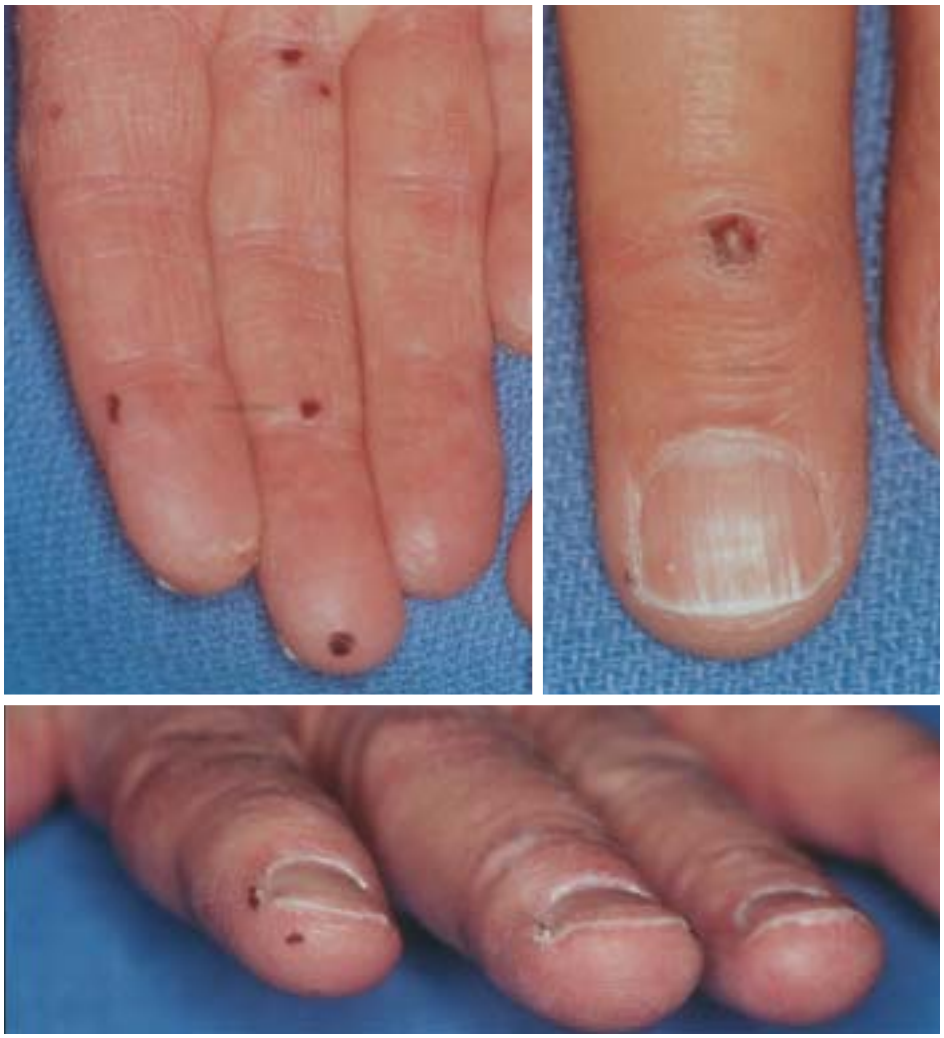
What Is Your Diagnosis?

