Scleromyxedema

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A 52-year-old man presented to our outpatient dermatologic clinic with a 6 month history of generalized asymptomatic eruptions. Physical examination revealed multiple 0.1 cm to 0.3 cm firm, waxy, and closely spaced papules with partial confluence on the dorsal hands, forearms, scalp, face, neck, upper back, and thighs (Figure 1: Panels A–D). On the dorsal part of the proximal interphalangeal joints, a central depression was surrounded by an elevated rim, which presented a donut-like appearance (Panel A). The skin of the glabella, upper back, and proximal arms was thickened and indurated with deep, longitudinal furrows, presenting typical characteristics of a “leonine face” (Panel C) and “Shar-Pei sign” (Panel D). Skin biopsy revealed scleromyxedema. Laboratory tests revealed normal thyroid function, and serum protein immunoelectrophoresis and immunofixation revealed IgG monoclonal gammopathy. After treatment with 2 g/kg intravenous immunoglobulin and glucocorticoid pulse therapy, the skin eruptions improved considerably.

Keywords: Dermatologic clinic; Scleromyxedema.

Declaration of Interests

The authors declare that they have no competing interests.

Figure 1: Scleromyxedema Panel A: Donut-like appearance in Fingers Panel B: Donut-like appearance in Neck Panel C: Leonine face Panel D: Shar-Pei sign.

Clinical Image

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