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Leonine facies presenting as scleromyxedema

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Figure Caption:

A 61-year-old man previously in good health was referred to Scleroderma Clinic for a 2-year history of slowly progressive cutaneous eruption involving his dorsal hands, extremities, and central face. On physical examination, the patient had nodular, erythematous indurated lesions of his forehead and erythematous papular lesions on his 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, 1-2 mm slightly translucent papules with background erythema (face and hand, shown in Panel A). Skin biopsy demonstrated a spindled fibroblastic proliferation in the dermis with increased mucin and variable fibrosis (Panel B). The clinical and histologic findings are diagnostic of scleromyxedema. Scleromyxedema is a rare disorder of unknown pathogenesis characterized by a generalized lichenoid papular cutaneous eruption resulting in diffuse skin induration that may simulate scleroderma. Rarely, larger

exophytic nodules, as seen in this patient, may be present. The vast majority of cases of scleromyxedema occur in association with a monoclonal gammopathy. Our patient was found to have an IgG lambda M-protein spike. The patient did not exhibit any CRAB features (hypercalcemia, renal insufficiency, anemia, and bone lesions) and evaluation with hematology culminated in diagnosis of IgG lambda monogammapathy of unclear significance (MGUS), with plans for ongoing observation. For his scleromyxedema, the patient received intravenous immunoglobulin (IVIG) [1] at doses up to 2g/kg/month, with significant improvement in appearance of his lesions after 16 months (Panel C). He continues to receive a maintenance dose of 1 gram/kg IVIG every 4 weeks.

Reference:

1. Blum M, Wigley FM, Hummers LK. Scleromyxedema: A Case Series Highlighting Long-Term Outcomes of Treatment With Intravenous Immunoglobulin (IVIG). *Medicine (Baltimore)*. 2008;87(1):10. doi:10.1097/MD.0b013e3181630835

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