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Images in Rheumatology

Cutaneous calcinosis in a patient with limited scleroderma: CREST Syndrome

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Cutaneous calcinosis in a patient with limited scleroderma: CREST Syndrome

Calcinosis or dystrophic soft-tissue calcification occurs in damaged or devitalized tissues in the presence of normal calcium/phosphorus metabolism. It is a known manifestation in the subcutaneous tissues of patients with connective tissues diseases, especially scleroderma, systemic lupus erythematosus, or dermatomyositis, and may involve a relatively localized areas or present as widespread calcinosis (1). Little is known about its physiopathology. It occurs in tissues that are under chronic stress, such as local trauma or damage associated with underlying inflammatory processes (2).

Subcutaneous calcinosis occurs in all subsets of scleroderma but is more prominent in patients with limited scleroderma and in those with anticentromere antibody. CREST syndrome is a limited form of scleroderma, characterized by calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia (3).

A 46-year-old woman suffered from CREST syndrome for 15 years. She had sclerotic cutaneous findings on her face and fingers (sclerodactyly), prominent facial telengectasia, Raynoud's phenomenon, and esophageal reflux. There were also multiple hardened erythematous-whitish nodules, some having a chalky appearance, around her knees (Figure 1). X-ray showed extensive calcinosis in the soft tissue of the knees (Figure 2). Pulmonary arterial pressure without pulmonary fibrosis was found to be 40

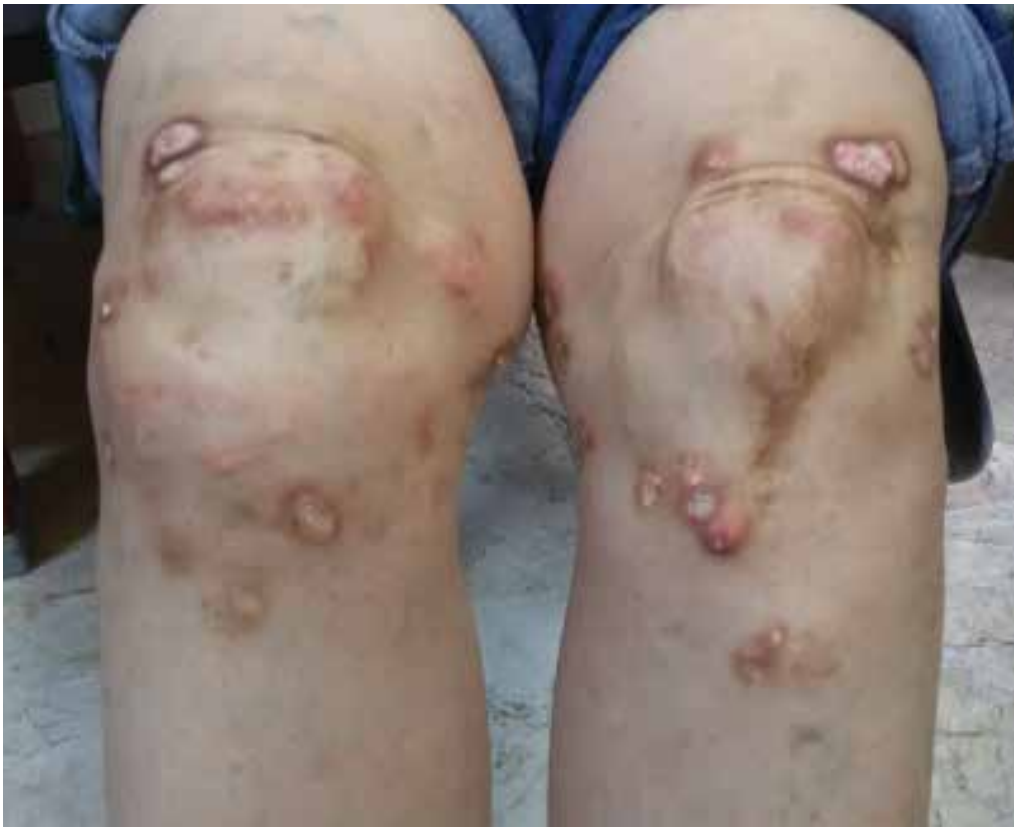

Figure 1. Multiple erythematous and whitish papules/nodules, some of which have a chalky white material on the around knees

