

## Case Report

# Rare case of pseudogout in the scapular region

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### ABSTRACT

Calcium pyrophosphate dihydrate crystal deposition disease (CPPD) also referred to as pseudogout is an inflammatory arthritis produced by the deposition of calcium pyrophosphate crystals in the synovium and periarticular soft tissues. This report documents a case of a 41 year old female who presented with a painful swelling in the back. MRI revealed a complex swelling with lobulation of size 7.4×7.4×7 cm in the posterolateral aspect of chest wall just superficial to the ribcage. A surgical excision biopsy of the swelling was done and patient improved symptomatically. Serum uric acid levels were measured and found to be within limits.

**Keywords:** CPPD, Periarticular, Pseudogout

### INTRODUCTION

Pseudogout is a widely recognized form of acute calcium pyrophosphate deposition (CPPD) disease, presenting as acute arthritis of monoarticular or oligoarticular joints.<sup>1</sup> Although articular cartilage and soft-tissue calcification secondary to CPPD deposition is a common radiographic finding, tophus formation has seldom been described in this condition.<sup>2-4</sup> It is rare under the age of 55, but occurs in 10-15% of people between the ages of 65 and 75 years and in 30-60% of those over 85.<sup>5</sup> The pathogenesis of pseudogout requires formation of calcium pyrophosphate crystals in the pericellular matrix of cartilage.<sup>1</sup> Once generated, these deposits induce inflammation but also have direct catabolic effects on chondrocytes and synoviocytes, resulting in production of metalloproteinases and prostaglandins that can destroy the pericellular matrix.<sup>1,6,7</sup> The calcium crystals additionally affect the mechanical properties of the cartilage and may accelerate joint damage.<sup>1,8</sup> Common signs include erythema, warmth, and swelling of the affected joint, with a clinical presentation similar to gout or septic arthritis.<sup>9</sup> Patients often present with fevers,

chills, and constitutional symptoms lasting weeks to months.<sup>1</sup> The knee (hyaline cartilage and menisci) is by far the most common site, followed by the wrist (triangular fibrocartilage) and pelvis (symphysis pubis).<sup>5</sup> In many patients chondrocalcinosis is asymptomatic and an incidental finding on X-ray examination, but others present with an acute inflammatory arthritis (pseudogout) or a chronic inflammatory arthropathy superimposed on a background of osteoarthritis, especially at the knee, associated with joint damage and functional limitation.<sup>10</sup> Pathological examination demonstrates positively birefringent rhomboid-shaped crystals in the synovial fluid of the affected joint.<sup>1</sup> Therapy is directed toward reducing inflammation and includes intraarticular glucocorticoids, systemic glucocorticoids, oral colchicine, and nonsteroidal antiinflammatory drugs.<sup>10</sup>

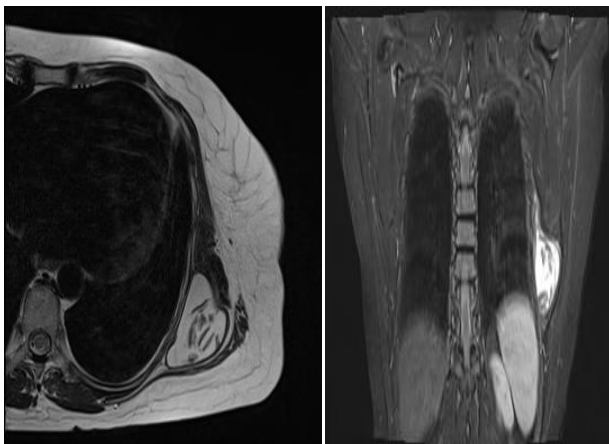
### CASE REPORT

A 41 year old female came with complaints of a swelling on the back noticed 6 months back. The swelling progressively increased in size and was associated with back pain for the past 10 days. History of multiple joint

pain for the past 1 year on symptomatic treatment. No history of discharge from the swelling, fever, trauma, cough with expectoration, breathlessness, restriction of movements or loss of weight.

On clinical examination, a 7\*6 cm soft, mobile, palpable swelling was present in the intramuscular plane of the left scapular region, slip sign was positive, non transilluminent. No other swellings were palpable, no warmth or tenderness, no cough impulse.

On investigation, baseline investigations were within normal limits. Serum uric acid levels were found to be within the normal range. MRI revealed a complex swelling with lobulation of size 7.4x7.4x7 cm in the posterolateral aspect of chest wall just superficial to the ribcage (Figure 1).



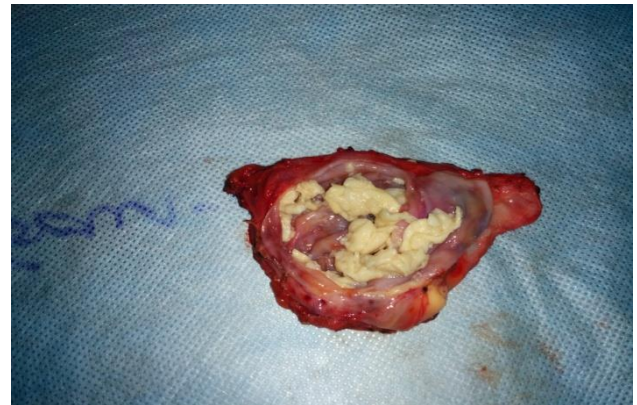
**Figure 1: MRI reveals a complex swelling with lobulation of size 7.4x7.4x7cm in the posterolateral aspect of chest wall just superficial to the ribcage.**

Small mediastinal lymphadenopathy noted. Fine Needle Aspiration Cytology was negative for malignant cells. A surgical excision biopsy of the swelling was done under general anaesthesia (Figure 2).



