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CASE REPORT

Calcium Pyrophosphate Dihydrate Crystal Deposition Disease–A Case Report

FONSECA A *, CRASTA J**, SWAMY M***

ABSTRACT

We report here, a case of calcium pyrophosphate dihydrate deposition disease, presenting as progressive bilateral knee pain that simulated osteoarthritis. A 52 year old male patient presented with gradually progressing bilateral knee pain of 2 year's duration. Plain X-ray films showed osteoarthritic changes and osteochondral loose bodies. The nodular crystal deposits with the characteristic features of CPPD crystals in the synovium at histology, aided in arriving at the correct diagnosis.

Key Words: pseudogout, calcium pyrophosphate dihydrate deposition

* MBBS, DCP,Postgraduate
**MD, DNB, Assosciate Professor
***MS, Professor,Department of Pathology and OrthopedicsSt.John`s Medical College Bangalore-34(India)
Corresponding Author:
Julian Crasta, MD, DNB
Associate Professor
Department of Pathology
St.John`s Medical College
Bangalore-560034
Karnataka
E-mail:ravinjul35@hotmail.com

Introduction

Calcium pyrophosphate dihydrate crystals (CPPD) were first described by Mc Carty et al in 1962 [1],[2]. The crystals were identified in the synovial fluid of patients who had gout like symptoms, without sodium urate crystals. Later, this entity was designated as 'pseudogout' and from then on the terminology has been applied to conditions which are characterized radiologically by prominent multifocal calcification in the cartilages of joints and the intervertebral disc space [3].Recently, the disease has been designated as CPPD crystal deposition disease and the term encompasses pseudogout, chondrocalcinosis and pyrophosphate arthropathy [4].

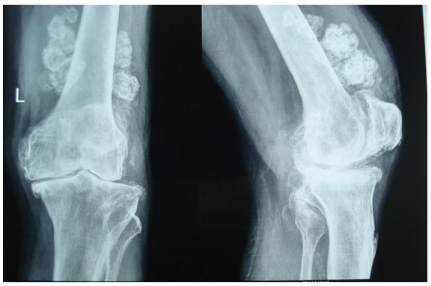
The clinical manifestations of calcium pyrophosphate dihydrate crystal deposition disease vary widely. Most of the cases are asymptomatic and are detected incidentally. Calcium pyrophosphate dihydrate deposition occurs in articular and periarticular tissues. In symptomatic cases, the clinical presentation mimics gout, osteoarthritis and rheumatoid arthritis. Rarely may it present as solitary tophaceous pseudogout [5].

Numerous clinical, radiological and epidemiological studies of calcium pyrophosphate dihydrate deposition disease have been reported. But few reports exist on its pathological findings, which have not been adequately described. We report here a case of calcium pyrophosphate dihydrate crystal deposition with radiological and pathological findings of the synovium.

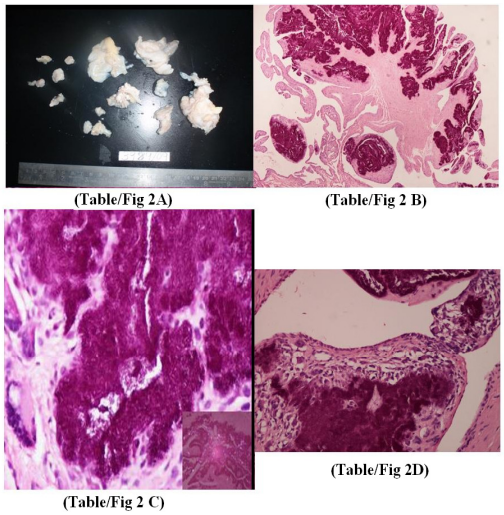
Case Report

A 52 year old male patient presented with gradually progressing bilateral knee pain of 2 year's duration . On examination,the left knee joint was swollen. Joint line tenderness with crepitus was noted on movement and a varus deformity was detected. No distal

neurovascular deficits were noted.Plain X-ray films showed osteoarthritic changes with osteochondral loose bodies [Table/Fig 1] . The clinical diagnosis was made as osteoarthritis with synovial chondromatosis and synoviectomy was done. Gross examination revealed two flat fragments of synovial tissue measuring 9x7x0.5cm and 7x3x0.5 cm exhibiting surface papillary excrescences with chalky white deposits [Table/Fig 2] (Fig 2A). Multiple bony fragments ranging in size from 1.5x1.5x0.6cm to 0.7x0.6x0.6cm were seen. Light microscopic examination revealed hyperplastic synovial lining membrane with prominent synovial lining [Table/Fig 2](Fig 2B). Beneath the synovial lining nodular deposits of basophilic calcified crystalline tophaceous deposits were seen. These crystals were colourless, rhomboid; needle shaped and exhibiting weak birefringence [Table/Fig 2] (Fig 2C).). The inflammatory response surrounding the deposits composed of lymphocytes and histiocytes, with occasional foreign body giant cells. . Synovial chondrometaplasia with nuclear atypia of chondrocytes was observed [Table/Fig 2] (Fig 2 D).



