

Primary Anetoderma

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The authors report a case of primary anetoderma in a 27-year-old black woman. Primary anetoderma is characterized by circumscribed oval plaques with overlying loose, wrinkled skin appearing as a depression or pouch-like protrusion of the skin.

Anetoderma is caused by the loss of normal elastic fibers without any other apparent changes in the skin. It has been classified as a localized form of elastolysis. Anetoderma has been divided into primary and secondary forms. Primary anetoderma is characterized by circumscribed oval plaques with overlying loose, wrinkled skin appearing as a depression or pouch-like protrusion of the skin. Passing over a rim of unaffected skin, the examining finger dips into the lesion, as if it were a bladder-like sac. Areas of predilection include the trunk and proximal extremities. Secondary anetoderma is preceded by an identifiable lesion at the same site. Rare associations with systemic lupus erythematosus, syphilis, leprosy, acrodermatitis chronica atrophicans, and tuberculosis have been reported. We report a case of primary anetoderma in a 27-year-old black woman.

Case Report

A 27-year-old black woman presented with a progressive, asymptomatic eruption of several years' duration. No family members had similar lesions and she denied any trauma to the area prior to the eruption. On physical examination there were in excess of 100 non-inflamed, hypopigmented, atrophic plaques, 0.5 to 2 cm in diameter, predominantly on her thighs and upper



FIGURE 1. Non-inflamed, hypopigmented, atrophic plaque on the upper arm.



FIGURE 2. Demonstration of looseness of the skin.

arms, but also on her trunk (Figures 1 and 2). A skin biopsy was taken from an abdominal plaque, which included perilesional normal skin. Microscopic findings with hematoxylin-eosin section show sparse perivascular lymphocytic infiltrate with scattered dilated capillaries and a diminished number of adnexal structures

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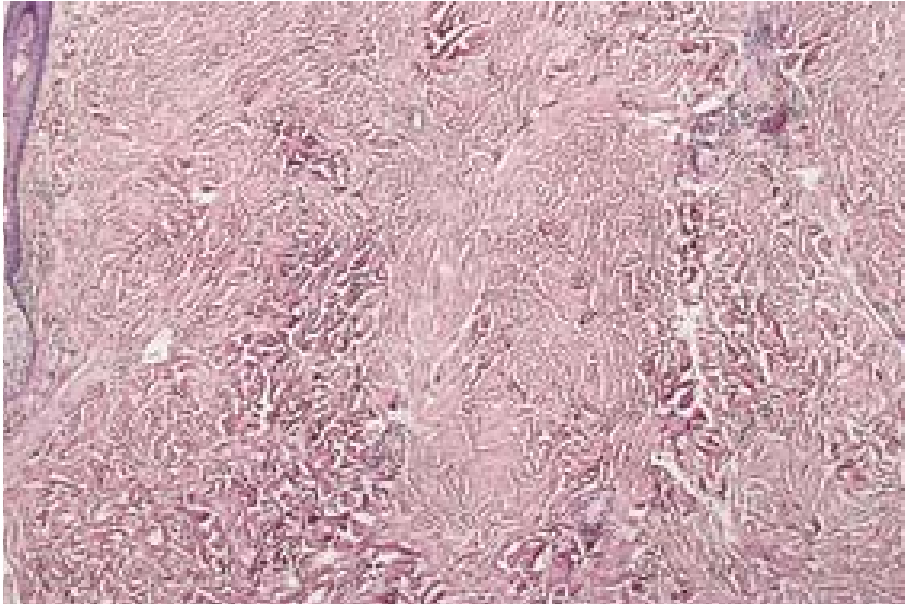


FIGURE 3. Microscopic findings show sparse perivascular lymphocytic infiltrate with scattered dilated capillaries and a diminished number of adnexal structures. The collagen bundles are in haphazard array (H&E; original magnification, X 50).

