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Images in Rheumatology

Scleroderma Renal Crisis: Clues From the Physical Exam

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Scleroderma renal crisis (SRC) is a rare, life-threatening complication of systemic sclerosis (SSc) and can sometimes be the first manifestation of the disease.[1]

A 56-year-old female presented with acute encephalopathy requiring intubation and a systolic blood pressure of 230 mmHg; no information was available about her medical history. Physical examination revealed foreshortened fingers (Figure 1A) and salt-and-pepper skin changes (Figure 1B) without sclerodactyly. Hand radiographs (Figure 2) demonstrated acro-osteolysis and calcinosis cutis. Laboratory studies showed renal failure, low haptoglobin, elevated lactate dehydrogenase, and thrombocytopenia; schistocytes were observed on peripheral blood smear. Based on these findings, she was diagnosed with SRC. She was started on captopril but subsequently developed angioedema, requiring a switch to losartan. She achieved normotension, and her encephalopathy resolved. At that time, she reported experiencing distal tuft loss for the past 3 years and Raynaud phenomenon for 1 year.

Although the patient required renal replacement therapy for 28 days, her renal function recovered while being treated with losartan. Three months after discharge, she maintained independent renal function with a serum creatinine of 2.2 mg/dL. Further evaluation ultimately revealed a positive test result for Scl-70 antibodies and nonspecific interstitial pneumonia.

SSc is one of the most common causes of salt-and-pepper skin changes and acro-osteolysis, characterized by resorption of the distal phalanx leading to foreshortened fingers.[2] SRC should always be considered in patients with new or rapidly progressive renal injury and hypertensive encephalopathy. These unique findings can help elucidate this diagnosis quickly, permitting the prompt initiation of renal protective and lifesaving therapies.

REFERENCES

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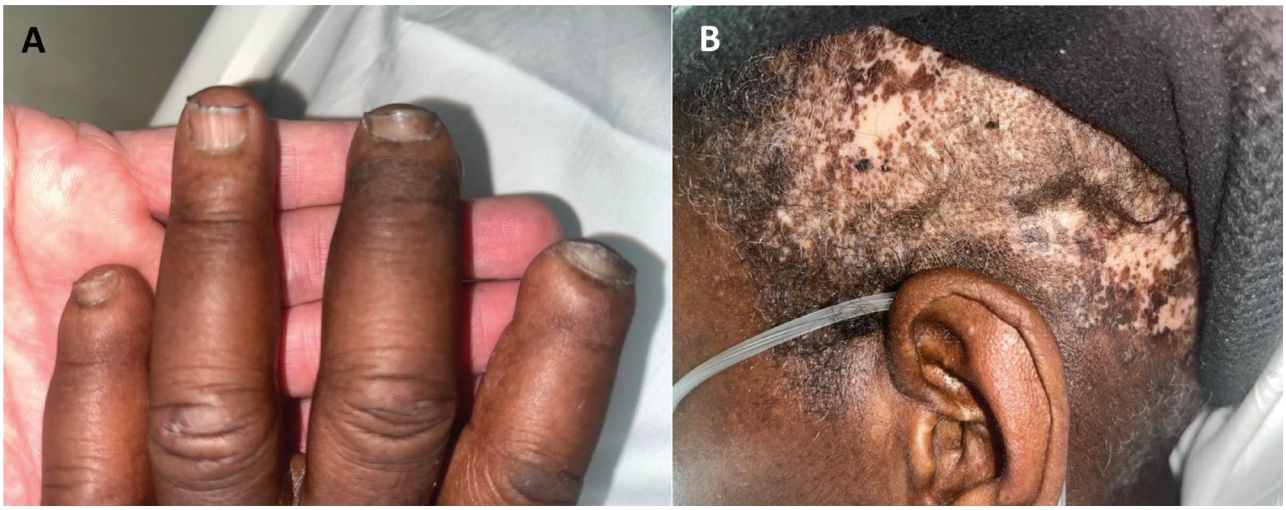

Figure 1. Physical exam revealed (A) loss of the terminal tufts of both hands as well as (B) salt-and-pepper changes involving the scalp.

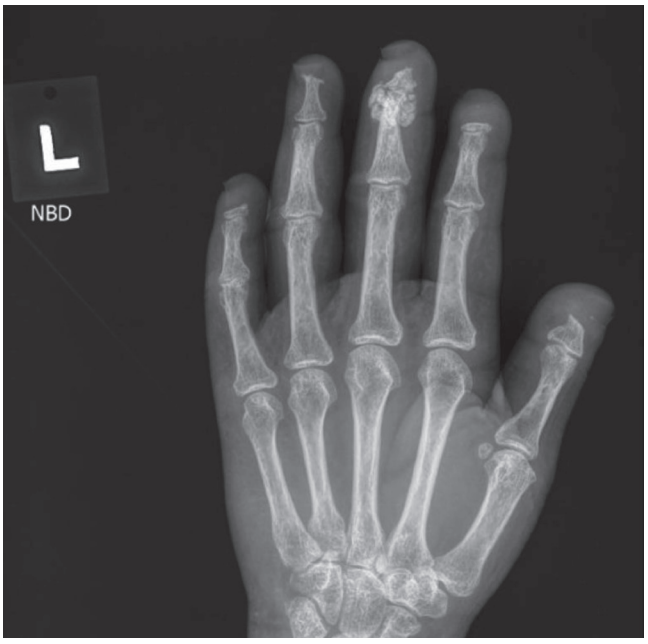

Figure 2. Hand radiographs demonstrated calcinosis cutis and acro-osteolysis most profoundly affecting the left hand.