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CASE REPORT

Arthritis Mutilans of the Shoulder: A Rare Cause for Rapidly Destructive Arthritis of the Shoulder

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ABSTRACT

Psoriatic arthritis (PsA) mutilans is a rare form of PsA that is extremely destructive, mimicking a charcot-like arthropathy. To date, the mutilans form of PsA has been described in the knee, spine, and elbow, but, to the best of our knowledge, this is the first report of this condition in the shoulder. This is a case presentation of a 59-year-old male with PsA demonstrating near complete and rapid disappearance of his right proximal humerus over the span of a few weeks. After a thorough diagnostic work-up, the patient was treated definitively with a hemiarthroplasty with improvement in his pain and range of motion (ROM) and satisfactory functional outcomes. Currently, there have not been any published recommendations for the diagnosis and management of arthritis mutilans of the shoulder, and we review our diagnostic and treatment strategy.

Keywords: Charcot-like arthropathy, Psoriatic arthritis mutilans, Rapidly destructive arthritis, Shoulder hemiarthroplasty.

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INTRODUCTION

Psoriatic arthritis mutilans is a rare form of PsA that is extremely destructive—mimicking a Charcot-like arthropathy. To date, the mutilans form of PsA has been described in the knee, hip, spine, and elbow, but, to the best of our knowledge, never in the shoulder. This is a case presentation of a 59-year-old male with PsA demonstrating near complete and rapid disappearance of his right proximal humerus over the span of a few weeks. After a thorough diagnostic work-up, the patient was treated definitively with a hemiarthroplasty with improvement in pain and ROM and satisfactory functional outcomes. Currently, there are no published recommendations for the diagnosis and management of arthritis mutilans of the shoulder. Herein, we review our diagnostic and treatment strategy.

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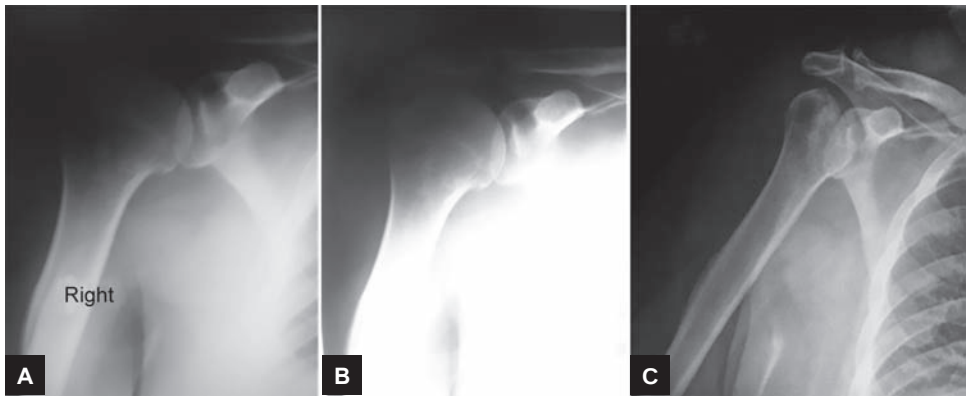
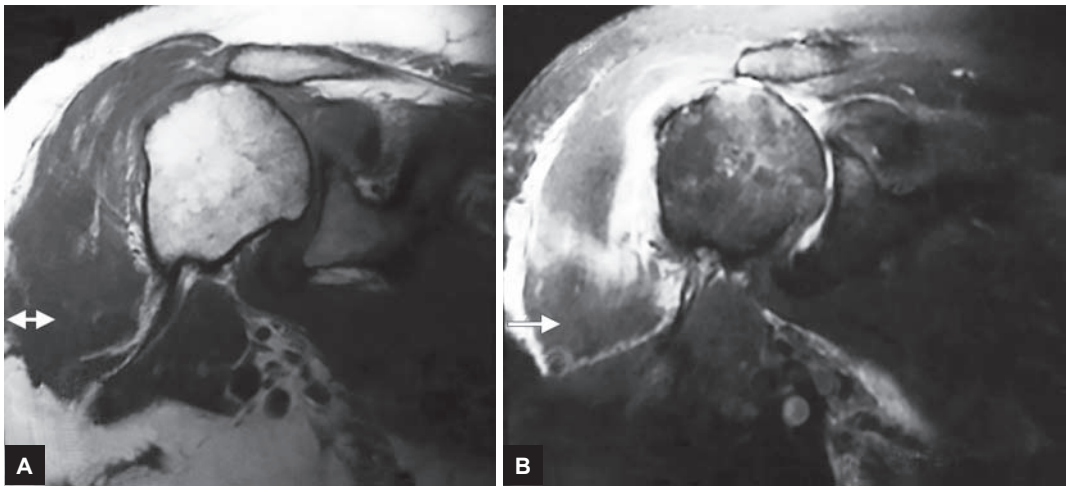
CASE REPORT

A 59-year-old man presented to our office for evaluation of right shoulder pain and decreased ROM that had developed suddenly with no apparent injury or trauma. His medical history was significant for PsA [rheumatoid factor (RF) negative] with intermittent joint pain in his elbows, shoulders, hips, knees, and feet. This condition required multiple treatments with oral steroids in the past for PsA flares. A Grashey anteroposterior (AP) view X-ray performed 3 months previously, when the right shoulder pain began (Figs 1A and B), demonstrated essentially normal bony anatomy and normal contour of the humeral head. The patient was managed with two cycles of cortisone injections to the right shoulder and acromioclavicular joints, which had given temporary pain relief and improved ROM, but the beneficial effects of the injections lasted for a maximum of 2 weeks.

The patient had a magnetic resonance imaging (MRI) of the right shoulder approximately 2 weeks prior to initial presentation at our office, demonstrating significant bone edema of the humeral head (Fig. 2) and a large retracted rotator cuff tear involving the supraspinatus and infraspinatus. There was also collapse of the humeral head and some evidence of superior humeral head migration. No evidence of avascular necrosis was detected. Repeat X-ray of the right shoulder performed during his evaluation, 9 weeks after his initial X-rays and 2 weeks after the MRI (Fig. 1C), revealed significant changes from the previous two studies. The humeral head was collapsed into a flat appearance with significant superior elevation of the humerus. There was no evidence of infection or recent risk factors for avascular necrosis that might explain the rapid decline in the appearance of his right shoulder. Given the rapid nature of the right shoulder changes, the differential diagnosis included Charcot arthropathy, septic arthritis, avascular necrosis, and Milwaukee shoulder. Given the patient's history of



Figs 1A to C: Grashey AP view of the right shoulder at initial presentation (A, B) and 9 weeks later (C) illustrating the rapid progression of the disease



Figs 2A and B: Coronal MRI images in T1 Fast Spin Echo sequence (A) and T2 Fast Spin Echo sequence (B) showing significant bone edema

PsA affecting other joints, atypical presentation of PsA of the shoulder was also entertained, although never described, and the patient's rheumatologist was consulted.

An MRI of the cervical spine was performed, which ruled out the presence of a syrinx. A rapid plasma reagin test for syphilis ruled out posterior column degeneration. The patient also had serum labs performed and a tagged white blood cell scan which demonstrated that an infection in the shoulder was unlikely. The patient then underwent a right shoulder arthroscopy to obtain biopsy and culture specimens. The arthroscopic appearance of the residual humeral head can be seen on the right side of the image in Figure 3. The pathology review of the biopsy specimen demonstrated vascularized edematous synovium with small foci of chronic inflammation, no granuloma or rheumatoid changes, no evidence of osteomyelitis, and hyperplastic fibrotic tenosynovium investing cancellous and cortical bone fragments (Fig. 4). The presence of fibrosis, sparse inflammatory cells, synovial hyperplasia, and increased vascularity in the biopsy specimen, rendered the diagnosis of mutilans form of PsA. There was no evidence of crystals when evaluating with polarized microscopy and special stains. Multicentric reticulohistiocytosis, a rare entity that can cause a similar presentation, was also ruled out. One culture specimen obtained during the arthroscopy demonstrated no growth and the other culture specimen demonstrated a rare bacillus species and *Staphylococcus epidermidis*