**Figure 2: Surgical excision biopsy of the swelling.**

Post-operatively the patient improved symptomatically. Drain was removed on day 2. Wound appeared to be healthy. Histopathological examination was reported as pseudogout.



**Figure 3: Tophaceous deposits-chalky white appearance.**

**Table 1: Classification of CPPD disorders.**

Groups	Disorder
<b>Group A</b>	Pseudogout
<b>Group B</b>	Pseudorheumatoid
<b>Group C and D</b>	Pseudoosteoarthritis
<b>Group E</b>	Lanthanic/asymptomatic
<b>Group F</b>	Pseudoneurotrophic
<b>Others</b>	Tophaceous, spinal CPPD deposition, crowned dens syndrome, spinal stenosis

Mukhopadhyay, Sudiptamohan & Guha, Abhijit & Perera, Anthony (2011). Monoarticular pseudogout of the hip presenting as septic arthritis: a case report. Acta orthopaedica et traumatologica turcica. 45. 200-2. 10.3944/AOTT.2011.2318.

**DISCUSSION**

Onset of pseudogout occurs typically in the elderly at an average age of 72 years.<sup>11</sup> Symptomatic CPPD, however, has been reported to be more common in females.<sup>12</sup> It is known that the probability of CPPD increases with age.<sup>13</sup> There are valid associations with hypophosphatasia, hypomagnesaemia, and hyperparathyroidism, but the association with hypothyroidism is controversial.<sup>14</sup> Clearly, an abnormality of pyrophosphate and calcium metabolism is likely to be the precipitating factor.

Clinical studies have shown that pyrophosphate levels are raised in patients with CPPD crystal deposition disease, possibly due to over-production, but why this occurs is unclear. In hypophosphatasia, the predisposing factor is thought to be impaired degradation of pyrophosphate due to mutations in the genes that encode ALP.<sup>5</sup> Osteoarthritis is thought to predispose to CPPD crystal deposition disease because of a reduction in the amounts of proteoglycan and other natural inhibitors of crystal formation in the abnormal cartilage.<sup>5</sup>

Investigations will include examination of synovial fluid using compensated polarized microscopy to demonstrate CPPD crystals and to distinguish from gout.<sup>5</sup> The aspirated fluid is often turbid and may be uniformly blood stained reflecting the severity of inflammation.<sup>5</sup> Since sepsis and pseudogout can exist, Gram stain and culture of the fluid should be performed to exclude sepsis, even if CPPD crystal are identified in synovial fluid.<sup>5</sup> X-rays of the affected joint may show calcification in hyaline cartilage and/or fibrocartilage, although absence of calcification does not exclude the diagnosis.<sup>5</sup> Screening for secondary causes should be undertaken, especially in patients who present under the age of 25 and those with polyarticular disease.<sup>5</sup>

A pseudogout differential diagnosis may include Wilson's disease, hemochromatosis, hemophilia, hypothyroidism, arthritis (rheumatoid, infectious, traumatic, and degenerative), amyloidosis, acromegaly, diabetes mellitus, ochronosis, and gout.<sup>15</sup> A tophaceous pseudogout differential diagnosis may include synovial chondromatosis, infectious arthropathies, cholesteatoma, acute otitis, osteoma of the mandible, and parotid neoplasms, as well as malignant lesions, such as chondrosarcoma and chondroid chordoma, and benign lesions, such as chondromas and chondroblastomas.<sup>15-17</sup> It is essential to rule out tophaceous pseudogout from the differential diagnosis to avoid unnecessary radical treatment for malignant conditions.

Joint aspiration would provide symptomatic relief. Patients with persistent symptoms can be treated with intraarticular corticosteroids, colchicine or NSAIDs. Colchicine at a dose of 0.6 mg once or twice daily may be effective as a prophylactic measure to reduce the number of attacks in a year, especially in patients who experience three or more attacks a year.<sup>18</sup> Unlike gout, however, there are no hypouricemic equivalents to improve the long-term control of acute attacks or to prevent or reverse CPPD disease. Early active mobilisation is important as well. The outcome of patients with CPPD disease is influenced by genetic predisposition, extent of crystal deposition and joint degeneration, and aggravating factors from the underlying associated diseases. A study that followed 104 patients with pyrophosphate arthropathy for a mean of 4.6 years found that patients presenting with acute attacks have a good prognosis.<sup>19</sup> They also found that some patients did not have progressive disease.

## CONCLUSION

Finally, tophaceous pseudogout appears to be a local rather than generalized process. Except for the involved joint, almost no crystal deposition is seen in other joints, and the clinical syndrome is confined to the affected joint. Thus, tophus formation, although rare, should be included in the spectrum of clinical and radiographic manifestations of CPPD crystal deposition disease.

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