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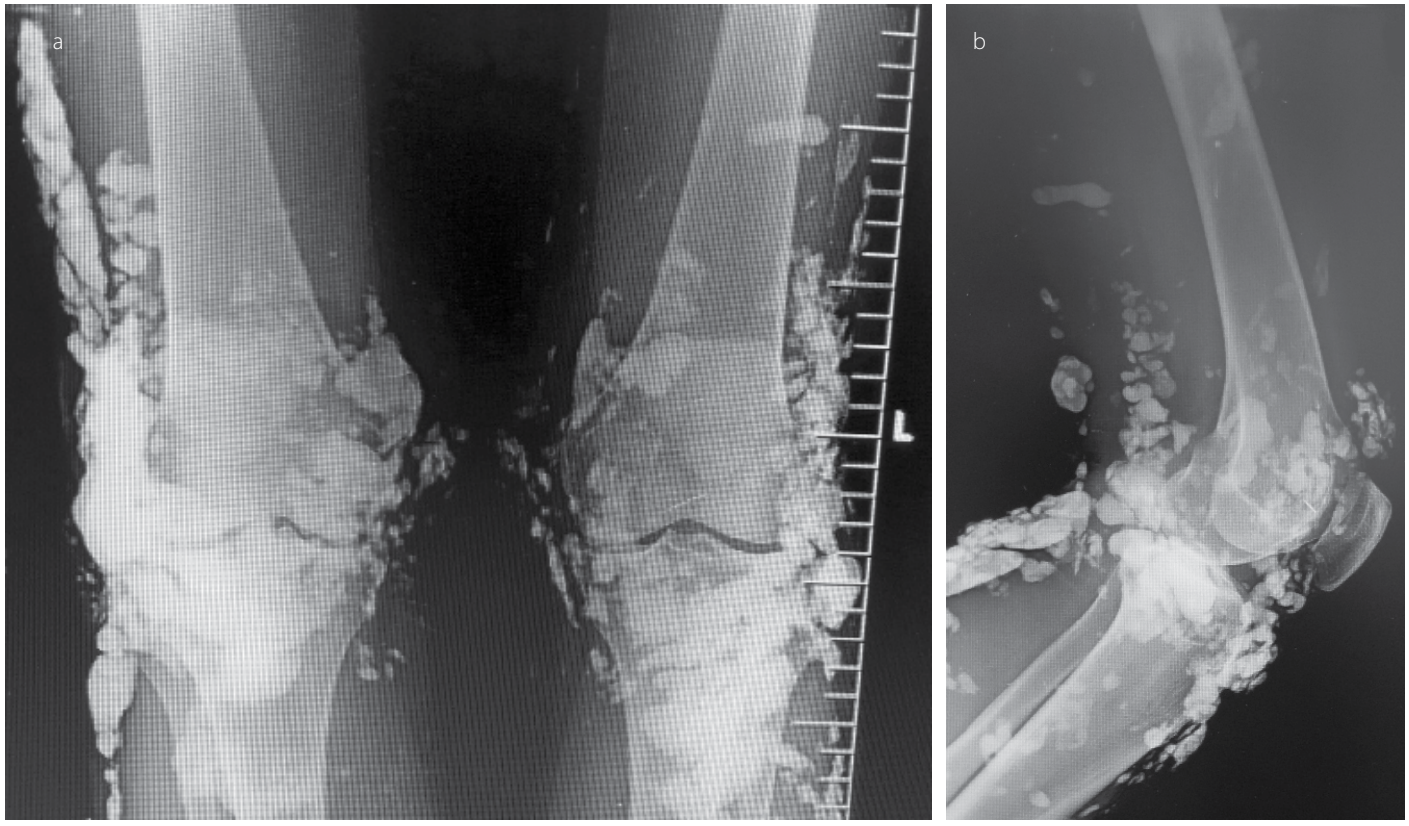

Figure 2. a, b. Axial (a) and lateral (b) radiographic images of knee show wide-spread and multiple radio-opacities on the periarticular area

mmHg using echocardiography. Antinuclear and anti-centromere antibodies were positive. Serum calcium and phosphorus levels were within the normal ranges. She had been treated with nifedipine, low dose aspirin, and colchicine for many years. Treatment was initiated with aluminum hydroxide. Medical therapy for cutaneous calcinosis is limited and has variable benefits. Multiple treatment approaches with diltiazem, disodium etidronate, probenecid, colchicine, minocycline, low-dose warfarin, and intralesional adrenal steroids have been explored, but no standard treatment has convincingly prevented or reduced calcinosis.

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