Case 8100

Milwaukee Shoulder Syndrome
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Patient: 80 years, female

Clinical History:
An 80-year-old woman was referred to our institution due to worsening of long-standing pain and limited motion of the left shoulder. She reported a history of a fall on that shoulder six weeks earlier. The laboratory studies were unremarkable.

Imaging Findings:
An 80-year-old woman was referred due to worsening of long-standing pain and limited motion of the left shoulder. She reported a history of a fall on that shoulder six weeks earlier. The laboratory studies were unremarkable. Radiographs of the left shoulder revealed bony erosion with complete destruction of the humeral head, joint space narrowing, and a large soft tissue swelling with extensive amorphous calcifications (Figure 1). Computer tomography (CT) of the left shoulder was performed to assess the extent of bony destruction which was proven to also involve the left glenoid cavity, acromion, and coracoid process (Figure 2). Magnetic resonance imaging (MRI) examination was limited due to patient movement. Proton density (PD) transverse and T2 fat-suppressed paracoronal images were acquired. MRI confirmed the radiographic findings and better demonstrated the massive tear with retraction of the rotator cuff (Figure 3). Bacterial culture of the joint fluid was negative. Due to the extensive bony destruction no surgical treatment was undertaken. Subsequently the patient developed similar complaints on the right knee without history of trauma. The right knee radiographs demonstrated extensive bony destruction of all joint components with similar semiology to that depicted in the shoulder radiographs (Figure 4).

Discussion:
Milwaukee shoulder syndrome (MSS) is an uncommon and enigmatic entity. It is characterized by rapid and severe joint destruction, which associates rotator cuff tear and atrophic osteoarthritis, and bears resemblances to neuropathic and neuropathic-like arthropathies. MSS has been called rapidly progressive osteoarthritis, apatite associated destructive arthritis, cuff-tear arthropathy, rapid destructive arthritis, idiopathic chondrolysis, and senile hemorrhagic shoulder syndrome. This diversity of nomenclature to address the same disease relates to the controversy surrounding its pathogenesis which still remains unclear. The role of hydroxyapatite crystals on MSS has motivated much controversy, most likely being a marker of osteolysis or a secondary event rather than a primary cause. MSS preferentially affects female, elderly, and osteopenic patients. Although the shoulder involvement predominates, the knees and the hips are also frequently affected. Bilateralism is observed in the majority of cases. The knees and the shoulders are implicated together in 50% of cases. Other sites as elbows, ankles, wrists, and intertarsal joints are seldom affected. Most patients complain of pain, joint swelling, and movement restriction that can date from several months to years.
In 25% of cases, MSS is preceded by overuse or trauma, including recurrent subluxation, a fall, or a motor vehicle accident.

There is striking structural joint damage associated with rotator cuff tears and severe instability. Joint effusion is often voluminous, blood-stained (80%), and contains hydroxyapatite and less commonly pyrophosphate crystals. The best documented and mainstay technique for the diagnosis of MSS is plain film. Early radiographic changes consist of a high-riding humeral head due to rotator cuff tear, with mild subchondral bone sclerosis, and narrowing of the glenohumeral joint space, with little or no osteophytosis. These changes may stabilise or show minimal cartilage erosions for several years, followed by sudden and dramatic deterioration. The bones on both sides of the joint are severely damaged, with extension into the undersurface of the acromion, the coracoid process, and the distal clavicle. Pseudoarthrosis between the humeral head, coracoid, and acromion is common.

Although ultrasound can demonstrate the rotator cuff tear and marked joint distension with fluid and echogenic debris reflecting synovial proliferation, blood clots, calcified deposits, and osteolysis, it cannot accurately differentiate MSS from the more common rotator cuff disease related to osteoarthritis.

MRI can have a complementary role in this behalf, more objectively and extensively demonstrating the soft-tissue associated changes. It should be noted that it requires correlative analysis with the radiographic studies, to avoid misleading diagnosis due to the extensive soft tissue changes. CT can be helpful in detailing the bony destruction and pre-operative planning.

Therapy can include analgesia and repeated arthrocentesis followed by intra-articular steroid administration. In the advanced disease, shoulder arthroplasty may be considered.

The case presented perfectly exemplifies the MSSs’ characteristic epidemiology, clinical setting, location, evolution, as well as the distinctive combination of destructive and atrophic joint changes observed by imaging. Indeed, the diagnosis of this uncommon and fascinating destructive arthropathy benefits greatly of the integrated analysis of all these distinctive data.

Differential Diagnosis List: Milwaukee shoulder syndrome

Final Diagnosis: Milwaukee shoulder syndrome

References:

Description: Frontal view - revealed complete destruction of the humeral head, with cephalic migration of the proximal humerus, joint space narrowing and erosion of the glenoid. A large soft tissue swelling with extensive amorphous calcifications is observed. In addition, note the rib fractures also shown.

Origin:
Description: Lateral view Origin:
Description: Coronal volume-rendered image - extensive bony destruction of the scapular and humeral aspects of the left glenohumeral joint as well as extra-articular involvement of the acromion and coracoid process are shown. Notice also innumerous loose calcified bodies. Origin:
**Description:** Left oblique sagittal volume-rendered image - better depicts the extent of glenoid and acromion erosion. **Origin:**
Figure 3

Description: Frontal view - valgus deformity of the right knee with extensive bony destruction more apparent in the lateral tibial plateau, is shown. Origin:
**Description:** Lateral view - documents marked joint narrowing and extensive bony destruction of all the articulation surfaces. **Origin:**
Figure 4

Description: PD transverse image - demonstrates extensive bony destruction and a large joint effusion containing abundant loose calcified bodies. Origin:
Description: PD transverse image at a lower level - documents the extensive osseous and soft tissues changes. Origin:
Description: T2 fat-suppressed paracoronal image – documents the massive rotator cuff tear with tendinous retraction (arrow) and communication of joint space with the subacromial subdeltoid bursa (*). Synovial thickening and increased signal intensity of both surrounding soft tissues and bone marrow are visible. Origin: