

## CASE REPORT

# Arthritis Mutilans: Case Report of an Uncommon Squeal of Psoriatic Arthritis

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### ABSTRACT:

Psoriatic arthritis (PsA) is a seronegative spondyloarthropathy that presents clinically in different forms. Arthritis mutilans is a rare yet the most aggressive and disabling form of PsA that requires early diagnosis and aggressive therapy preferably in the form of biological agents to prevent or stop the irreversible destruction of small joints. We describe here, report of a case of long standing PsA who developed arthritis mutilans resulting in disability, compromised mobility and a poor quality of life. **Keywords:** Rheumatology, Hand deformity, Psoriasis, Arthritis, Joints

### INTRODUCTION:

Psoriatic arthritis (PsA) is a chronic inflammatory arthropathy associated with psoriasis, and included among the seronegative spondyloarthropathies<sup>1</sup>. Psoriasis features associated with a higher likelihood of PsA include nail dystrophy, scalp lesions, and intergluteal/perianal psoriasis<sup>2</sup>. Arthritis mutilans is a severe, deforming and destructive arthritis that affects fewer than 5 percent of people with psoriatic arthritis<sup>3</sup>. Although it is not necessarily a diagnostic challenge given its characteristic features, it is disabling and mutilating to a significant degree. Early diagnosis and adequate management can prevent the progression of disease and development of irreversible deformities. We present a case of long standing psoriatic arthritis who developed disabling arthritis mutilans resulting in restriction of mobility and a poor quality of life.

### CASE REPORT:

A 54-year-old man, with 18 years' history of psoriasis presented with progressive pain and deformities in both hands and feet. He had a poor response to methotrexate therapy and was only on non-steroidal anti-inflammatory drugs (NSAIDs) at the time of presentation. He had extensive scaly, erythematous lesions consistent with psoriasis on trunk and all limbs. On hand examination, there was synovial thickening and soft tissue swelling of the wrists and metacarpophalangeal joints (MCPs), dactylitis, and boutonnière and swan neck deformities in both hands (Figure-1). Characteristic nail changes in the form of nail pits, onycholysis, oil drop sign and nail-bed hyperkeratosis were present (Figure-2). Similar changes were also present in both feet (Figure-3).

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X-rays of the hand and feet revealed bilateral, asymmetrical, fusiform soft-tissue swelling, erosive changes and new bone formation in the distal joints, "pencil-in-the-cup" appearance, sub-luxation of the right second metacarpophalangeal joint and joint-space narrowing in the interphalangeal joints (Figure-4). Results of the routine haematologic and biochemical profiles were unremarkable. His rheumatoid factor was negative.

He was advised NSAIDs, retinoid and narrow-band ultraviolet-B (NB-UVB) phototherapy. The skin lesions responded well but the relief in joint pain was not adequate. The patient is on follow-up visits. Biological agents and surgery were not considered because of financial constraints of the patient and the systemic steroids were avoided because of possible rebound of the skin disease upon withdrawal.

### DISCUSSION:

Five main clinical patterns of psoriatic arthritis have been identified; distal interphalangeal (DIP) predominant, symmetrical polyarthritis ( $\geq 5$  involved joints), asymmetrical oligoarthritis ( $\leq 4$  involved joints) and monoarthritis, predominant spondylitis, and arthritis mutilans<sup>4</sup>. The diagnosis of psoriatic arthritis is easy in the presence of typical skin lesions, however it can also be made in absence of skin lesions using Classification of Psoriatic Arthritis criteria. Arthritis mutilans (AM) has been described in association with a wide variety of arthropathies, including rheumatoid arthritis, psoriatic arthritis, juvenile idiopathic arthritis, systemic sclerosis and Systemic lupus erythematosus<sup>5</sup>. Bruzees et al published a case series of arthritis mutilans and presented a literature review of the last decade<sup>6</sup>. They documented 120 cases of psoriatic arthritis mutilans (age range 34-71 years), all from the Americas, Europe and New Zealand. This does not suggest that such complications of psoriatic arthritis do not occur in developing world (particularly Africa, Middle East and Asia). It only indicates a low rate of reporting and documentation from these areas. The duration of psoriasis ranged from 10-52 years and of the arthritis mutilans from 0.3 to 30 years. Most of them were managed with DMARDs

(disease-modifying antirheumatic drugs) while only few (7 cases) were given etanercept.

Arthritis mutilans is an aggressive form of arthritis featured by extensive erosion and destruction of small joints of hands and feet. The resulting finger and toe disfigurements impair the patients' functional capacity<sup>6</sup>.

The development of PsA is significantly associated with the duration of arthritis and involvement of DIP joints.

Local corticosteroid injections and non-steroidal anti-inflammatory drugs are recommended in milder forms of PsA. Leflunomide and methotrexate are usually the first choice in peripheral psoriatic arthritis<sup>7</sup>. Biological therapies with tumor necrosis factor- $\alpha$  (TNF- $\alpha$ ) blockers should be employed if active psoriatic arthritis fails to adequately respond to one or more of DMARDs, or if the patient has active, predominantly axial disease. Arthritis mutilans does not respond to the traditional DMARDs, however, TNF- $\alpha$  blockers, have shown

improvement in resolution of bone and joint destruction and improvement in functional capacity in patients with Arthritis mutilans. It has been suggested that these might be more effective than methotrexate in management of PsA<sup>8</sup>. Nevertheless, decision to use biological therapies depends on patient's comorbidities, preferences and affordability<sup>9,10</sup>.

#### CONCLUSION:

Arthritis mutilans is the most aggressive and disabling form of PsA. In patients where methotrexate fails, Tumor necrosis factor  $\alpha$  blockers can be the treatment of choice. However, keeping in view the high cost of biological agents, this might not be a feasible option for most of the patients with PsA in developing countries like Pakistan. Therefore in order to prevent permanent joint damage, early recognition and treatment of PsA are critical.



**Figure 1:** Hands showing psoriatic lesions, synovial thickening of MCPs, dactylitis, boutonniere and swan neck deformities in both hands



**Figure 2:** Nail pits, onycholysis and nailbed hyperkeratosis in finger nails.



**Figure 3:** Psoriatic skin lesions, nail changes and arthritis mutilans in feet.



**Figure 4:** Radiological changes in both hands suggestive of arthritis mutilans

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