cutis® Photo Quiz

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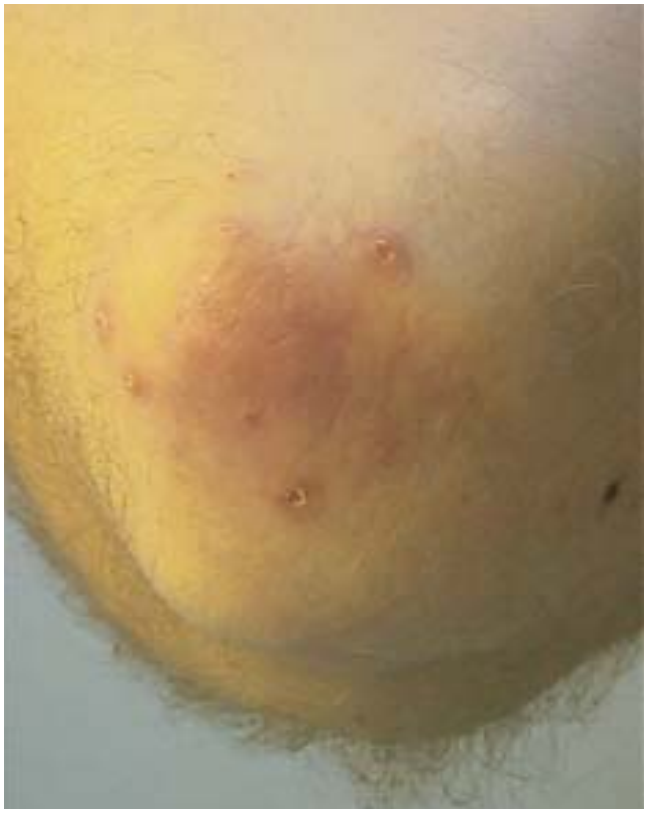
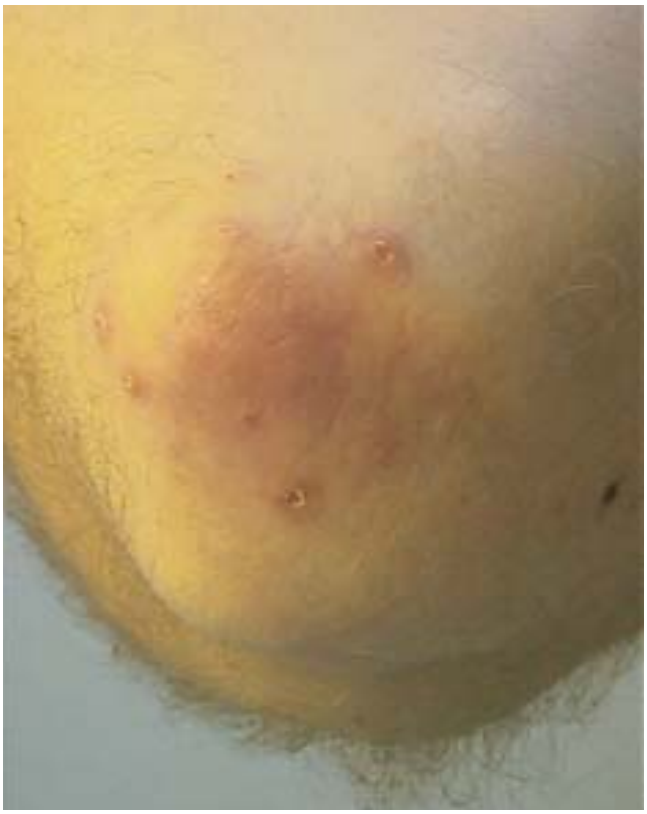


A 33-year-old pharmacy technician and farmer was being treated for Wegener's granulomatosis and later presented for evaluation of tender areas on the left elbow.

What is your diagnosis?

