30. MILWAUKEE SHOULDER SYNDROME - AN INCIDENTAL FINDING BUT IMPORTANT CAUSE OF ACUTE SHOULDER ARTHROPATHY

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Introduction: McCarty et al first described the Milwaukee shoulder in 1981 after encountering four elderly women with similar patterns of destructive shoulder arthropathy, rotator cuff loss and large recurrent joint effusions containing basic calcium phosphate crystals. A variety of descriptive terms have since been adopted including apatite-associated destructive arthritis and idiopathic destructive arthritis of the shoulder and although it does most commonly affect the shoulders, it can affect any large joint. Knee disease predominantly involves the patellofemoral and lateral tibiofemoral compartments and results in valgus deformity as described by the McCarty group in 1986.2 A few years later, in 1990, reports for a total of 72 patients with the Milwaukee Shoulder Syndrome were amalgamated and comprise the largest dataset to date for this condition.3-7 Women were predominantly affected with a ratio of 4:1 and a mean age at onset of 72 years. Both shoulders were affected in 64% of patients and nearly a third had experienced trauma or joint overuse preceding the symptoms. From these historical reports we now know that patients typically present with an insidious onset (over months or years) of joint swelling associated with pain and restriction of movement. Examination reveals large shoulder effusions, often extending to the subdeltoid region, reduced active range of movement and pronounced joint instability which reflects the radiographic findings in this condition. There is characteristically superior subluxation of the humeral head, sometimes forming a pseudoarthrosis with the distal clavicle and acromion which is only possible due to widespread rotator cuff loss. Aspirated synovial fluid is typically haemorrhagic, non-inflammatory and can be positive for calcium apatite crystals. Treatment is largely supportive and oral anti-inflammatories and intra-articular corticosteroid injections can provide symptomatic relief. Its natural history remains unclear but cases seem to stabilise after 1-2 years with reduction of symptoms, joint effusions and radiographic progression.8

Case description: A 73-year-old lady presented initially to the Acute Medical Unit with a 6-month history of generalised joint pain and 3-stone weight loss following her husband's death. She used to be her husband's carer and 6 months prior to presenting to us she suffered a traumatic fall onto her right side when her husband fell onto her. A month after the fall she presented to A&E with right hip pain, she underwent hip and chest radiography and was reassured there was no fracture. However when the plain films were reviewed after discharge anterior subluxation of the right shoulder was noted and the patient was recalled urgently for assessment. By the time she presented to us, although she complained of widespread arthralgia affecting the hips, knees and right ankle, the worst joints were certainly the shoulders. Here she described quite longstanding symptoms (for at least 3 years) of pain and restriction of movement (right worse than left) with associated joint swelling. She had been furniture walking for the last month and this may indicate some overuse of both shoulder joints. On examination she was very frail and there were bilateral cool shoulder effusions (right larger than left) with significant restriction of movement. All other joints were unremarkable. We proceeded to aspirate the right shoulder and obtained 30ml of haemorrhagic non-inflammatory synovial fluid; no crystals were seen on polarised microscopy and cultures proved negative. Alizarin red staining was not undertaken. A malignancy screen was also done and was unremarkable. Plain films showed chronic significant bilateral anterior dislocation of the humeral heads indicative of severe rotator cuff destruction. There was also marked soft tissue swelling and calcification. Milwaukee shoulder syndrome was diagnosed and intra-articular corticosteroid injection provided for symptomatic relief. We will follow-up with the patient in due course and consider if further imaging and orthopaedic input are appropriate given both her frailty and the fact that shoulder prostheses tend not to be successful in joints where the rotator cuff has been lost.

Discussion: Milwaukee shoulder is a rare arthropathy characterised by the presence of intra-articular or peri-articular hydroxyapatite crystals and rapid rotator cuff as well as glenohumeral joint destruction. It differs from osteoarthritis given the widespread loss of cartilage, marked softening of the humeral head and presence of relatively small osteophytes; whereas osteoarthritis here is characterised by sclerosis of the humeral head, prominent osteophytes and an intact rotator cuff. It is also important to note that although our patient presented with pain and restriction of movement, the Milwaukee shoulder can often be painless and the joint deformity tends to be far worse than the degree of pain experienced – almost as if it were a Charcot joint (but with no evidence of neurological impairment). Alizarin red staining, although not used in our patient, is a simple and quick screening method to identify microcrystalline or non-crystalline calcium phosphate salts in synovial fluid. A historical study9 showed that this technique can pick up crystals missed by polarised light. Treatment, as mentioned above, is currently supportive and the focus is on symptomatic relief. Physiotherapy plays a major role in keeping the joint mobile and strengthening the surrounding muscles. Drug therapies such as non-steroidal anti-inflammatory drugs and intra-articular corticosteroids are widely used with varying degrees of benefit. Phosphacitrate has been shown to reduce crystal deposition in animal studies10 but, to date, no human studies have been possible and therefore this hypothesis remains untested.

Key learning points: The Milwaukee shoulder tends to come on insidiously, typically over months or years and is often preceded by trauma or joint overuse. Examination reflects rotator cuff loss with large joint effusions and humeral head subluxation. Synovial fluid alizarin red staining can be used to confirm the diagnosis and plain films are sufficient imaging and often depict significant humeral head subluxation as well as soft tissue calcification. We would like to highlight that as the syndrome often develops insidiously and the plain radiograph appearance is dramatic, the Milwaukee shoulder can easily be mistaken for other serious pathology, such as a traumatic subluxation, haemarthrosis or sepsis; however the clinical picture is characteristic allowing for a diagnosis based on clinical assessment and radiographs. This well documented but rare syndrome is an important differential diagnosis of shoulder disease presenting acutely.