cutis® Photo Quiz

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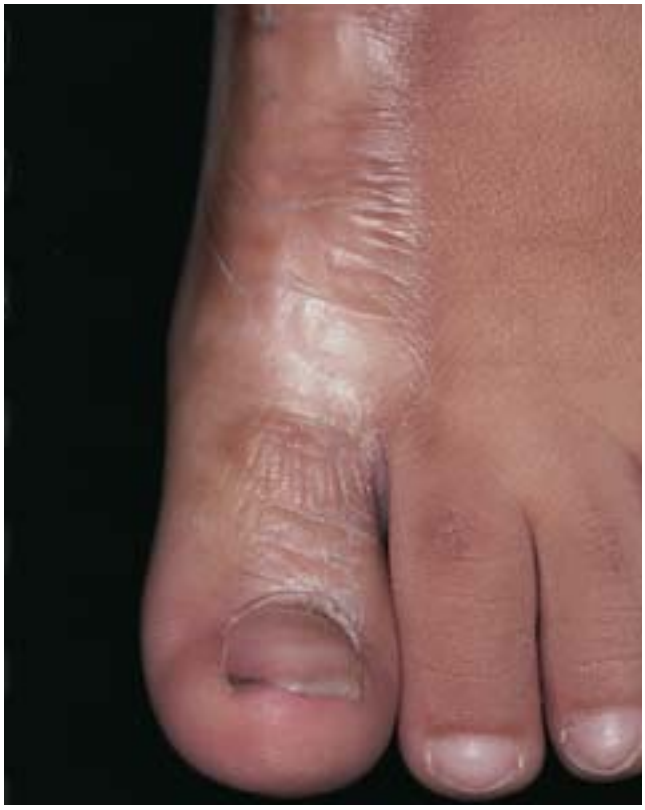

A 5-year-old black girl presented with a several-year history of a linear sclerotic plaque that progressed from her left proximal thigh to her distal left first toe. She also had hypopigmented, round-to-oval plaques located on her left trunk.

What is your diagnosis?

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The Diagnosis: Linear Scleroderma

Linear scleroderma consists of linear, bandlike, sclerotic lesions that usually are asymmetric and often are associated with atrophy of the soft tissue, muscle, and underlying bone. The lesions commonly affect the lower extremities (Figure 1). Scleroderma in children is rare, can affect all races, and can begin at any age, with boys being affected almost as frequently as girls.[1-3] Children are more likely to develop localized forms of scleroderma (morphea, generalized morphea [Figure 2], guttate morphea, nodular morphea, subcutaneous morphea, and linear scleroderma) than systemic sclerosis.[1,4-7] The etiology of scleroderma is unknown. The role of *Borrelia burgdorferi* has been explored but not confirmed definitively.[4]

Plaque morphea, in its inflammatory stage, is characterized by an area of induration surrounded by a violaceous halo. As the lesions progress, the affected skin develops a waxy ivory color, and atrophic hypopigmented or hyperpigmented areas may occur.[3] Scleroderma en coup de sabre describes frontoparietal involvement of the face and scalp, often with alopecia of the scalp and eyelids. This condition may progress down the face, with intracranial abnormalities such as calcification and atrophy of underlying soft tissue and bone.[4] The symptoms of scleroderma en coup de sabre may overlap with those of Parry-Romberg syndrome.

Diagnosis of scleroderma is established by clinical morphology and biopsy. Histopathologic examination reveals compact, hyalinized, sclerotic collagen in the dermis and subcutis, with atrophy of appendages. Perivascular lymphoid infiltrates, particularly at the dermal-subcutaneous junction are noted in the inflammatory phase. The linear and deep morphea types of scleroderma (morphea profunda) are characterized by deeper sclerosis, with involvement of fascia and muscle.[1] Speckled or homogeneous antinuclear antibody patterns, and ssDNA antibodies may be noted.