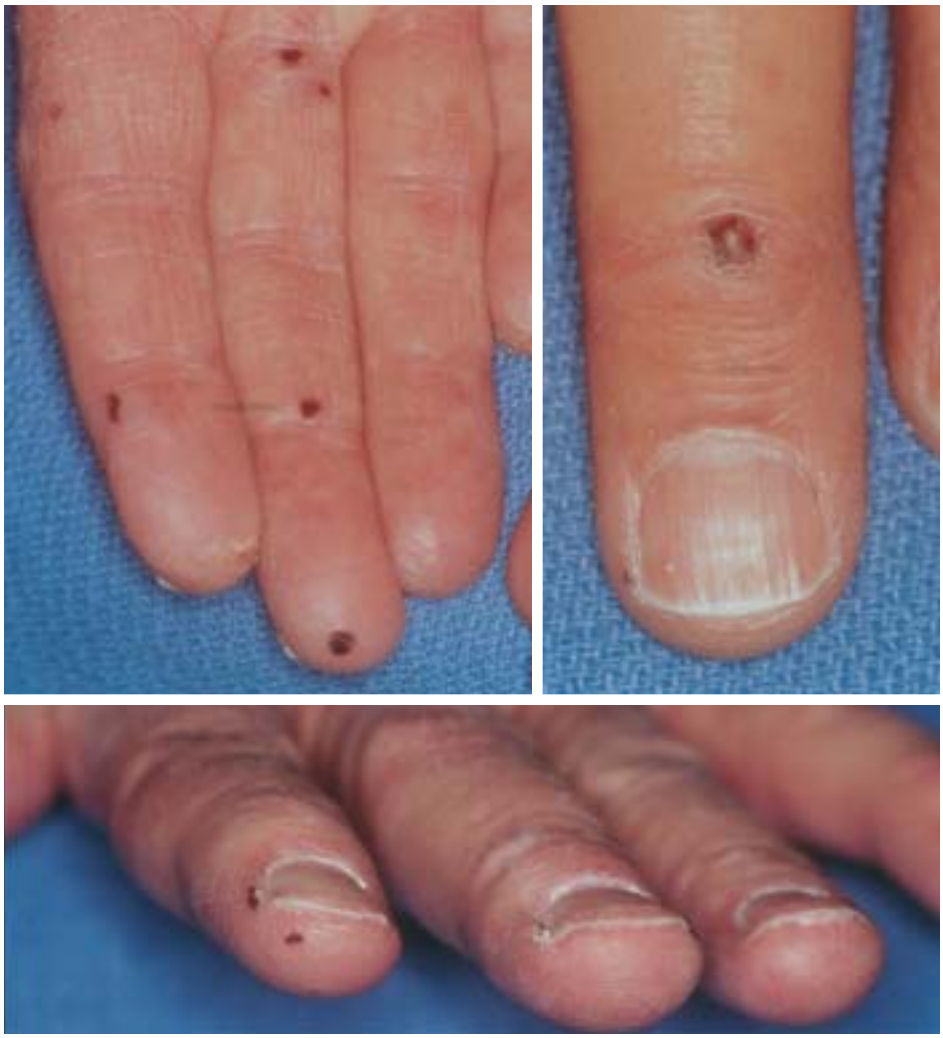


This patient has active rheumatoid arthritis. His rheumatoid factor is elevated, and he presented with multiple acral reddish-brown papules on the digital pulp and nail folds of both hands.

PLEASE TURN TO PAGE 462 FOR DISCUSSION

Mark D. Herron, MD; Scott Florell, MD; Department of Dermatology, University of Utah School of Medicine, Salt Lake City.
The authors report no conflict of interest.

The Diagnosis: Bywaters Lesions of Rheumatoid Vasculitis



In 1949, Bywaters[1] reported 6 patients with clinical and radiological evidence of rheumatoid arthritis presenting with small nodules on the terminal pads of the fingers, the free edge of the nails, and the dorsum of the fingers. In addition, the patients displayed nail-fold capillary thrombosis and necrosis. These digital nodules healed leaving a brown eschar. Two of these cases progressed early to gangrene, and the patients died from complications of the vasculitis. Four cases did not evolve into gangrene.[1]

In 1957, Bywaters[2] described 10 cases of rheumatoid arthritis with cutaneous vessel obliteration manifesting as a spectrum from brown lesions of the nail fold, the nail edge, and the digital pulp progressing to gangrene of the extremities, limbs, and viscera. Biopsy results showed intimal obliteration within vessels as a primary feature. In those cases with transient cutaneous involvement, the clinical signs were similar to those seen in the 2 cases in which the patients died—nail-fold, nail-edge, and finger-pulp lesions resulting in brown keratinizing spots with no tissue loss.[2]

