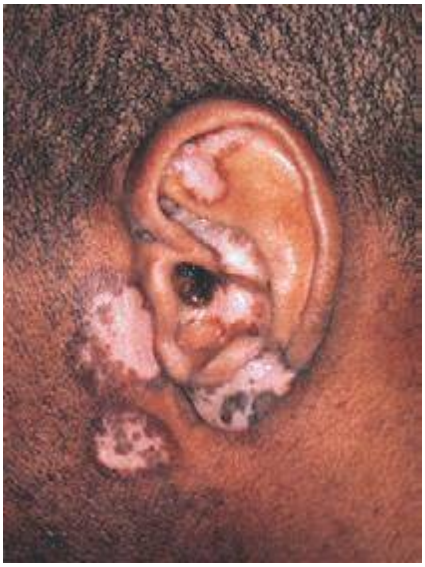

FIGURE 73-6 Discoid (chronic cutaneous) lupus erythematosus. Violaceous, hyperpigmented, atrophic plaques, often with evidence of follicular plugging that may result in scarring, are typical.

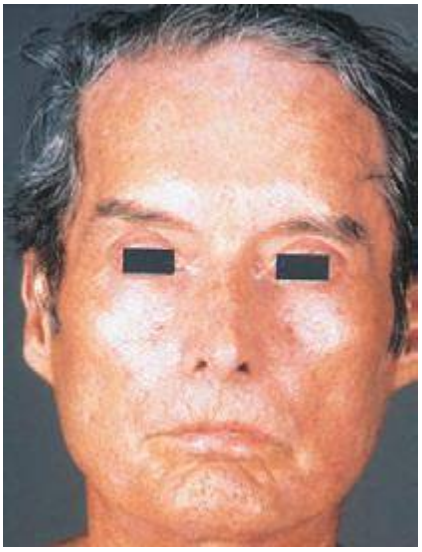

FIGURE 73-8 Scleroderma often eventuates in development of an expressionless, masklike facies.

become sausage-shaped, and, because the fingernails are usually unaffected, they may curve over the end of the fingertips. Periungual telangiectases are usually present, but periungual erythema is rare. In advanced cases, the extremities show contractures and calcinosis cutis. Facial involvement includes a smooth, unwrinkled brow, taut skin over the nose, shrinkage of tissue around the mouth, and perioral radial furrowing (Fig. 73-8). Matlike telangiectases are often present, particularly on the face and hands. Involved skin feels indurated, smooth, and bound to underlying structures; hyper- and hypopigmentation are common as well. *Raynaud's phenomenon* (i.e., cold-induced blanching, cyanosis, and reactive hyperemia) is documented in almost all patients and can precede development of scleroderma by many years. *Linear scleroderma* is a limited form of disease that presents in a linear, bandlike distribution and tends to involve deep as well as superficial layers of skin. The combination of calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia has been termed the *CREST syndrome*. Centromere antibodies have been reported in a very high percentage of patients with the CREST syndrome but in only a small minority of patients with scleroderma. Skin biopsy reveals thickening of the dermis and homogenization of collagen bundles. Direct immunofluorescence microscopy of lesional skin is usually negative.

Morphea is characterized by localized thickening and sclerosis of skin; it dominates on the trunk. This disorder may affect children or adults. Morphea begins as erythematous or flesh-colored plaques that become sclerotic, develop central hypopigmentation, and have an erythematous border. In most cases, patients have one or a few lesions, and the disease is termed *localized morphea*. In some patients, widespread cutaneous lesions may occur without systemic involvement (*generalized morphea*). Many adults with generalized morphea have concomitant rheumatic or other autoimmune disorders. Skin biopsy of morphea is indistinguishable from that of scleroderma. Scleroderma and morphea are usually quite resistant to therapy. For this reason, physical therapy to prevent joint contractures and to maintain function is employed and is often helpful. Treatment options for early, rapidly progressive disease include phototherapy (UVA1 or PUVA) or methotrexate (15–20 mg/week) alone or in combination with daily glucocorticoids.

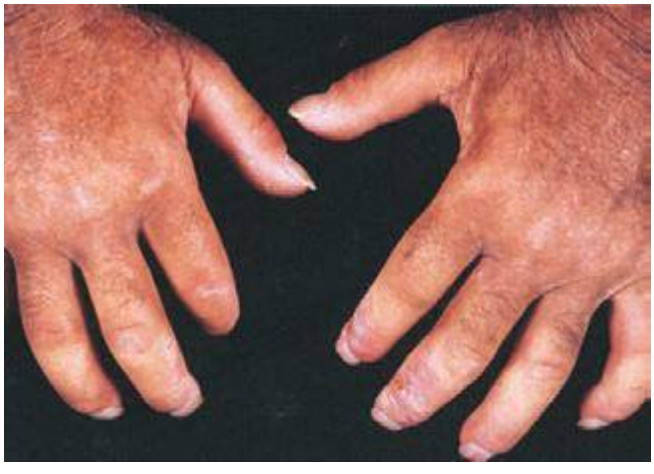

FIGURE 73-7 Scleroderma showing acral sclerosis and focal digital ulcers.

Diffuse fasciitis with eosinophilia is a clinical entity that can sometimes be confused with scleroderma. There is usually a sudden onset of swelling, induration, and erythema of the extremities, frequently following significant physical exertion. The proximal portions of the extremities (upper arms, forearms, thighs, calves) are more often involved than are the hands and feet. While the skin is indurated, it usually displays a woody, dimpled, or "pseudocellulite" appearance rather than being bound down as in scleroderma; contractures may occur early secondary to fascial involvement. The latter may also cause muscle groups to be separated and veins to appear depressed (i.e., the "groove sign"). These skin findings are accompanied by peripheral-blood eosinophilia, increased erythrocyte sedimentation rate, and sometimes hypergammaglobulinemia. Deep biopsy of affected areas of skin reveals inflammation and thickening of the deep fascia overlying muscle. An inflammatory infiltrate composed of eosinophils and mononuclear cells is usually found. Patients with eosinophilic fasciitis appear to be at increased risk for developing bone marrow failure or other hematologic abnormalities. While the ultimate course of eosinophilic fasciitis is uncertain, many patients respond favorably to treatment with prednisone in doses of 40–60 mg/d.

The *eosinophilia-myalgia syndrome*, a disorder with epidemic numbers of cases reported in 1989 and linked to ingestion of L-tryptophan manufactured by a single company in Japan, is a multisystem disorder characterized by debilitating myalgias and absolute eosinophilia in association with varying combinations of arthralgias, pulmonary symptoms, and peripheral edema. In a later phase (3–6 months after initial symptoms), these patients often develop localized sclerodermatous skin changes, weight loss, and/or neuropathy (Chap. 382). The precise cause of this syndrome, which may resemble other sclerotic skin conditions, is unknown. However, the implicated lots of L-tryptophan contained the contaminant 1,1-ethylidene bis[tryptophan]. This contaminant may be pathogenic or may be a marker for another substance that provokes the disorder.