

Arthritis Mutilans in a Patient with Psoriasis

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Arthritis is reported to be a feature of psoriasis in approximately 7% of cases.¹ The most dramatic and severe form of arthritis associated with psoriasis is arthritis mutilans (AM), which is a rare disorder, affecting fewer than 5% of patients with psoriasis.² AM has a predilection for the small joints of the hands and feet. It is generally characterized by seronegative degenerative joint disease, leading to osteolytic changes in the carpal and digital bones. The bone and joint lesions rapidly and progressively cause bone lysis and joint ankylosis with loss of digits, soft-tissue deformities, telescoping of fingers and toes, and the hallmark “la main en lorgnette” deformity (*opera-glass hand*).³

This article reports the case of a 55-year-old man with a more than 20-year history of psoriasis and AM. The epidemiology, clinical features, pathology, and management of AM are discussed.

CASE PRESENTATION

Initial Presentation and History

A 55-year-old man came to the emergency department of a Veterans Affairs hospital in 2001 for treatment of arthritis affecting the joints of his hands and feet. He was not taking any medication at the time of presentation.

Medical history was significant for psoriasis and arthritis of more than 20 years' duration, with skin lesions first appearing in 1973. In 1981, he had been treated with methotrexate at a local clinic. Despite improvement of symptoms with the drug, he had stopped taking it because he could not afford the cost of treatment. He had not been specifically followed for his medical condition since that time. Approximately 6 years ago, he had to stop working because of loss of strength and flexibility of the fingers and thumbs in both hands. There was no other significant historical finding.

Physical Examination

The patient appeared well, with a normal body habitus and no signs of being obese or underweight. He walked with some difficulty (but without assistance) because of painful feet. Pain in the joints of his hands and feet was assessed as grade 5/10. Temperature was 36.1°C (96.9°F), and blood pressure was 144/86 mm Hg.

Breath sounds were normal, and cardiovascular examination was unremarkable. Bowel sounds were normal. His abdomen was soft and nontender. There was an extensive area on the trunk, extending from the lower chest to below the umbilicus and across the lower third of the back, of a scaly, silver-colored, confluent skin lesion consistent with psoriasis (Figure 1). Both hands showed swelling and shortening of the digits. On the left, there was significant swelling and shortening of the thumb and the second, fourth, and fifth digits. There also was pitting of the nails of the right third and fourth digits. Onycholysis of the other nails on the right was also noted (Figures 2 and 3). Both feet exhibited marked shortening of the toes and soft-tissue swelling, bilaterally (Figures 4 and 5). These findings were consistent with AM associated with the patient's psoriasis.

Laboratory Examination

Results of routine hematologic and biochemical profiles were unremarkable. Rheumatoid factor was within normal limits. Urinalysis showed no abnormalities.

Radiographic Examination

Detailed radiographic imaging of both hands and feet was performed. In the hands, extensive resorption and deformities were noted in the distal and middle phalanges, more so on the left, involving all the digits. The right index and ring fingers were relatively spared. Marked foreshortening of all the digits was seen. (Figure 6)

Radiographs of the feet showed acro-osteolysis of tarsal bones on both sides. The first metatarsophalangeal joints in both feet were narrowed and showed articular erosions. There was radiographic evidence of diffuse soft-tissue swelling on both sides as well. (Figures 7 and 8).

Treatment and Outcome

The patient was referred to the rheumatology service at the hospital, and treatment was initiated with

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Figure 1. Clinical photographs showing a scaly, silver-colored, confluent skin lesion consistent with psoriasis covering an extensive area of the trunk of the case patient. The lesion extends from the lower chest to below the umbilicus (A) and across the lower third of the back (B).

administration of nonsteroidal anti-inflammatory drugs and methotrexate, which had proved helpful in the past. Treatment of the skin lesions with a topical corticosteroid preparation and calcipotriene resulted in considerable improvement. Because of a suboptimal response to a 12-week trial of methotrexate, the patient's drug regimen was switched to etanercept, which resulted in some improvement in symptoms. It is still too early to tell, however, if remission will be achieved in this patient.

