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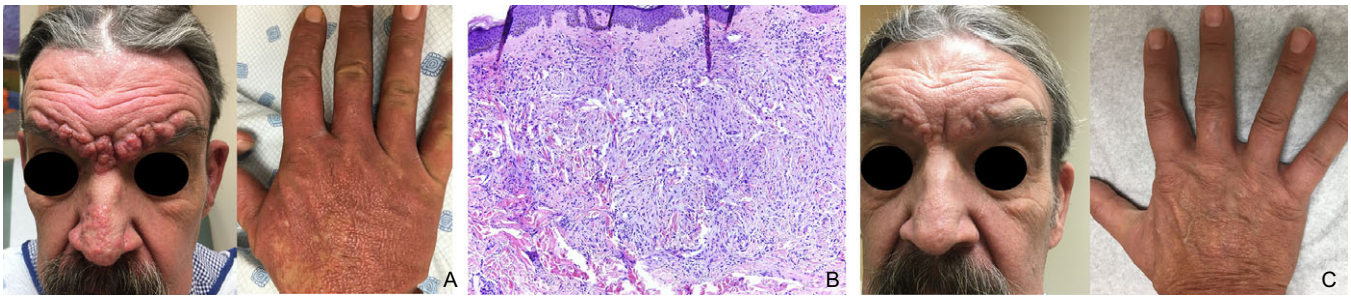
Clinical Images: Monoclonal gammopathy–associated scleromyxedema presenting as leonine facies



The patient, a 61-year-old man previously in good health, was referred to the scleroderma clinic for a 2-year history of slowly progressive cutaneous eruption involving the dorsal hands, extremities, and central area of the face. Physical examination revealed nodular, erythematous, indurated lesions on the forehead and erythematous papular lesions on the nose with coalescence of firm erythematous papulonodules, resulting in a leonine facies. On the dorsal hands, arms, and legs were numerous, shiny, firm, closely set, slightly translucent papules measuring 1–2 mm with background erythema (**A**). Skin biopsy demonstrated a spindled fibroblastic proliferation in the dermis with increased mucin and variable fibrosis (**B**). The clinical and histologic findings were diagnostic of scleromyxedema. Scleromyxedema is a rare disorder of unknown pathogenesis characterized by a generalized lichenoid papular cutaneous eruption and resulting in diffuse skin induration that may simulate scleroderma. Rarely, larger exophytic nodules, as seen in this patient, may be present. The majority of scleromyxedema cases occur in association with a monoclonal gammopathy. The patient was found to have an IgGλ M protein spike. This patient did not exhibit any CRAB features (hypercalcemia, renal insufficiency, anemia, and bone lesions), and evaluation including hematologic studies culminated in a diagnosis of IgGλ monogammopathy of unclear significance, with plans for ongoing observation. For his scleromyxedema, the patient received intravenous immunoglobulin (IVIG) at doses of up to 2 gm/kg/month (1), with significant improvement in the appearance of lesions after 16 months (**C**). He continues to receive a maintenance dose of 1 gm/kg IVIG every 4 weeks.

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