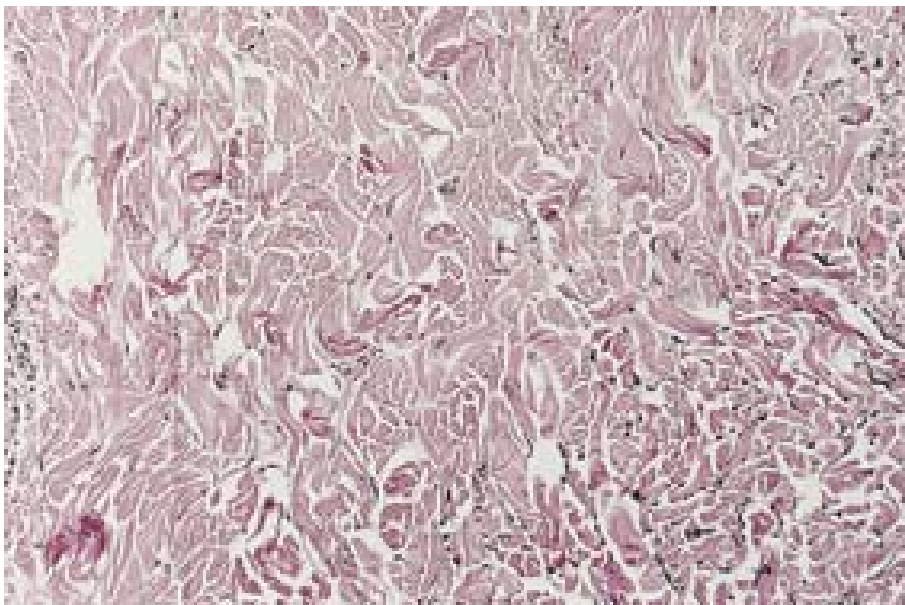


FIGURE 4. Nearly complete loss of elastic tissue throughout the lesion, in contrast to the adjacent built-in normal "control" (Elastin stain; original magnification, X 125).

(Figure 3). The collagen bundles were in haphazard array. Although this was subtle on hematoxylin-eosin-stained sections, elastin stain showed nearly complete loss of elastic tissue throughout the lesion, in contrast to the adjacent built-in normal "control" (Figure 4). Complete blood count, serum chemistry panel, anti-nuclear antibody, rapid plasma reagin, and lyme titer were within normal limits.

Comments

Primary anetoderma has been subclassified into a Jadassohn type, which is preceded by inflammation, and a Schweninger-Buzzi type, in which lesions appear without preceding inflammation. However, both types exhibit identical courses and prognoses. Fur-

thermore, histologic variations in inflammatory infiltrate and elastic tissue loss do not correlate with clinical appearance, disease course, or associated diseases.¹

The pathogenic mechanism underlying localized elastolysis has not been completely elucidated. Studies have demonstrated C3 and IgM in a fibrillar and granular pattern at the basement membrane zone and in the reticular and papillary dermis.^{2,5} Based on these findings, an immunologic mechanism of elastolysis has been suggested. However, these nonspecific findings also may be seen in systemic lupus erythematosus and lepromatous leprosy.⁶ Monoclonal antibody studies have shown the presence of substantial numbers of T cells, predominantly of the helper type (CD4+), which could represent a cell-mediated immune reaction directed against

elastic fibers or antigens uncovered by fragmentation of the fibers. Because of the localized destruction of elastin, attention has been focused on the role of elastase in the process. However, serum and tissue elastase levels have not been consistently elevated.^{3,7} Inflammatory cell (eg, macrophage) elastase release, which would not produce any appreciable elevation in serum or tissue elastase levels, could be the cause of the elastolysis.^{8,9}

Electron microscopy has confirmed the paucity of elastic fibers.⁷ The existing elastic structures appear ragged and irregular. The concentration of desmosine is decreased in lesional skin compared with unaffected perilesional skin and control subjects.⁷ The reduced desmosine concentration reflects the decrease in cross-linked elastin.

In most patients, disease activity persists for at least 15 years. Multiple treatment modalities, such as penicillin, phenytoin, dapsone, vitamin E, and nicotine, have been unsuccessful.²

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REFERENCES

1. Venecie PY, Winkelmann RK: Histopathologic findings in anetoderma. *Arch Dermatol* 120: 1040-1044, 1984.
2. Venecie PY, Winkelmann RK, Moore BA: Anetoderma: clinical findings, associations, and long-term follow-up evaluations. *Arch Dermatol* 120: 1032-1039, 1984.
3. Kossard S, Kronman KR, Dicken CH, et al: Inflammatory macular atrophy: immunofluorescent and ultrastructural findings. *J Am Acad Dermatol* 1: 325-334, 1979.
4. Hodak E, Shamaï-Lubovitz O, David N, et al: Primary anetoderma associated with a wide spectrum of autoimmune abnormalities. *J Am Acad Dermatol* 25: 415-418, 1991.
5. Hodak E, Shamaï-Lubovitz O, David M, et al: Immunologic abnormalities associated with primary anetoderma. *Arch Dermatol* 128: 799-803, 1992.
6. Quismorro FP, Rea TH, Levan NE, et al: Immunoglobulin deposits in lepromatous leprosy skin: presence of deposits in apparently uninvolved skin and occurrence of serum antiepithelial antibodies. *Arch Dermatol* 111: 331-334, 1975.
7. Oikarinen AL, Palatsi R, Adomian GE, et al: Anetoderma: biochemical and ultrastructural demonstration of an elastin defect in the skin of three patients. *J Am Acad Dermatol* 11: 64-72, 1984.
8. Venecie PY, Winkelmann RK: Monoclonal antibody studies in the skin lesions of patients with anetoderma. *Arch Dermatol* 121: 747-749, 1985.
9. Venecie PY, Winkelmann RK, Moore BA: Ultrastructural findings in the skin lesions of patients with anetoderma. *Acta Derm Venereol* 64: 112-120, 1984.