A case of generalized scleromyxedema that responded to IVIG monotherapy
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INTRODUCTION

• Generalized scleromyxedema is an idiopathic papular mucinosis that typically affects middle-aged adults.
• It is often associated with a concurrent IgG monoclonal gammopathy and elevated lambda light chains. The role of this gammopathy in this condition remains a debate.
• Due to the rarity of this disease and a lack of clinical trials, treatment guidelines have not yet been established.

CASE PRESENTATION

• A 49-year-old African-American female presented initially with a mildly pruritic rash that originated on her bilateral thighs.
• The lesions subsequently disseminated to her bilateral upper and lower extremities and face over several months.
• Physical examination revealed erythematous monomorphic papules coalescing into thick pebbly plaques on her upper and lower extremities, more prominent on her left forearm and bilateral anterior thighs. Similar lesions were also present on the bridge of her nose and bilateral helices of the ears.
• A biopsy demonstrated fibroblastic proliferation within the dermis, mild dermal lymphocytic inflammation and increased mucin in the superficial dermis. The Alcian blue stain for mucin was positive.
• Laboratory work up revealed a normal thyroid stimulating hormone (TSH) and IgG kappa monoclonal gammopathy on serum protein electrophoresis (SPEP).
• Further work up by hematology demonstrated a 10-15% plasma cell myeloma that did not require treatment.
• Patient was started on subsequent IVIG 2g/kg spread over five days, once a month for six months. She had 90% improvement of her skin lesions.

FIGURES 1-4: Lesions on the body

FIGURES 5 & 6: Histopathology

DISCUSSION

• Scleromyxedema is characterized by symmetrical distribution of small waxy or lichenoid papules coalescing to form thick plaques with sclerodermoid changes.
• Patients with scleromyxedema can develop internal disease involving neurologic, cardiac, rheumatologic, muscular, respiratory and renal organs.
• Four criteria are needed to make the diagnosis of scleromyxedema: 1) generalized papular eruption or sclerodermoid features, 2) histologic features of mucin deposition, fibroblast proliferation and fibrosis, 3) monoclonal gammopathy, and 4) absence of thyroid disorder.
• The association between IgG monoclonal lamda gammopathy or plasma cell dyscrasia and scleromyxedema remains unclear. However, serum levels of these proteins do not correlate with disease progression or severity. In fact, only 10% of the patients progress to frank multiple myeloma.
• IVIG has been used as first line monotherapy and is administered as 2g/kg/month. The patients treated on this regimen experience improvement in skin tightness and thickness after the first or second cycle.
• For patients that do not respond to IVIG monotherapy, addition of systemic steroids or other treatments can result in partial or complete remission.
• More recently, combinations of thalidomide and dexamethasone, lenalidomide, bortezomib and autologous hematopoetic stem cell transplant have been tried with varying levels of success.

REFERENCES