(Table/Fig 1) X-ray knee joint showing osteoarthritic changes with articular calcification and osteochondral loose bodies



(Table/Fig 2A) (Table/Fig 2 B)

(Table/Fig 2 C) (Table/Fig 2D)

(Table/Fig 2)

(Table/Fig 2A)Synovial membrane exhibiting papillary excrescences with chalky white deposits

(Table/Fig 2 B and C) basophilic calcified deposits with characteristic crystals of CPPD showing weak birefringence(inset)

(Table/Fig 2D) inflammatory response with foreign body giant cells.

Discussion

Intraarticular CPPD calcium pyrophosphate dihydrate crystal deposition is a common idiopathic condition of the elderly and is present at the time of death in about 5% of adults [2]. Though it is asymptomatic in many, others may be severely affected and it clinically presents commonly as osteoarthritis and gout [3]. It is important to distinguish calcium pyrophosphate dihydrate arthropathy from other arthropathies that it mimics, because the clinical course, prognosis and management may be quite different in all these presentations..

Calcium pyrophosphate dihydrate crystal deposition disease mainly affects middle aged and elderly people, with a female preponderance [5]. Pseudogout most often involves the knee and less commonly the wrist or ankle with the sudden onset of severe pain, swelling and redness [6]. However tophaceous pseudogout is common in the temporomandibular joint. Calcium pyrophosphate dihydrate crystal deposition disease is known to be associated with hyperparathyroidism, hypoparathyroidism, hemochromatosis and hemosiderosis [7].

The clinical features that should alert one to the likelihood of CPPD arthropathy include the following: an unusually severe or destructive arthropathy, a history of other joint involvement or of a previous joint operation, clinical evidence or a history of a disorder known to be associated CPPD and a family history of joint disease. There may also be evidence of chondrocalcinosis or other distinctive radiological signs of CPPD [3]. Our patient was an elderly male who presented with symptoms of osteoarthritis with no other associated clinical conditions. Approximately 20% of the patients with CPPD calcium pyrophosphate dihydrate have hyperuricaemia and some have coexistent gout. Though clinically CPPD crystal deposition disease mimics various arthropathies the gross and histological features are characteristic. Grossly, the chalky white deposits on the synovial villi produce an appearance which can be likened to snow covered whiskers [3]. Pseudogout exhibits nodular deposits of CPPD crystals which are similar to tophaceous gout and tumoral calcinosis. The CPPD crystals range in size from 2 to 40 micrometres. They are pleomorphic and are mostly rhomboid shaped,although long or short rods and small squares are also seen [8],[9]. However, the sheaf like arrangement is never observed,which distinguishs them from sodium urate crystals. Unlike urate crystals, CPPD crystals are retained in tissues after fixation with formaldehyde and after processing with alcohol. By polarized light microscopy, these crystals are found to have weaker birefringence in contrast to the stronger birefringence of urate crystals. The calcified material in tumoral calcinosis is amorphous granular and lacks crystalline material. The exact nature of these crystals can be determined by radiographical diffraction or electron probe analysis [9],[10].

The chondroid metaplasia with cytological atypia which is seen in calcium pyrophosphate dihydrate crystal deposits may superficially resemble chondrosarcoma. Particularly, in decalcified sections from which CPPD calcium pyrophosphate dihydrate crystals are lost, atypical features in metaplastic chondrocytes with the presence of myxoid stroma may lead to the histological misdiagnosis of chondrosarcoma. However the presence of crystals, histiocytes and foreign body giant cells helps in distinguishing it from chondrosarcoma [5].

The potential for overlooking or misinterpreting cases of CPPD is heightened by the frequent lack of clinical suspicion or by inadequate communication between the surgeon and the pathologist. Therefore, pathologists who examine articular tissue must maintain a keen awareness of the possibility of CPPD whenever they receive an arthroplasty or synovial biopsy specimen. On gross examination of an arthroplasty specimen, the presence of osteochondral loose bodies and the presence of characteristic crystals at microscopy should alert the pathologist towards the correct diagnosis.

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