In a final study in 1963, Bywaters and Scott[3] observed a close association between episodes of small vascular lesions and a positive rheumatoid factor. They also found a uniform presence of

CONTINUED ON PAGE 464

CONTINUED FROM PAGE 462

rheumatoid nodules on the elbows and a positive rheumatoid factor. Of the 35 patients in their comparative study, 20 patients with vascular lesions on the nail folds, nail edges, and digital pulp managed to maintain functional lives, with no progression of the vasculitis. Only 4 of these patients died of systemic vasculitis.[3]

Bywaters lesions signify cutaneous vasculitis occurring with rheumatoid arthritis. Clinically, this form of vasculitis manifests as nail-fold thromboses and nail-fold telangiectasia, with accompanying small brown infarcted papules of the nail fold, the nail edge, or the digital pulp. Infiltrates of neutrophils, nuclear debris, fibrin deposition within vessels, and thrombi within the vascular lumen are nonspecific findings of Bywaters lesions.[4] These lesions show leukocytoclastic vasculitis and differ from rheumatoid nodules in that no palisading granulomas can be found. This milder form of rheumatoid vasculitis occurs without systemic evidence of vasculitis.

Moderate rheumatoid vasculitis initially presents with palpable purpura on dependent sites, such as the lower extremities. Systemic symptoms may involve malaise, myalgia, arthralgia, pleurisy, and gastrointestinal pain. Fever, proteinuria, hematuria, pulmonary infiltrates, pericarditis, retinal hemorrhages, and neuropathy may accompany the cutaneous vasculitis.[5]

Severe rheumatoid vasculitis exhibits peripheral neuropathy; gastrointestinal bleeding; and involvement of the lungs, kidneys, and heart. Vasculitis of the small and medium vessels causes digital gangrene, nail-fold infarcts, and cutaneous ulceration. Of these, digital gangrene is most notable with severe rheumatoid arthritis.[5] In a comparative study, 94% of patients with rheumatoid vasculitis tested positive for rheumatoid factor, and 88% tested positive for the presence of HLA-DR4.[6] The presence of the HLA-DR4 haplotype has been associated with severe disease, and its absence has been associated with milder disease.[7]

Patients with active rheumatoid arthritis have a high incidence of cutaneous vasculitic lesions. In addition, patients with rheumatoid nodules and high titers of rheumatoid factor appear to be predisposed to rheumatoid vasculitis.[3] Bywaters lesions are regarded as a manifestation of mild rheumatoid vasculitis and frequently occur without systemic signs of vasculitis. Nail-fold and finger-pulp papules are a distinct feature of this type of rheumatoid vasculitis. These papules can progress to larger painful hemorrhagic papular lesions of the digital pads and represent infarction at the region of the nail fold.

REFERENCES

1. Bywaters EGA. Variant of rheumatoid arthritis characterized by recurrent digital pad nodules and palmar fascitis, closely resembling palindromic rheumatism. *Ann Rheum Dis*. 1949;8:1-21.
2. Bywaters EGA. Peripheral vascular obstruction in rheumatoid arthritis and its relationship to other vascular lesions. *Ann Rheum Dis*. 1957;16:84-103.
3. Bywaters EG, Scott JT. The natural history of vascular lesions in rheumatoid arthritis. *J Chron Dis*. 1963;16:905-914.
4. Craig SD, Jorizzo JL, White WL, et al. Cutaneous signs of rheumatic disease: acral purpuric papules in a patient with clinical rheumatoid arthritis. *Arthritis Rheum*. 1994;37:957-959.
5. Jorizzo JL, Daniels JC. Dermatologic conditions reported with rheumatoid arthritis. *J Am Acad Dermatol*. 1983;8:439-457.
6. Scott DG, Bacon PA, Tribe CR. Systemic rheumatoid vasculitis: a clinical and laboratory study of 50 cases. *Medicine (Baltimore)*. 1981;50:288-297.
7. Weyand CM, Hicok KC, Conn DL, et al. The influence of HLA-DRB1 genes on disease severity in rheumatoid arthritis. *Ann Intern Med*. 1992;117:801-806.