Multicentric Reticulohistiocytosis: Diagnosis at the Nailbeds

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We evaluated a 67-year-old woman for bilateral hand swelling and pain for 3 months. During this time she developed small erythematous papules on her wrists, fingers, and ears. On examination she had superficial nodulosis on the pinnae of her ears, elbows, and fingers, and along her cuticles, which had a "beaded string" appearance (Figure 1). She had synovitis involving her metacarpophalangeal joints and wrists bilaterally. Her laboratory values were remarkable for elevated rheumatoid factor and a normocytic anemia. Anticyclic citrullinated peptide antibodies were also present. Hand radiographs showed no erosive changes. She underwent biopsy of a finger nodule that revealed proliferation of histiocytes in the dermis that were large, angled, and multinucleated (Figure 2).

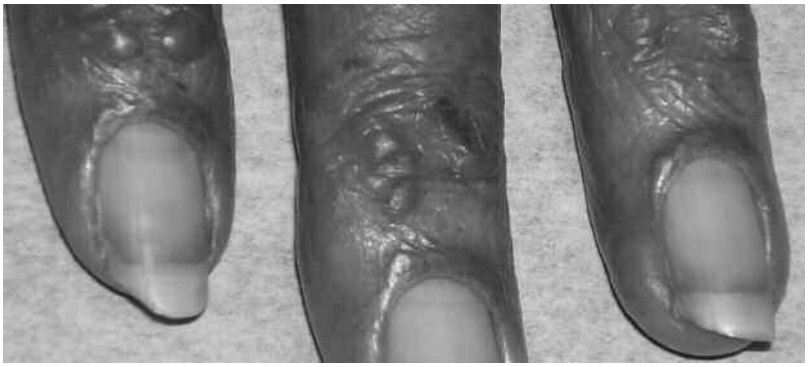

Figure 1. Nodules on the distal phalanges along with coalescing nodules at the periungual area with "beaded string" appearance.

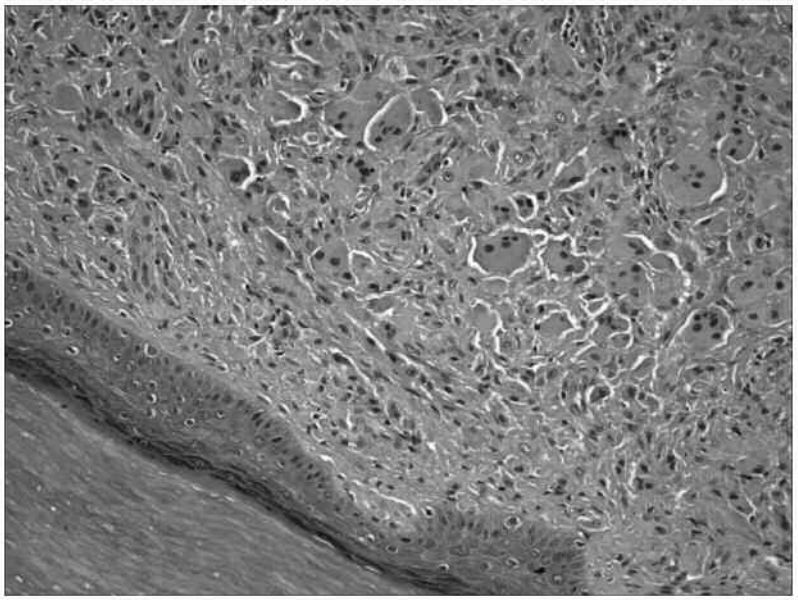

Figure 2. Biopsy revealed a diffuse multinucleated histiocytic giant-cell infiltration throughout the dermis. Overlying epidermis is mildly to moderately hyperkeratotic with atrophy and flattening of rete ridges. Some areas show bands of collagen below the dermal-epidermal junction that separate the infiltrate from the epidermis (H&E ×40).

Multicentric reticulohistiocytosis is a rare systemic disorder with multiple periungual smooth and shiny erythematous papules that can be associated with a polyarticular, erosive arthritis. Women are affected more commonly than men and usual onset is in the fourth decade. The disease is associated with an underlying malignancy in 15%–28% and a coexisting autoimmune disorder in 6%–17%. The disease typically remits within 5–10 years. Diagnosis is based on characteristic findings on skin or synovial biopsy[1]. Treatment is difficult because the disease does not typically respond well to disease modifying antirheumatic drugs such as methotrexate and hydroxychloroquine. However, recent case reports suggest improvement with anti-tumor necrosis factor-α therapy, etanercept, or aminobisphosphonate therapy[2,3].

Our patient had a negative screening for age-appropriate cancers. Additionally, she had normal computed tomography of the chest, abdomen, and pelvis. She has responded well to methotrexate and aminobisphosphonate therapy.

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