Case Report

Musculoskeletal Ultrasound Diagnosis in Calcium Pyrophosphate Dihydrate Crystal Deposition Disease

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ABSTRACT: Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease is extremely versatile. We present the case of a 52 years old patient referred for inflammatory pain of the left knee and ankle of recent onset with intense inflammatory syndrome that mimics rheumatoid arthritis. Musculoskeletal ultrasound was able to detect effusion, hyperechoic crystals within the cartilage and fibrocartilage, inside tendons, suggestive for CPPD disease and to guide fluid aspiration as well as in sustaining a rapid diagnosis corroborating images suggestive for CPPD disease to biological data – hypercalcemia and hyperparathyroidism in finding the predisposing condition, the parathyroid adenoma.

KEYWORDS: *CPPD disease, musculoskeletal ultrasound, crystals, primary hyperparathyroidism*

Introduction

Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease frequently referred in a simplistic manner as chondrocalcinosis is a metabolic disorder determined by calcium pyrophosphate dihydrate crystal precipitation at the level of intra and periarticular structures (hyaline cartilage, fibrocartilage, synovial membrane) often manifested through latent joint structural lesions rather than symptomatic arthropathy. [1] This extremely versatile disease makes positive and differential diagnosis quite difficult as usually mimics or may coexist with gout, rheumatoid arthritis, septic arthritis or primary osteoarthritis.

Case presentation

We report the case of a 52-years-old patient MA referred for inflammatory pain of the left knee and ankle started for 48 hours. The patient has a personal history of chronic symmetric polyarthropathy with frequent episodes of joint pain, swelling and local erythema at the knee, ankle, wrist and metacarpophalangeal joints developed over the last year associated with intense biological inflammatory syndrome and prior diagnosed as seronegative rheumatoid arthritis treated with leflunomide and non-steroidal antiinflamatories. Arthritic flares occurred over an intercritic period that resembled mostly chronic degenerative arthritis with rapid onset of inflammatory flares over 24 hours followed by remission of symptoms after 5 - 7 days of steroidal and NSAID therapy.

Clinical examination at presentation revealed an underweight patient, with low grade fever, fatigue, confused, synovial and dorsal extensor tenosynovial swelling of both wrists, second and third metacarpophalangeal joints together with distal hand osteoarthritis, pain and crepitation at glenohumeral passive mobilisation, left knee and ankle with peryarticular erythema, warmth and local swelling with pain and limited motion upon active and passive mobilisation, muscle weakness, upper abdominal pain, nausea and vomiting, constipation.

Biological profile at admission revealed intense inflammatory syndrome ESR 110mm/hour, CRP 10.2 mg/l, hemoglobin 9.5g/dl, leukocytosis up to 12000/mm^3, absent rheumatoid factor, negative anti-CCP antibody testing, elevate serum calcium concentration 16.5mg/dl, transaminases and creatinine within normal range. Hypercalcemia rises the suspicion of hyperparathyroidism so intact parathyroid hormone (PTH) is tested.

Before PTH results were available imaging studies were performed. Knee X-ray showed narrowing of the joint space, sclerotic borders, osteophytes. Ankle X-ray: decreased bone mineralisation and osteophytes.

Musculoskeletal ultrasound was performed with a Prosound α7 scanner (Aloka, Tokyo, Japan), using a multifrequency linear array transducer (7-18MHz).

Knee ultrasound scanning showed linear and punctate deposits within the femoral hyaline cartilage present both in suprapatellar longitudinal and transverse view (Fig.1a), hyperechoic spots within the distal portion of the patellar tendon (Fig.2a), synovial proliferation protruding into the joint effusion consistent with "pseudo rheumatoid arthritis" (Fig.1b),

aggregates of medial meniscal calcification (Fig.2b). Presence of effusion allowed ultrasound-guided aspiration with fluid analysis. [2]

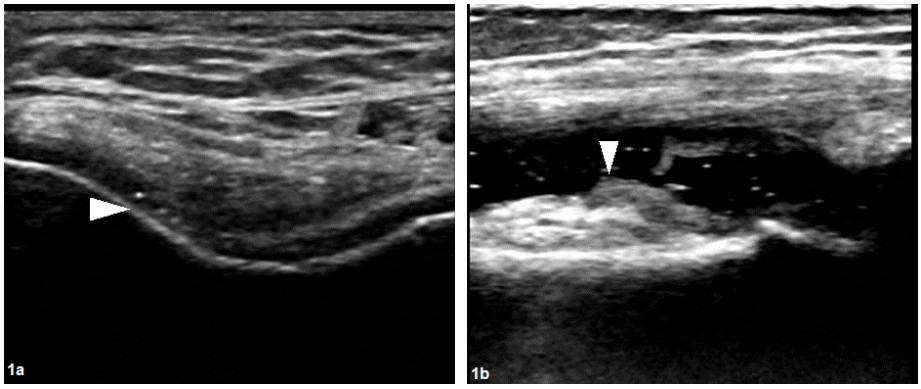

Fig.1a.Knee suprapatellar transverse view with isolated hyperechoic spots within the hyaline cartilage.
Fig.1b.Knee suprapatellar lingitudinal view with joint effusion and synovial proliferation