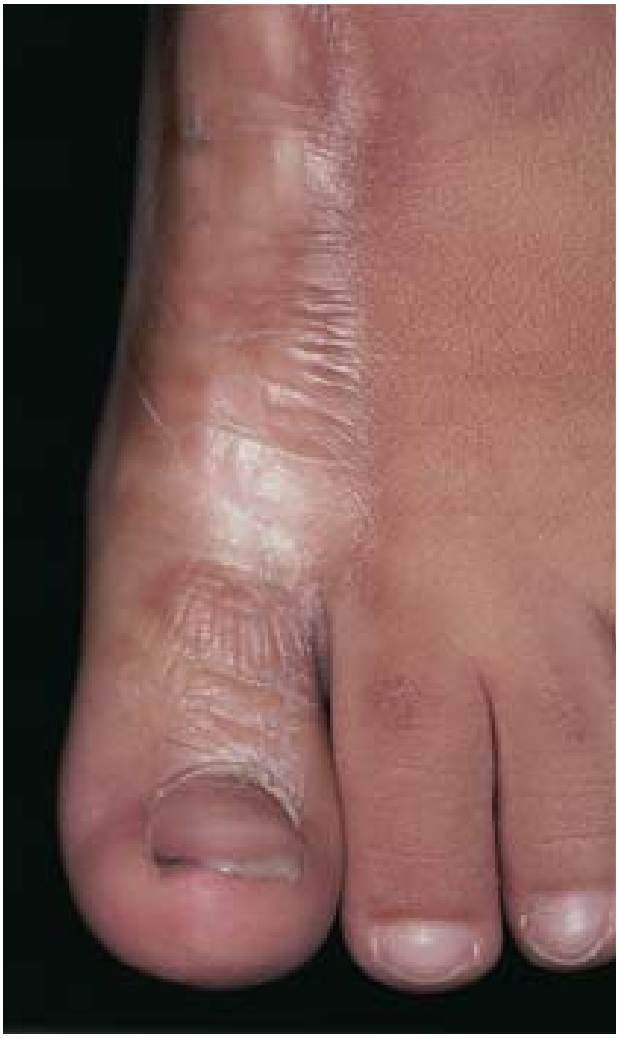

Figure 1. Sclerotic lesion of morphea involving skin and soft tissue.

Scleroderma may cause serious deformities, especially in small children. Loss of function and contractures are common around joints in linear scleroderma. Cranial involvement may be associated with uveitis, seizures, hemiplegia, and learning problems.[2] Localized scleroderma is difficult to treat. However, mild cases of localized scleroderma may be self-limiting and may not require treatment. Early physical therapy should be used to preserve range of motion and function. For more

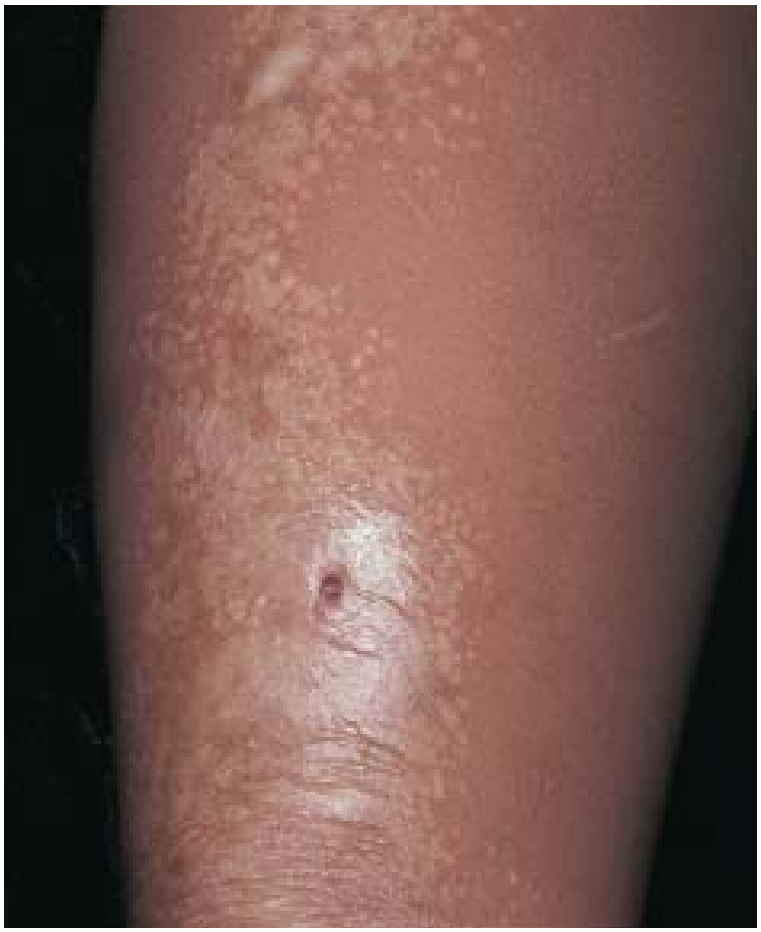

Figure 2. Upper portion of lesion with guttate pattern.

widespread or rapidly progressive cases, systemic steroids, penicillamine, plasmapheresis, chloroquine or hydroxychloroquine, PUVA, and methotrexate have been used with anecdotal success.[1,4,6] Surgical intervention rarely is indicated, and poor wound healing can be common. However, surgery may be beneficial to relieve flexion contractures and for facial scleroderma.

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