

SHORT COMMUNICATION

Clinicopathologic Correlation: Differentiating Between Localized Lichen Myxedematosus And Scleromyxedema

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Scleromyxedema, also known as sclerodermoid or generalized lichen myxedematosus, is a rare type of primary cutaneous dermal mucinosis associated with monoclonal gammopathy, systemic symptoms, and normal thyroid studies.¹ Clinically, scleromyxedema can have a variable presentation with potential for relapse. Histologically, scleromyxedema can be indistinguishable from localized lichen myxedematosus (LM).² Unlike patients with LM, patients with scleromyxedema may develop severe complications such as dermato-neuro syndrome, which is a potentially fatal condition characterized by a flu-like prodrome followed by fever, seizures, and coma.³

It is important to differentiate scleromyxedema from LM early due to the risk of progressive systemic disease. IVIG is generally regarded as first-line therapy for scleromyxedema. Second line-agents include thalidomide or lenalidomide paired with systemic glucocorticoids. Melphalan, Bortezomib with dexamethasone, or autologous hematopoietic stem cell transplantation can be utilized for severe or refractory disease.⁴

We report the case of a 38-year-old female with chronic arthritis who presented with an

11-month history of biopsy-proven LM that was recalcitrant to treatment with topical steroids and methotrexate. On physical exam, she had numerous, 2mm, monomorphic, dome-shaped papules on the lateral cheeks, upper dorsolateral arms, and anterior legs. Repeat punch biopsy of the right upper arm demonstrated increased mucin deposition separating thickened collagen fibers and increased fibroblasts in the superficial dermis consistent with “scleromyxedema/lichen myxomatous.”

Upon further review of her medical records, our patient had a history of epilepsy and monoclonal IgG gammopathy. Given the patient’s constellation of symptoms and prior medical history, she was diagnosed with scleromyxedema. We referred her to oncology and began treatment with IVIG, after which she experienced an improvement in symptoms and complete resolution of her skin lesions.

Within 5 months of discontinuing IVIG, our patient’s skin disease relapsed. On repeat examination, she had scattered 2mm dome-shaped papules on the dorsal hands similar to her prior presentation. She also had scattered 5-8mm faintly violaceous macules on the upper and lower dorsal extremities. Biopsies of both lesion types revealed mildly

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increased fibroblasts and dermal mucin, reconfirming the diagnosis of scleromyxedema. Our patient is scheduled to reinstitute IVIG therapy with the goal of achieving clearance of her skin lesions.

In the absence of long-term, continuous treatment, scleromyxedema has the potential to progress and evolve clinically.⁴ Scleromyxedema can be distinguished from LM through careful history and physical exam. Earlier correlation of this patient's dermatologic presentation with her systemic signs including monoclonal gammopathy, epilepsy, and arthritis may have prompted sooner initiation of the appropriate treatment for scleromyxedema. Given that prognosis and management of LM and scleromyxedema differ, patients with histologic evidence of a cutaneous mucinosis should be screened for evidence of systemic involvement.

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