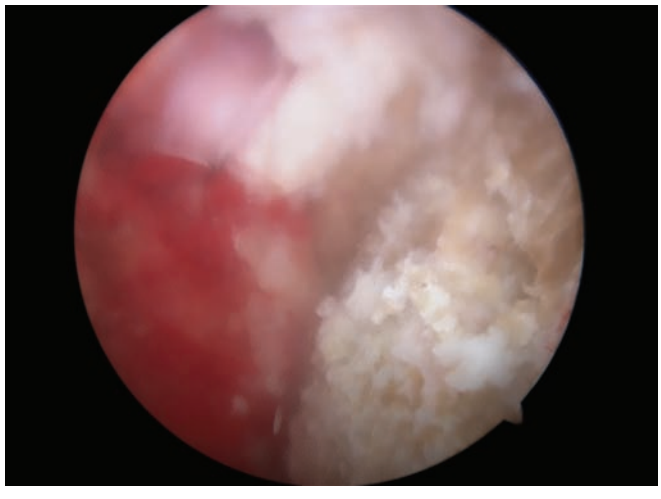
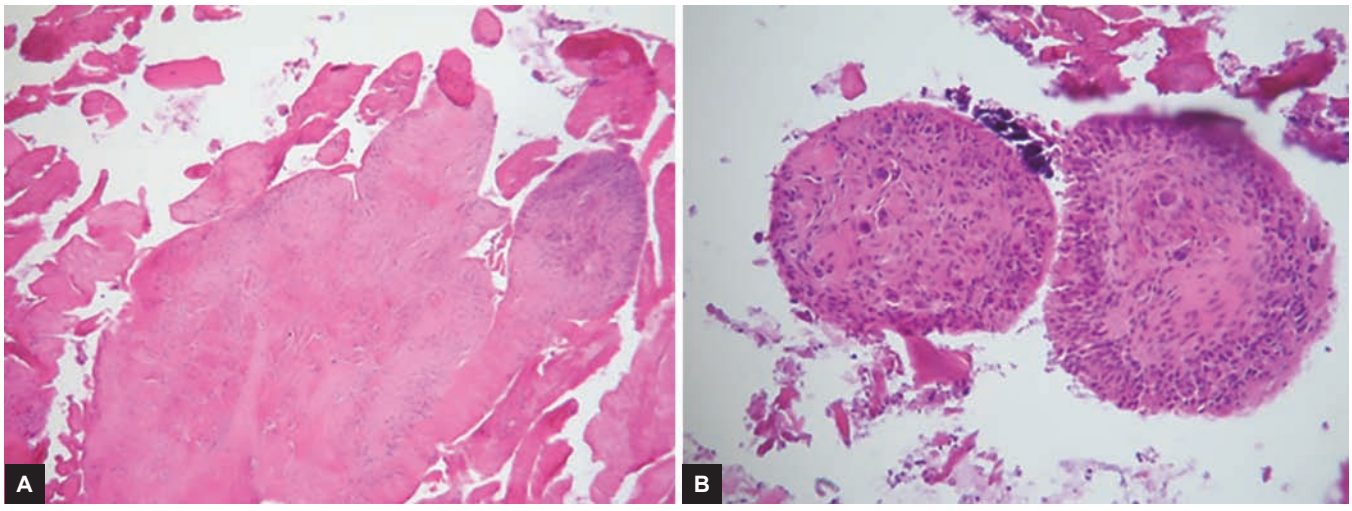


Fig. 3: Arthroscopic appearance of the residual humeral head



Figs 4A and B: Vascularized edematous synovium with small foci of chronic inflammation fibrosis, sparse inflammatory cells, synovial hyperplasia, and increased vascularity, rendering the diagnosis of mutilans form of PsA