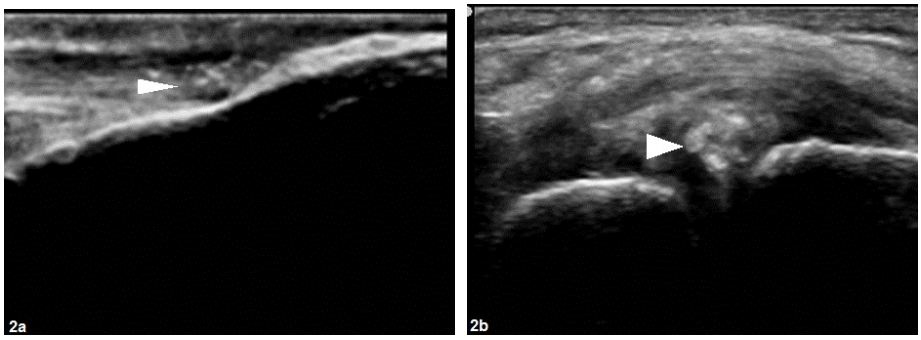

Fig.2a Intratendinous hyperechoic spots in the distal insertion of the patellar tendon, lingitudinal view.
Fig.2b. Medial longitudinal view of the knee with aggregates of meniscal calcifications.

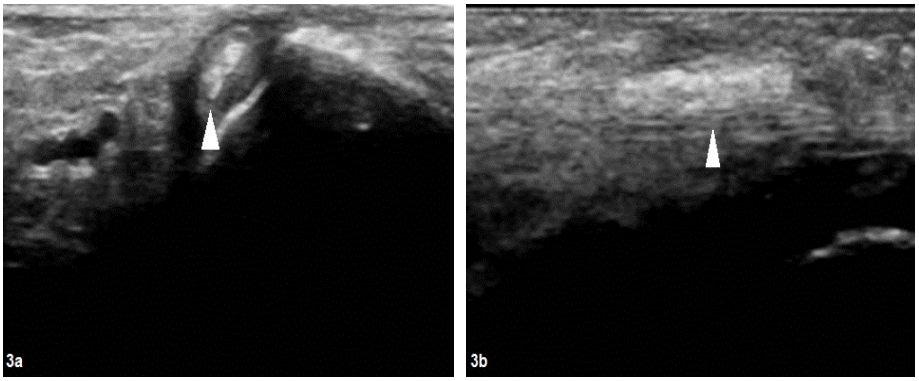

Fig.3a Ankle joint medial transverse view with large calcification inside tibialis posterior tendon.
Fig.3b. The same calcification inside tibialis posterior tendon in medial ankle joint longitudinal view.

Ankle ultrasound showed moderate effusion in the tibiotalar joint, gross intratendinous hyperechoic bands visualised in two perpendicular planes at the level of tibialis posterior tendon (Figure 3a, 3b).

Hand ultrasound identified calcifications within the triangular fibrocartilage of the wrist and cortical bone irregularities consistent with osteophytes at metacarpophalangeal joints (Figure 3a).

Shoulder ultrasound showed multiple ovalar, linear calcifications of the rotator cuff mostly in supraspinatus tendon with tendon dishomogenity and partial-thickness tears, probably mixed crystals – CPP and basic calcium phosphate BCP. [3]

Subsequently, small part ultrasound for thyroid and parathyroids was performed and detected the presence of a hypoechoic homogeneous ovalar structure of 2.14cm,

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confirmed through computed tomography examination.

Intact PTH values of 120.2pg/ml sustained the presence of the parathyroid adenoma while fluid analysis by compensated polarised light microscopy showed parallelepiepedic predominantly intracellular crystals with weak positive birefringence.

Final diagnosis: Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease. Left knee and ankle acute arthritis CPPD-induced. Primary hyperparathyroidism. Parathyroid adenoma. Osteoarthritis with CPP deposition – knee, ankle, hand osteoarthritis. [4]

Discussion

The particularities of the case are represented by chronic polyarticular evolution with acute arthritic flares symmetrical in evolution at the wrist, metacarpophalangeal, knee, ankle joints with intense inflammatory syndrome that mimics rheumatoid arthritis, lack of information after conventional radiography, the advantages of ultrasound in the ability to detect effusion, synovitis, hyperechoic crystals within fibrocartilage, linear or punctate aggregates inside tendons, hyperechoic bands within the cartilage or homogeneous hyperechoic nodular or ovalar deposits in bursae suggestive for CPPD disease and to guide fluid aspiration especially when fluid analysis depends on a difficult processing technique and a well-trained observer. [5] Musculoskeletal ultrasound was the first available method to evaluate and recognize diagnosis corroborating images suggestive for CPPD disease to biological data – hypercalcemia and hyperparathyroidism in finding the predisposing condition, the parathyroid adenoma. It represents an increasingly used imaging tool in the diagnosis and evaluation of rheumatic diseases that allows multiplanar dynamic assessment in real time, guiding local procedures, with the advantage of lack of radiation, low cost, easily accepted by the patient and with good reproducibility. [6]

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