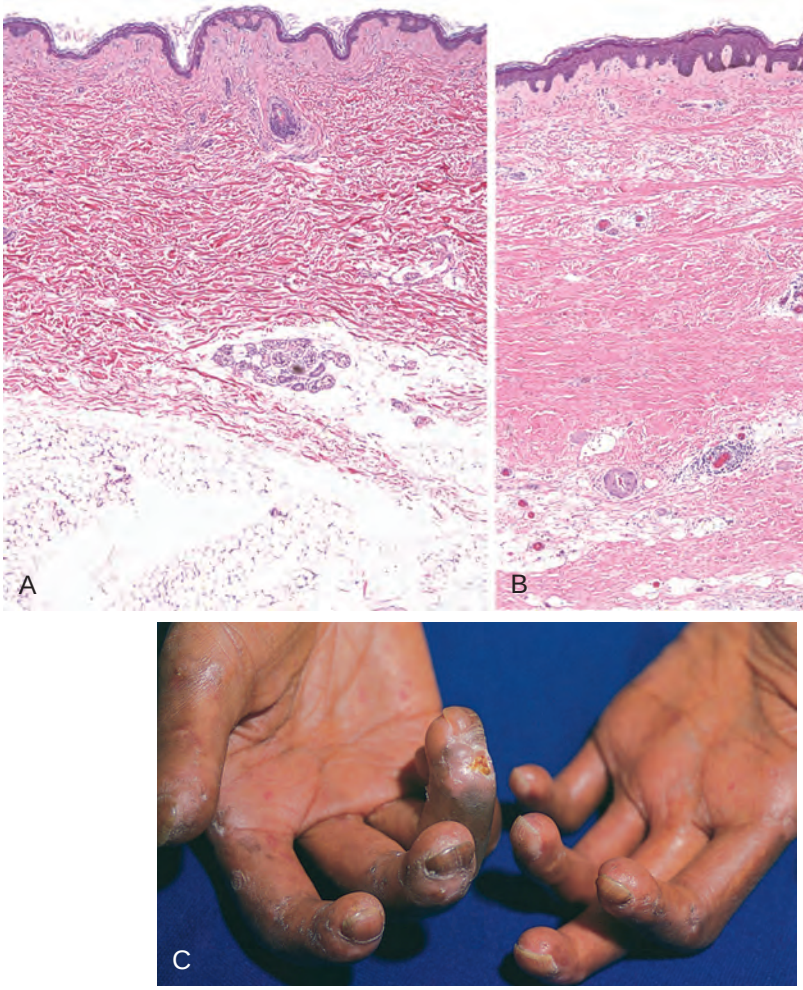

Figure 6-31 Systemic sclerosis. **A,** Normal skin. **B,** Skin biopsy from a patient with systemic sclerosis. Note the extensive deposition of dense collagen in the dermis with virtual absence of appendages (e.g., hair follicles) and foci of inflammation *(arrow)*. **C,** The extensive subcutaneous fibrosis has virtually immobilized the fingers, creating a clawlike flexion deformity. Loss of blood supply has led to cutaneous ulcerations. (**C,** Courtesy Dr. Richard Sontheimer, Department of Dermatology, University of Texas Southwestern Medical School, Dallas, Texas.)

atony and dilation, especially at its lower end. Abdominal pain, intestinal obstruction, or malabsorption syndrome with weight loss and anemia reflect involvement of the small intestine. Respiratory difficulties caused by the pulmonary fibrosis may result in right-sided cardiac dysfunction, and myocardial fibrosis may cause either arrhythmias or cardiac failure. Mild proteinuria occurs in as many as 30% of patients, but rarely is the proteinuria severe enough to cause a nephrotic syndrome. The most ominous manifestation is malignant hypertension (Chapter 11), with the subsequent development of fatal renal failure, but in its absence progression of the disease may be slow. The disease tends to be more severe in blacks, especially black women. As treatment of the renal crises has improved, pulmonary disease has become the major cause of death in systemic sclerosis.

Virtually all patients have ANAs that react with a variety of nuclear antigens. Two ANAs strongly associated with systemic sclerosis have been described. One of these, directed against *DNA topoisomerase* I (anti-Scl 70), is highly specific. Depending on the ethnic group and the assay, it is present in 10% to 20% of patients with diffuse systemic sclerosis. Patients who have this antibody are more likely to have pulmonary fibrosis and peripheral vascular disease. The other, an *anticentromere antibody*, is found in 20% to 30% of patients, who tend to have the CREST syndrome. Patients with this syndrome have relatively limited involvement of skin, often confined to fingers, forearms, and face, and calcification of the subcutaneous tissues. Involvement of the viscera, including esophageal lesions, pulmonary hypertension, and biliary cirrhosis, may not occur at all or occur late. In general these patients live longer than those with systemic sclerosis with diffuse visceral involvement at the outset.

KEY CONCEPTS

Systemic Sclerosis

- Systemic sclerosis (commonly called scleroderma) is characterized by progressive fibrosis involving the skin, gastrointestinal tract, and other tissues.