A

B

FIGURE 382-5 Cutaneous vascular changes. ***A.*** Capillary changes at the nailfold in a patient with limited cutaneous systemic sclerosis (lcSSc). ***B.*** Telangiectasia on the face.

to firm adherence of skin to the underlying platysma muscle interfere with neck extension. The face assumes a characteristic "mauskopf" appearance with taut and shiny skin, loss of wrinkles, and occasionally an expressionless facies due to reduced mobility of the eyelids, cheeks, and mouth. Thinning of the lips with accentuation of the central incisor teeth and fine wrinkles (radial furrowing) around the mouth complete the picture. Reduced oral aperture (microstomia) interferes with eating and oral hygiene. The nose assumes a pinched, beak-like appearance.

In established SSc, the skin is firmly bound to the subcutaneous fat (tethering) and undergoes thinning and atrophy. Telangiectasias are dilated skin capillaries 2–20 mm in diameter frequently seen in lcSSc. These lesions, reminiscent of hereditary hemorrhagic telangiectasia, are prominent on the face, hands, lips, and oral mucosa (Fig. 382-5). A greater number of telangiectasias correlates with the extent of microvascular complications, including PAH. Breakdown of atrophic skin leads to chronic ulcerations at the extensor surfaces of the proximal interphalangeal joints, the volar pads of the fingertips, and bony prominences such as the elbows and malleoli. Ulcers are painful and may become secondarily infected, resulting in osteomyelitis. Healing of ischemic fingertip ulcerations leaves characteristic fixed digital "pits." Loss of soft tissue at the fingertips due to ischemia is frequent and may be associated with striking resorption of the terminal phalanges (acro-osteolysis) (Fig. 382-6).

Calcium deposits (calcinosis) in the skin and soft tissues occur in patients with lcSSc who are positive for anticentromere antibodies. The deposits, varying in size from tiny punctate lesions to large conglomerate masses, are composed of calcium hydroxyapatite crystals and can be readily visualized on plain x-rays. Frequent locations include the finger pads, palms, extensor surfaces of the forearms, and

FIGURE 382-6 Acro-osteolysis. Note dissolution of terminal phalanges in a patient with long-standing limited cutaneous systemic sclerosis (lcSSc) and Raynaud's phenomenon.

the olecranon and prepatellar bursae (Fig. 382-7). They may occasionally ulcerate through the overlying skin, producing drainage of chalky white material, pain, and local inflammation. Paraspinal soft tissue calcifications may cause neurologic complications.

PULMONARY FEATURES

Pulmonary involvement is frequent in SSc and is the leading cause of death. The two principal forms are ILD and pulmonary vascular disease. Patients with SSc frequently develop some degree of both complications. Less frequent pulmonary manifestations include aspiration pneumonitis complicating chronic gastroesophageal reflux, pulmonary hemorrhage due to endobronchial telangiectasia, obliterative bronchiolitis, pleural reactions, restrictive ventilatory defect due to chest wall fibrosis, spontaneous pneumothorax, and drug-induced lung toxicity. The incidence of lung cancer is increased.

Interstitial Lung Disease (ILD) Evidence of ILD can be found in up to 90% of patients with SSc at autopsy and 85% by thin-section high-resolution computed tomography (HRCT). In contrast, clinically significant ILD develops in 16–43%; the frequency varies depending on the detection method used. Risk factors include male gender, African-American race, diffuse skin involvement, severe gastroesophageal reflux, and the presence of topoisomerase I autoantibodies, as well as a low forced vital capacity (FVC) or single-breath diffusing capacity of the lung for carbon monoxide (DLco) at initial presentation. In these patients, the most rapid progression in lung disease occurs early in

FIGURE 382-7 Calcinosis cutis. Note large calcific deposit breaking through the skin in a patient with limited cutaneous systemic sclerosis (lcSSc).