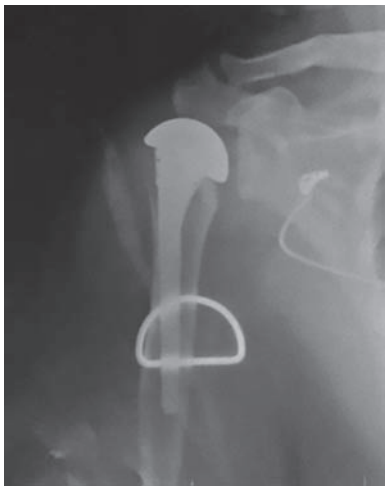


Fig. 5: Postoperative x-ray of the right shoulder

Fig. 6: ROM at 2-year follow-up

bacteria in broth only. The positive culture result was considered contaminant. Based on the pathology results and the findings of the diagnostic arthroscopy, the patient was diagnosed with a destructive form of psoriatic arthropathy affecting the shoulder. After considerable discussion, the recommendation given for definitive treatment of the shoulder was a hemiarthroplasty, with systemic medical management of PsA to start after the shoulder had fully healed.

A right shoulder hemiarthroplasty was performed by the senior author GEG utilizing a standard deltopectoral approach. A press-fit humeral stem (Affiniti, Tornier, Bloomington, Minnesota, USA) was chosen with an extended coverage head designed for articulation with the glenoid as well as the acromion in shoulders with massive rotator cuff tears (Fig. 5). The bone of the proximal humerus was very dense and demonstrated an abnormal dark discoloration. The hemiarthroplasty was implanted uneventfully. The remainder of the patient's operative course was without complication. The patient progressed very well postoperatively and demonstrated good, pain-free, functional ROM at his outpatient follow-up visits. At the final follow-up 2 years later, the patient required no pain medication, his American Shoulder and Elbow Surgeons Score was 62/100 at 2-year follow-up and his single assessment numeric evaluation score improved from 10% preoperatively to 50% at 2 years postoperative. His Penn shoulder score at 2 years was 55, with good pain subscale scores, but suboptimal overhead activity domain, with function essentially the same as his preoperative status. The short-form (SF)-36 physical scale score was 34.9/100; however, this low score was attributed to decreased ROM and pain at other body sites, likely due to arthritis. The SF-36 mental scale score was 65.3/100 at 2 years. Although ROM was decreased compared with the uninvolved shoulder, the patient had returned to work activities and was satisfied with the short-term result (Fig. 6).

DISCUSSION

Psoriatic arthritis is a seronegative spondyloarthropathy estimated to develop in 1 to 39% of patients with psoriasis.[1] Five subsets of PsA are recognized, according to the Moll and Wright classification[2]: Distal interphalangeal (DIP) joint involvement, peripheral asymmetric arthropathy, polyarticular symmetric involvement, psoriatic spondyloarthropathy, and arthritis mutilans.[3] The mutilans form is the rarest clinical form, accounting for less than 5% of all cases. Presenting with a rapid and impressive osteolysis and joint destruction, it is regarded as the most crippling and destructive of the subtypes leading to irreversible deformity.[3,4] Although acute onset of arthritis mutilans (3 months) was reported in 1 patient,[5] its development has been associated with longer arthritis duration.[6,7]

The diagnosis of PsA is suggested when a patient presents with psoriatic skin lesions, inflammatory type arthritis, and a negative serum RF. Along with Reiter's syndrome, ankylosing spondylitis and enteropathic arthropathies, it is one of the seronegative spondyloarthropathies associated with human leukocyte antigen (HLA)-B27.[8] Patients with psoriasis who carry the HLA-B7, HLA-B27, HLA-DR7, and HLA-Cw*0602 alleles are more likely to develop a destructive arthritic condition.[8] As 10 to 15% of the normal population is RF positive, it is possible to have PsA and be RF positive, making diagnosis challenging.

Chandran et al[9,10] defined arthritis mutilans radiographically, using the modified Steinbrocker scoring method, as ≥5 joints with grade 4 radiographic damage.[11] Although the initial description by Moll and Wright and the definition by Chandran et al[9,10] included ankylosis, osteolysis is generally accepted as a defining feature of this condition.[12] Several earlier attempts to define the disease focused mainly on the digital characteristics and changes in fingers and toes.[13,14] However, it has been since recognized to present in other skeletal locations, including the cervical spine, knee, and hip.[15-18] The typical radiographic features include severe osteolysis and bone resorption. As the most common locations of arthritis mutilans are the foot and hand, it is characterized by erosion of the DIP joints, periostitis, and joint ankylosis, resulting in whittling and cupping of both joint ends, leading to the characteristic "pencil-in-cup" deformity and even digital telescoping ("opera glass finger") in severe osteolysis.[3]

The MRI features in arthritis mutilans reveal higher erosion scores as well as bone edema and proliferation scores.[19] This suggests that bone edema visible on MRI could be used as a biomarker for early identification of arthritis mutilans.

