A CALCIFIED INTRAARTICULAR MASS IN A MAN WITH SEVERE SHOULDER OSTEOARTHRITIS: WHAT ARE THE IMPLICATIONS?
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SUMMARY
Rapidly progressive osteoarthritis in the shoulder in the elderly is often associated with chronic rotator cuff calcifications and damage and with apatite crystals identifiable in the joint fluid. The key roles of the crystals and rotator cuff lesions although suspected have been disputed. We describe a 57-year-old man with severe degenerative changes at the right shoulder and other joints. A calcified mass 2-cm in length was found on radiographs medially in relation to the proximal humeral diaphysis. At arthroscopy, the mass was confirmed to be in the joint and due to calcified synovium.

Biopsy revealed synovium with apatite like crystal clumps in this mass. Calcium pyrophosphate crystals were also found but in the cartilage only. This case with the apatite crystals only in synovium and with destructive arthritis without a complete rotator cuff tear raise the possibility that synovium as a primary site for apatite deposition might be important in the destructive arthritis. Management of this patient like many with rotator cuff tear arthropathy has been difficult.

Rapidly destructive osteoarthritis at the shoulder, much like that in the patient reported here, has been described under a variety of terms that suggest implications for pathogenesis. Neer et al used the term cuff tear arthropathy to describe glenohumeral degenerative arthritis and a rotator cuff tear in
twenty-six patients who had required a total shoulder replacement. McCarty et al described 4 elderly women with destructive arthropathy of the shoulder, large effusions, apatite crystals present in the joint effusions and massive tears of the rotator cuffs and coined the term Milwaukee shoulder syndrome. Dieppe et al suggested the terms apatite-associated destructive arthropathy and idiopathic destructive arthritis. Calcifications have been noted in the rotator cuff structures but have not been reported in the joint or synovium. We describe a patient with a similar destructive arthropathy, who had a calcified mass about 2-cm in length in the right shoulder, well visible by X-ray and arthroscopy, that was localized to synovium at arthroscopy. Since this patient did not have prominent rotator cuff disease, our case suggests that intraarticular crystals can be associated with difficult to manage progressive shoulder osteoarthritis without a prominent primary rotator cuff cause.

**CASE REPORT**

A 57-year-old right-handed caucasian man, presented with a 3-year history of chronic severe persistent pain of his right shoulder. Pain was present at rest, aggravated on motion and awakened him from sleep when he rolled over onto his right shoulder. Symptoms increased with rainy weather and he had stiffness that awakened him in the morning but could last all day. He had left shoulder pain, but not nearly as much as his right; he had also pain and swelling at the right wrist, bilateral second and third MCP joints, intermittent discomfort at the left wrist and occasional right knee pain.

Past medical history was significant for this arthritis. He had a history of right carpal tunnel syndrome documented on electromyogram in 11/97. A left wrist ganglion was removed in 1995 and he had severe heel pain that had been treated by several corticosteroid injections. Two years earlier, because of a car accident, he required fusion of L5-S1, and C3-T1. In 1997, the left wrist was fused due to narrowing of the radiocarpal joint, subchondral sclerosis and chondrocalcinosis. No examination for calcium pyrophosphate (CPPD) crystals in synovial fluid or tissue was done. He had no past history of overuse or neurological diseases. Family history was positive for hypertension.
Fig. (1): Radiography
Severe glenohumeral joint degeneration. Severe calcific deposits
Calcification below the humeral head. A calcified mass 2cm in length below the
humeral diaphysis. Irregularities of the glenoid. Mild osteopenia.

Fig. 2 (a): Arthroscopy, 1998
The joint was extremely arthritic with a very visible calcified mass filling the joint.
Specimen was obtained for biopsy.
Calcified. Synovium showed later (apatite) crystals.
Intra-articular biceps tendon.
Fig. 2 (b): Arthrosopy, 1998
The glenoid devoid of articular Cartilage.
Calcified cartilage (CPPD).

Fig. (3): Arthrosopy 1998
Aggregates of (apatite) crystals (alizarin S red).
General examination revealed blood pressure of 150/92 mmHg, weight 200 pounds, and height was 72". He had well healed scars at the neck, over the lumbo-sacral area and at the left wrist. He had Heberden's and Bouchard's nodes in both hands and Dupuytren's contracture on the left hand. The right shoulder had muscle atrophy, local warmth and erythema, severe tenderness over the anterior leading edge of the acromion, acromioclavicular joint and right biceps tendons, mild pain with forward flexion and moderate pain with external rotation and abduction as well as limited range of motion (ROM). Crepitus was present at the right shoulder. The left wrist was fused. The right wrist was swollen and had moderate tenderness and limited ROM. Right first, second through fifth metacarpophalangeal and right carpometacarpal joints were tender but not swollen. Lower limb joints were unremarkable.

Laboratory investigation included normal serum calcium, phosphate, creatinine, sedimentation rate, complete blood count, thyroid and intact parathyroid function hormones, negative rheumatoid factor, normal C3 and C4, and negative SSA, SSB and ANA.

A chest X-ray dated 11/13/1996 showed several calcific densities projecting over the right humeral head and just inferior to the proximal humerus. Right shoulder radiographs showed degenerative changes in the glenohumeral articulation, irregularities of the glenoid fossa inferiorly, mild osteopenia of the joint, and some calcification immediately below the humeral head. In addition, there was soft tissue calcification below the proximal humeral diaphysis medially measuring about 2cm in length (figure 1). Other X-rays showed extensive postoperative changes and fusion of the cervical and lumbar vertebrae and left wrist.

Pain was persistent, so arthroscopy was done in 1998. A calcified mass of synovial tissue was found partially filling the glenohumeral joint. There was also minor calcification over the posterior aspect of glenoid cartilage and the articular part of the glenoid and humeral head were devoid of articular cartilage (Figure 2). There was a 25% tear of the biceps tendon and 25% tear of the subscapularis tendon, and no visible tendon calcification. The subacromial space was markedly enlarged. Arthroscopic acromioplasty, biceps tendodesis and distal clavicle excision were done, and the specimens were sent for examination. The labrum cartilage demonstrated degenerative change with areas of positively birifringent CPPD crystals. In contrast, alizarin red S positive apatite like clumps were present through the fatty synovium (Figure 3). No CPPD were noted in this
Calcified Intraarticular Mass  Sonia Rashad et al.

tissue and inflammatory cells were not identified in the tissue. No synovial fluid was examined.

After arthroscopy the patient noticed improvement in his symptoms and was referred to physical therapy, but he quickly had increased pain so that he had to stop. Symptoms persisted so he received further local injections to the shoulder but with no benefit. The patient is now being considered for arthroplasty.

**DISCUSSION**

The rapidly progressive osteoarthritis, or Milwaukee shoulder syndrome, a condition that affects predominantly elderly woman, is characterized by predominant involvement of the dominant limb, severe glenohumeral joint destruction, and rotator cuff tear, and generally non-inflammatory effusions with evidence of crystal deposition in periarticular soft tissue. Characteristic radiographic findings have included superior migration of the humeral head with articulation with the overlying acromion, narrowing of the glenohumeral joint space, formation of osteophytes, and periarticular soft-tissue calcification. Radiographic evidence of a calcification is most common in the supraspinatus tendon part of the rotator cuff and interestingly has not been described in the joint despite the report of apatite crystals in joint fluid.

We describe an elderly man with similar arthropathy with a calcified mass about 2-cm in length in the right shoulder, well visible in X-ray and arthroscopically. As far as we know, this is the first report of such a calcified mass in a patient with a Milwaukee shoulder. Other authors described calcification only along periarticular soft tissues.

The cause of this unusual synovial apatite calcification and mass is unknown. The generation of extracellular inorganic pyrophosphate (ePPi) is considered crucial to the maintenance of cartilage function. However, excessive (ePPi) production may bear pathologic consequences because pyrophosphate forms the anionic portion of CPPD and elevated levels of ePPi favor the formation of CPPD crystals. Prostaglandin E1 and E2 were demonstrated to downregulate ePPi production. It is conceivable that inhibition of prostaglandin synthesis, as achieved with NSAIDs used to treat this and other OA patients, may act to elevate (ePPi) production and stimulate CPPD crystal formation. Whether these crystals are cause or the result of arthritis remains unanswered. The apatite like crystals might be an epiphenomenon resulting from changes in the matrix of damaged
cartilage with the crystals then are shed into the synovial fluid.\cite{9} We did not find apatite in cartilage in our patient but some cartilage had been denuded.

Ryan and Cheung have reviewed clinical and experimental evidence supporting a role for both CPPD and apatite crystals in cartilage degeneration in OA.\cite{7} Presence of apatite crystals correlates strongly with the severity of cartilage degeneration and the finding of large joint effusion. Apatite crystals, through activation of protein kinase C, phospholipase C, and other transcription factors can enhance tissue damage by at least two mechanism: stimulation of mitogenesis and prostaglandin synthesis in synovial fibroblasts and chondrocytes, and induction of synthesis and secretion of matrix metalloproteases (MMPs) by chondrocytes.\cite{7}

The possibility has been raised that repeated intraarticular steroid injections may precipitate crystals in the joint and may contribute to more rapid joint destruction. Weiss and Schumacher 1985 reported that the intraarticular injections in patients with Milwaukee shoulder can help maintain function,\cite{10} but they also are a recognized cause of periarticular calcification.\cite{11} In soft tissue, corticosteroids can cause atrophy and necrosis, and can facilitate crystal deposition.\cite{11,12} Might steroids have been a factor in this case? The freshly removed calcified mass was analyzed with polarized light and did not contain any obvious evidence of steroid crystals and the calcified mass predated the use of the most extensive of these injections.

The calcification of synovium appeared to be in fatty areas and not especially in macrophage rich sites. This must suggest the possibility of primary calcification at that location rather than sequestration in synovium after initial deposition in cartilage. Prominent mineralization in fatty synovium was also seen in patients with early osteoarthritis studies by Concoff and Kalunian (personal communication).

Although we emphasize the synovial apatite deposits in this case, the role of the crystals is still unclear. This patient like many others\cite{13} also had CPPD crystals in the cartilage and also had other joints involved. CPPD seemed likely to be factor in the other joints as chondrocalcinosis was seen at one wrist.

Management of this patient like many patients with cuff tear arthropathy has been difficult. Since the patient is elderly and nonoperative management has failed, a humeral hemiarthroplasty is the procedure of
choice as it can provide reliable relief of pain and improvement in function.[8]

REFERENCES