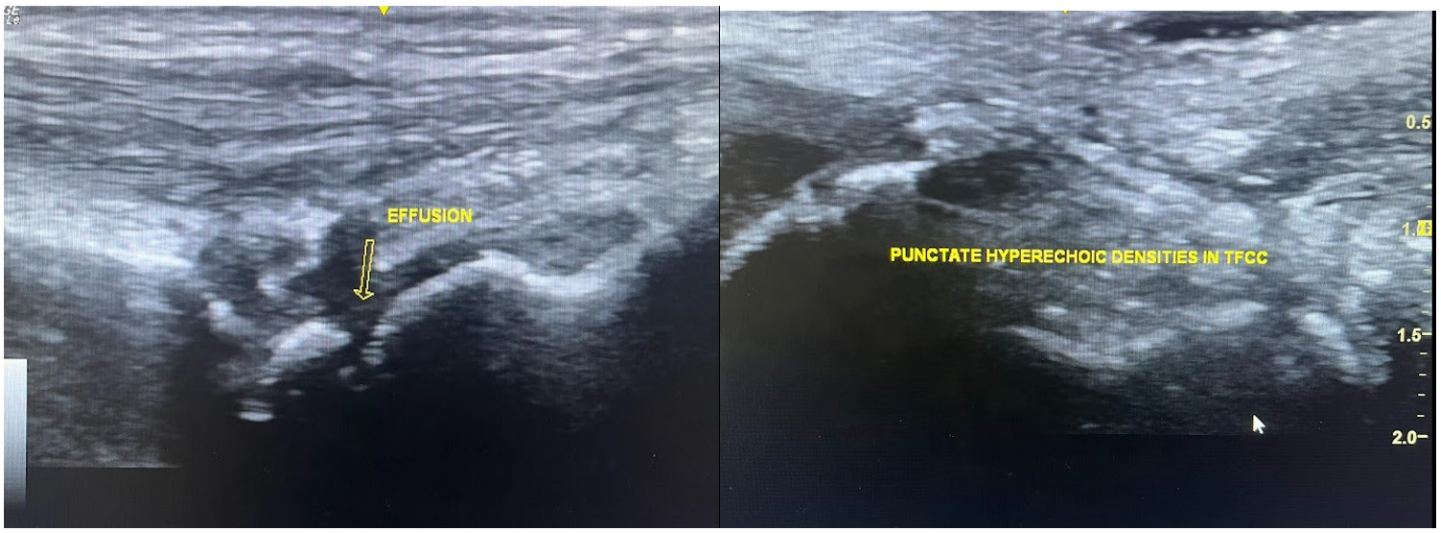

Figure 2: [A] Ultrasound [US] image of the dorsal aspect of the radiocarpal joint showing effusion between the radiolunate joint. [B] US image of the ulnar aspect of the wrist shows echogenic foci with punctate calcification in the TFCC.

was formerly called the 'pseudo-OA'. Following confirmation of CPPD diagnosis, consideration was given to initiating colchicine prophylaxis to potentially reduce the frequency of future acute attacks. Identifying the cause of the patient's asymmetric joint pain allowed for the development of a specific targeted treatment plan.

Discussion

CPPD is arthritis caused by CPP crystals. CPPD, which was referred to as pseudogout, had its roots in early medical descriptions of a condition that mimicked gout in terms of symptoms with acute arthritis. In 2011, the European League Against Rheumatism task force agreed that 'CPPD' should be the umbrella term that includes acute CPP crystal arthritis, OA with CPPD, and chronic CPP crystal inflammatory arthritis [6]. Acute CPP crystal arthritis is the most recognized form of CPPD and presents with a sudden onset of inflammation as monoarticular or oligoarticular arthritis. Chronic CPP crystal arthritis manifests in several forms. Common presentation involves a polyarticular arthritis that can closely resemble OA. While chronic CPPD arthritis can mimic OA, several features help distinguish them: unlike the consistent pain of OA, CPPD can cause episodes of increased inflammation and pain and can lead to more severe joint damage. The involvement of joints such as the glenohumeral joint, the wrist, and the metacarpophalangeal joints, which are not often affected by typical OA, should lead one to suspect the presence of CPPD disease. In OA, the most frequent radiographic abnormalities are asymmetric joint space narrowing, osteophyte formation, subchondral sclerosis, bone remodeling and subluxation [7]. CPP that occurs in joints with pre-existing degenerative changes can be a diagnostic challenge. Radiographic characteristics with chondrocalcinosis (linear or punctate calcification of the hyaline and fibrocartilage) is the characteristic finding on plain radiography that is associated with CPPD. Scapholunate calcifications showed a sensitivity of 98.2% and a specificity of 61.1% for CPPD [8-13]. CPPD in the knee is associated with scalloping of the anterior femoral cortex at the level of the patella [12] rather sparing the tibiofemoral and fibulofemoral compartments.

Ultrasonography can be a useful diagnostic modality in CPPD. It is important to emphasize that the presence of chondrocalcinosis supports the diagnosis of CPPD but the absence of it, does not exclude the diagnosis. New imaging technologies, including advanced MRI techniques, diffraction-enhanced synchrotron imaging, and dual-energy CT, hold promise for improved diagnostic accuracy [8]. CPPD is most accurately diagnosed by the finding of positively birefringent, rhomboid-shaped crystals in synovial fluid from the affected joint.

Conclusion

This case highlights the importance of considering CPPD in the differential diagnosis of patients presenting with OA-like symptoms. While the initial diagnosis might be OA, a careful evaluation can reveal underlying CPPD. This distinction is particularly valuable in scenarios where differential diagnosis is challenging, as it may carry important prognostic and therapeutic implications. The treatment of acute attack of crystal arthritis is with use of NSAIDs along with the possible use of intra-articular glucocorticoids. The daily use of oral colchicine at a low dose (0.6 to 1.2 mg) may be useful in reducing the frequency of acute attacks [9]. There is some data which support the use of hydroxychloroquine in patients with CPPD disease [10]. Interleukin-1 receptor blocking therapy could be taken into account in patients with acute and chronic CPPD with comorbidities, to reduce hospitalization times, or in patients where NSAIDs, colchicine and steroids are ineffective or contraindicated [11].

The limitations of current treatment options and diagnostic hurdles necessitate a more meticulous approach to managing suspected CPPD. This includes a heightened focus on evaluating joint pain in atypical locations and presentations, coupled with a comprehensive evaluation through synovial fluid analysis and advanced imaging modalities.

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