DISCUSSION

Epidemiology and Clinical Features

Psoriatic arthritis (PsA) is a seronegative, inflammatory arthropathy occurring in 5% to 42% of patients with psoriasis.³ PsA is differentiated from rheumatoid



Figure 2. Clinical photograph of the right hand of the case patient showing swelling and shortening of the digits.



Figure 3. Clinical photograph of the left hand of the case patient showing marked swelling and shortening of the thumb and the second, fourth, and fifth digits, as well as pitting of the nails of the third and fourth digits. Onycholysis of the other nails is also present.

arthritis (RA) by more common involvement of the distal interphalangeal (DIP) joints and axial skeleton and by the absence of systemic features (eg, vasculitis, Sjögren's syndrome, uveitis, rheumatoid nodules). Periarticular tissues are also affected in cases of PsA, producing enthesopathies, tenosynovitis, and the hallmark sign of dactylitis (ie, diffuse swelling of an entire digit). Joint disease occurs after the appearance of the skin lesion in 15% of affected patients and precedes the skin condition in 10% to 15% of patients.⁴

The arthropathy associated with psoriasis has been divided into 5 morphologic types: (1) asymmetric



Figure 4. Clinical photograph of the left foot of the case patient showing marked shortening of the toes and soft-tissue swelling, bilaterally.



Figure 5. Clinical photograph of the right foot of the case patient showing marked shortening of the toes and soft-tissue swelling, bilaterally.

oligoarthritis, (2) symmetric polyarthritis, (3) arthritis involving the DIP joints, (4) spondylitis, and (5) AM. Clinically, there is considerable overlap between these various types, with a general progression toward polyarticular disease seen in the majority of patients.⁵

Asymmetric oligoarthritis is the commonest form, affecting nearly 70% of patients with PsA.⁴ Enthesitis resulting in “sausage-shaped” digits is a typical finding. Symmetric polyarthritis is present in approximately 33%⁶ of patients with PsA and is characterized by erosive disease in the majority of patients. Arthritis involving the DIP joints occurs in approximately 16% of patients with PsA⁴ and is rare without other joint in-



Figure 6. Radiograph of the case patient's hands showing extensive resorption and deformities in the distal and middle phalanges, especially on the left, involving all the digits. The right index and ring fingers are relatively spared. Marked foreshortening of all the digits is apparent.



Figure 7. Radiograph of the case patient's feet showing acro-osteolysis of the tarsal bones on both sides. The first metatarsophalangeal joint in both feet is narrowed and shows articular erosions.

volvement. Spondylitis as a predominant form affects approximately 4% of patients with PsA and is clinically similar to ankylosing spondylitis.^{3,6} This variant is also classified as a spondyloarthropathy and, thus, predominantly involves the axial skeleton. Spondylitis also is primarily associated with asymmetric sacroiliac joint involvement, although peripheral joints may sometimes be affected.

AM is the most destructive form of PsA. Fortunately, it is rare, affecting only approximately 5%⁷ of patients with PsA. AM is characterized by an asymmetric pattern

of peripheral joint involvement, with a predilection for the interphalangeal and metacarpophalangeal joints of the hand and small joints of the feet.⁷ Characteristic features of AM are severe multijoint deformity of the hands, foreshortened fingers with excessive skin folds, hypermobile joints, and digits that can be elongated by traction.⁷ Radiologically, AM is characterized by severe resorption of the joint with an attendant loss of function, sometimes to a dramatic degree.