PLEASE TURN TO PAGE 37 FOR DISCUSSION

The Diagnosis: Palisaded Neutrophilic and Granulomatous Dermatitis With Underlying Wegener's Granulomatosis



Palisaded neutrophilic and granulomatous dermatitis describes a disorder seen in the setting of autoimmune disease.[1] Synonyms include Churg-Strauss granuloma, cutaneous extravascular necrotizing granuloma, and Winkelmann's granuloma.[2] Tender umbilicated papules on the extensor surface of the joints, especially on the elbows, are characteristic.[1] This unique presentation is associated with vasculitis, lupus erythematosus, and other autoimmune disorders. Although areas of necrobiosis are not identified, fibrinoid degeneration of collagen is evident and is surrounded by a mixed inflammatory infiltrate. This disorder was first reported by Finan and Winkelmann in 1983[2] and given its current name by Chu and colleagues in 1994.[1] The primary pathologic changes are in the dermis, unlike rheumatid nodules that primarily involve subcutaneous tissue. Small vessels may exhibit staining with IgM and C3 when studied by direct immunofluorescence. Different histologic patterns may be noted, depending on the age of the lesion. A mixed inflammatory infiltrate with numerous neutrophils may be seen in the early stages of lesion development, whereas a chronic infiltrate with giant cells

may be seen at a later point. Phosphotungstic acid–hematoxylin stain highlights perivascular fibrin, and extensive infiltration of neutrophils is found throughout the dermis in early lesions. Leukocytoclastic vasculitis is evident. As lesions develop, the number of neutrophils diminishes, and leukocytoclastic vasculitis may be absent. Fibrin cuffing around vessels may be minimal, but palisaded granulomas become more striking. Palisaded and neutrophilic granulomatous dermatitis may mimic granuloma annulare when viewed at scanning magnification.

In granuloma annulare, mucin is identified in areas of degeneration but not in palisaded neutrophilic granulomatous dermatitis. Palisaded neutrophilic granulomatous dermatitis does not exhibit the extensive areas of necrobiosis and plasma cell infiltration characteristic of necrobiosis lipoidica. Rheumatoid nodules are characterized by fibrin in the center. Infectious palisaded granulomas may show mixed patterns of inflammation, as well as stellate abscesses. Special stains, culture, and clinical history usually will help differentiate palisaded neutrophilic and granulomatous dermatitis from infectious etiologies. Rheumatoid neutrophilic dermatitis may appear to be similar to palisaded neutrophilic dermatitis histologically; however, rheumatoid neutrophilic dermatitis has a Sweet syndrome–like presentation clinically.[3]

Palisaded neutrophilic and granulomatous dermatitis may be associated with many different vasculitic or autoimmune diseases. Wegener's granulomatosis is characterized by necrotizing granulomatous inflammation and vasculitis of small and medium-sized vessels. The upper and lower respiratory tracts, the kidneys, and the eyes are most commonly affected. Sinusitis of several months' duration often is the initial manifestation. Skin biopsy results often are nonspecific. Lymphocytic infiltrates, purpura, leukocytoclastic vasculitis, and granulomatous vasculitis all may occur. Granulomatous vasculitis may closely resemble palisaded and neutrophilic dermatitis; however, granulomatous vasculitis is centered around blood vessels. Open lung biopsy results may exhibit geographic areas of necrosis. When glomerulonephritis is present, Wegener's granulomatosis is said to be generalized. Serologic evaluation typically reveals specific serum antineutrophil cytoplasmic antibodies directed to proteinase 3 and myeloperoxidase.

REFERENCES

1. Chu P, Connolly MK, LeBoit PE. The histopathologic spectrum of palisaded neutrophilic and granulomatous dermatitis in patients with collagen vascular disease. *Arch Dermatol*. 1994;130:1278-1283.
2. Finan MC, Winkelmann RK. The cutaneous extravascular necrotizing granuloma (Churg-Strauss granuloma) and systemic disease: a review of 27 cases. *Medicine (Baltimore)*. 1983;62:142-158.
3. Mashek HA, Pham CT, Helm TN, et al. Rheumatoid neutrophilic dermatitis. *Arch Dermatol*. 1997;133:757-760.