Extensive Calcinosis Cutis in Limited Cutaneous Scleroderma

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The CREST syndrome is a subset of limited scleroderma characterized by calcinosis cutis, Raynaud's phenomenon (RP), esophageal dysmotility, sclerodactyly, and telangiectasias. The calcinosis of skin and soft tissues results from deposition of calcium hydroxyapatite crystals and occurs to a different extent in 20%–40% of patients. The exact mechanism of calcinosis associated with scleroderma is not known and no effective drug therapy exists, therefore the therapeutic approach is mostly symptom-oriented. Sometimes a surgical intervention, especially in paraspinal calcifications, may be necessary.

A 71-year-old woman presented with calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and facial telangiectasias 20 years ago. She was diagnosed with CREST syndrome, now classified as lim-

ited cutaneous scleroderma (lcSSc)^{1,2}. Antinuclear antibodies and anticentromere antibodies were positive³. She had never received specific therapy, and concomitant arthralgias occasionally were treated with nonsteroidal antiinflammatory drugs. Physical examination revealed marked skin thickening of hands, forearms, feet, and the face, as well as multiple indurated, confluent, yellowish nodules on hands and forearms bilaterally. Radiographs of hands (Figure 1) and forearms (Figure 2) showed extensive confluent calcifications typically seen in lcSSc. Followup visits in our outpatient clinic during the last 2 years revealed a stable extent of calcification and skin thickening, but increasing complications caused by esophageal dysmotility.

Figure 1. Radiograph of hands shows extensive confluent calcifications typically seen in lcSSc.

Figure 2. Radiograph of forearms also shows extensive calcifications.

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