Given the variety of morphologic features associated with AM, it is not surprising that attempts have been made to quantify them objectively. For example, the Larson hand score of 0 to 110 has been proposed to evaluate the destruction of the joints of the hand; more recently, another hand score (0–10) has also been described.⁸

Etiology and Pathogenesis

The etiology and pathogenesis of PsA are not clearly understood. Evidence suggests an interplay between genetic, immunologic, and environmental factors.³ For example, studies indicate a concordance rate of 70% in monozygotic twins, compared with 20% in dizygotic twins.⁴ In addition, certain HLA markers appear to be related to disease progression and prognosis. HLA-B27, HLA-DR7, and HLA-DQw3 are predictive of disease progression, whereas HLA-B22 appears to be protective.³ Immune causality may also play a role, as is suggested by synovial membrane infiltration with immune-competent cells, such as macrophages and lymphocytes, in PsA. Furthermore, high levels of interleukin-1 and tumor necrosis factor (TNF)- α also suggest a cytokine-driven process. Environmental agents (eg, infectious agents, traumatic insults) also have been implicated in the etiology of arthritis in susceptible individuals.

Differential Diagnosis

AM is caused by a wide variety of arthropathies, with RA and PsA being the most commonly associated diseases.⁹ It also occurs in association with juvenile RA, neuropathic arthropathy, gout, porphyria, mixed connective tissue disease, multicentric reticulohistiocytosis, scleroderma, Raynaud's phenomenon, sarcoidosis, tuberculosis, vinyl chloride poisoning, tertiary syphilis, osteomyelitis, and erosive osteoarthritis.⁵

In the case patient, the most likely cause of AM was PsA, based on the extensive skin involvement and exclusion of other etiologies and associations.

Radiographic Features

Radiographically AM is characterized by the pres-



Figure 8. Radiograph of the case patient's left foot showing diffuse soft-tissue swelling.

ence of severe bone and joint resorption⁷ with a predilection for the DIP and proximal interphalangeal joints. In contrast to RA, new bone formation is seen in AM, with evidence of traction spurs, periostitis, and ankylosis of joints. A characteristic radiologic feature is the “pencil-in-cup” deformity, which is associated with the destruction of the head of a metacarpal bone with osteolysis and new bone formation. This process produces the characteristic cupped appearance in the bone of the adjacent phalanx.⁹

Pharmacologic Treatment

A large number of patients with PsA have fairly mild and nondestructive disease and can be treated with nonsteroidal anti-inflammatory drugs. Disease-modifying agents, such as gold and antimalarial drugs, although effective in cases of RA, may not be suited for the management of PsA, because they are known to aggravate the skin lesions of psoriasis.¹⁰ Methotrexate is effective in controlling cutaneous psoriasis and is therefore commonly used to treat PsA.^{11–13} Whereas cyclosporin effectively treats both skin and joint manifestations of PsA, renal toxicity limits its use.³ Studies of patients using sulfasalazine versus placebo have shown trends toward improvement.¹⁴ The retinoid etretinate appears to be effective in clearing the skin disease in open trials, with improvement reported in 55% to 100% of patients.¹⁵ Further randomized trials are clearly needed to assess these therapeutic models. Many other candidate compounds and techniques appear to have therapeutic potential in this disorder. Some that have been evaluated in recent years include colchicine, azathioprine, tacrolimus, and psoralen plus ultraviolet A.¹⁵

As advances are made in newer therapies for the treatment of RA, some are inevitably starting to influence the management of PsA and other immune arthropathies.³ Preliminary results of studies using

anti-TNF agents (eg, etanercept, infliximab) to treat PsA have shown promise and had positive results.^{16–18} Anti-TNF drugs certainly deserve further consideration in cases of PsA, particularly when the disorder is complicated by AM. However, it is unclear if this strategy will have an impact on the osteolysis and ankylosis that cause much of the morbidity in PsA. Specifically directed studies will be needed to prove the safety and efficacy of these drugs.

Surgical Treatment

For AM, surgical intervention is very challenging. However, attempts at correcting various deforming and disabling features of AM have shown some benefit. Generally, these surgical procedures involve distraction lengthening and bone grafting for telescoping of digits, various types of arthroplasties, arthrodesis, and joint manipulation. Because spontaneous joint fusion appears to have a protective effect in preventing further digital shortening, it has been suggested that early surgical intervention with prophylactic arthrodesis may be beneficial.⁵ The combination of arthrodesis and arthroplasty also has been shown to improve function in some patients.

