INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

A RARE CASE OF SCLEROMYXEDEMA TREATED WITH METHYLPREDNISOLONE AND CYCLOSPORINE

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Background: Scleromyxedema is a rare chronic cutaneous mucinosis of unknown etiology. Diagnosis of scleromyxedema established if there are generalized/diffuse papular and sclerodermoid eruption, microscopic triad (mucin deposition composed primarily of hyaluronic acid in dermis, fibrosis, and irregularly arranged fibroblast proliferation), monoclonal gammopathy, and absence of thyroid disorder. The etiology of this disease remains an enigma, the precise mechanisms whereby increased fibroblast activity results in mucin deposition remain to be defined. To date, there is no completely satisfactory therapeutic approach to scleromyxedema. The rarity of the disorder, combined with the lack of well-designed clinical trials studying the disease, translates to a "therapeutic ladder" based on anecdotal reports and small cases series. The objective of this report was to report a case of scleromyxedema treated with methylprednisolon and cyclosporine.

Observation: A male, 58-year-old, presented with generalized papular and sclerodermoid eruption accompanied with Shar-Pei sign on the back and both of knee, doughnat sign on the proximal interphalangeal joint of fingers hand. Upon histopathological examination of the skin lesion, fibroblast proliferation and fibrosis were found on hematoxylin eosin staining. Alcian blue staining was positif and showed deposition mucin on the dermis. Laboratoy examination showed monoclonal gammopathy and absence of thyroid disorder, wich supported the diagnosis of scleromyxedema. The patient was treated with oral methylprednisolon 28mg/day and cyclosporine 100mg/day. Clinical improvement was seen on day 39th after receiving therapy.

Key message: The efficacies of the various treatments that have been utilized remain unclear, furthermore no specific treatment appears to be uniformly effective. Corticosteroid and cyclosporine reported has a good result for treatment of scleromyxedema in several cases.