It is currently unclear what causes some patients with PsA to develop the rare, mutilans form, which resembles a charcot-like arthropathy. A neurotraumatic theory has been suggested as a possible predisposing factor.[3] This theory suggests that somatic muscular reflexes in charge of joint protection from extreme ROM may be lost secondary to neuropathy, subsequently leading to joint destruction.[3] However, only a minority of patients with severe peripheral sensory neuropathy develop Charcot joints and this theory remains controversial.[3] Alternatively, the neurovascular theory proposes a neutrally initiated vascular reflex, leading to active bone resorption by osteoclasts.[3] Further research is needed to understand the etiology, presentation, and treatment of this disease.

The challenges in diagnosis necessitate awareness in PsA patients, especially when presenting with signs of joint inflammation. The involved joints are often less tender compared with patients with RA,[20] explaining why the severity of their disease is sometimes underestimated. The present case emphasizes not only the need for awareness when joints other than the feet and hands are involved, but also the need for rapid and sometimes aggressive intervention. Conservative treatment consists of nonsteroidal anti-inflammatory drugs, disease-modifying antirheumatic drugs, and antitumor necrosis factor-α (anti-TNF-α) agents. It has been suggested that early treatment might prevent joint damage.[21] Treatment with bisphosphonates, such as zoledronic acid, has been shown to reduce bone edema, but not erosive changes in PsA.[22] Anti-TNF agents appear to be even more effective, showing promise to reduce joint destruction.[22-24] Structural joint damage in patients with PsA was significantly prevented by infliximab therapy,[25] and a dramatic repair of joint damage has been observed in a patient with PsA following 2 years of etanercept therapy.[26]

When confronted with a patient with rapidly progressing shoulder joint destruction, the physician is advised to perform several immediate investigations to rule out pathological conditions such as infection, tumor, avascular necrosis, Charcot arthropathy, Milwaukee shoulder, and arthritis mutilans (Table 1).

While PsA has been described in the shoulder,[27] to the best of our knowledge, this is the first description of arthritis mutilans of the shoulder. After other conditions are ruled out and non-operative treatment has failed, operative treatment can provide acceptable results. The results of hemiarthroplasty for rotator cuff-deficient shoulders show improvement, but certainly not return to a normal shoulder.[28] This is to be expected, given that nothing is done to address the massive rotator cuff tear and the prosthesis is simply designed to provide

Table 1: Differential diagnosis of rapidly progressing shoulder joint destruction

Condition/pathology	*Investigation*
Infection	Aspirate sent for culture
	Serum ESR, CRP
	Consider arthroscopic or open biopsy for culture and tissue pathology
Tumor	Shoulder radiographs
	ESR
	Shoulder CT
AVN	Shoulder radiographs
	Shoulder MRI
Charcot joint	MRI, cervical spine to evaluate for syrinx
	Consider other neurodegenerative conditions
Milwaukee shoulder	Aspirate sent for crystal analysis
Arthritis mutilans	Thorough skin examination
	Serum RF
	Rheumatology referral
	Dermatology referrals if skin lesions present

ESR: Erythrocyte sedimentation rate; CRP: C-reactive protein; CT: Computed tomography; AVN: Avascular necrosis

a smoother bearing surface. Consequently, pain relief is helpful, but functional gains are modest with this procedure. Another treatment consideration that likely would have provided improved pain relief and function is reverse total shoulder arthroplasty, as this option addresses the functional deficits from the massive rotator cuff tear. The decision in this case was made for hemiarthroplasty given that the diagnosis was still unclear and with the thought that a more robust implant would be safer should further joint destruction ensue. Physicians treating this impressively destructive condition are advised to perform a thorough diagnostic work-up and consider this new entity, PsA mutilans of the shoulder, as part of the differential diagnosis.

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