CONCLUSION

AM is a rare but compelling type of advanced joint disease in patients with psoriasis. Although it does not necessarily present a diagnostic challenge, given its distinctive morphology, it is mutilating and disabling to a significant degree. Furthermore, there appears to be no established method for treating AM effectively, although intermittent success has been seen with a variety of pharmaceutical and surgical options. There is a clear need to investigate the pathogenesis of AM further and to design and develop more effective therapies to circumvent its potential to destroy the locomotor appendages and ultimately disable patients permanently. It is not unreasonable to expect that, with additional empirical data, the various debilitating aspects of AM will be successfully remedied in patients with psoriasis.

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REFERENCES

1. Moll JM, Wright V. Psoriatic arthritis. *Semin Arthritis Rheum* 1973;3:55–78.
2. Roberts ME, Wright V, Hill AG, Mehra AC. Psoriatic arthritis. *Ann Rheum Dis* 1976;35:206–12.
3. Gladman DD, Brockbank J. Psoriatic arthritis. *Expert Opin Investig Drugs* 2000;9:1511–22.
4. Veys EM, Mielants H. Current concepts in psoriatic arthritis. *Dermatology* 1994;189 Suppl 2:35–41.
5. Walton RL, Brown RE, Giansiracusa DF. Psoriatic arthritis mutilans: digital distraction lengthening: pathophysiologic and current therapeutic review. *J Hand Surg (Am)* 1988;13:510–5.
6. Veale D, Rogers S, Fitzgerald O. Classification of clinical subsets in psoriatic arthritis. *Br J Rheumatol* 1994;33:133–8.
7. Belt EA, Kaarela K, Kauppi MJ, et al. Assessment of mutilans-like hand deformities in chronic inflammatory joint disease. A radiographic study of 52 patients. *Ann Rheum Dis* 1999;58:250–2.
8. Larsen A, Dale K, Eek M. Radiographic evaluation of rheumatoid arthritis and related conditions by standard reference films. *Acta Radiol Diagn (Stockh)* 1977;18:481–91.
9. Bruce I, Gladman DD. Psoriatic arthritis: recognition and management. *BioDrugs* 1998;9:271–8.
10. Trnavsky K, Zbojanova M, Vlcek F. Antimalarials in psoriatic arthritis. *J Rheumatol* 1983;10:833–4.
11. Jones G, Crotty M, Brooks P. Psoriatic arthritis: a quantitative overview of therapeutic options. The Psoriatic Arthritis Meta-Analysis Study Group. *Br J Rheumatol* 1997;36:95–9.
12. Black RL, O'Brien WM, Van Scott EJ, et al. Methotrexate therapy in psoriatic arthritis; double-blind study on 21 patients. *JAMA* 1964;189:743–7.
13. Willkens RF, Williams HJ, Ward JR, et al. Randomized, double-blind, placebo controlled trial of low-dose pulse methotrexate in psoriatic arthritis. *Arthritis Rheum* 1984;27:376–81.
14. Farr M, Kitas GD, Waterhouse L, et al. Treatment of psoriatic arthritis with sulphasalazine: a one-year open study. *Clin Rheumatol* 1988;7:372–7.
15. Piro MH, Cash JM. Treatment of refractory psoriatic arthritis. *Rheum Dis Clin North Am* 1995;21:129–49.
16. Hohler T, Kruger A, Schneider PM, et al. A TNF-alpha promoter polymorphism is associated with juvenile onset psoriasis and psoriatic arthritis. *J Invest Dermatol* 1997;109:562–5.
17. Mease P, Goffe B, Metz J, et al. Etanercept in the treatment of psoriatic arthritis and psoriasis: a randomized trial. *Lancet* 2000;356:385–90.
18. Yazici Y, Erkan D, Lockshin MD. Etanercept in the treatment of severe, resistant psoriatic arthritis: continued efficacy and changing patterns of use after two years [letter]. *Clin Exp Rheumatol* 2002;20:115.

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