

Discrete Papular Lichen Myxedematosus with an Unusual Segmental Presentation

Discrete papular lichen myxedematosus, a subtype of localized papular lichen myxedematosus, is an idiopathic cutaneous mucinosis. It typically presents symmetrically, involving the trunk and extremities. We report on an unusual case of discrete papular lichen myxedematosus with a segmental presentation.

An otherwise healthy 23-year-old man presented with a gradual accumulation of asymptomatic papules on the lower abdomen over the previous year. Physical examination revealed multiple, well-defined, discrete and coalescing, 1-5mm, smooth, indurated, dark brown, slightly erythematous papules, with variable surrounding hyperpigmentation, involving the skin of the right lower abdomen in a segmental distribution (Figure 1). A punch biopsy specimen was obtained. Histopathological examination revealed mucin deposition in the superficial to mid-reticular dermis, moderate proliferation of fibroblasts, and an



Figure 1. Segmentally distributed 1-5 mm, indurated, dark brown, slightly erythematous papules with variable surrounding hyperpigmentation.

inflammatory infiltrate composed of plasma cells and lymphocytes (Figures 2A, B). The clinical and pathologic features were consistent with discrete papular lichen myxedematosus.

The laboratory work-up, including a complete blood count with differential and liver function tests,

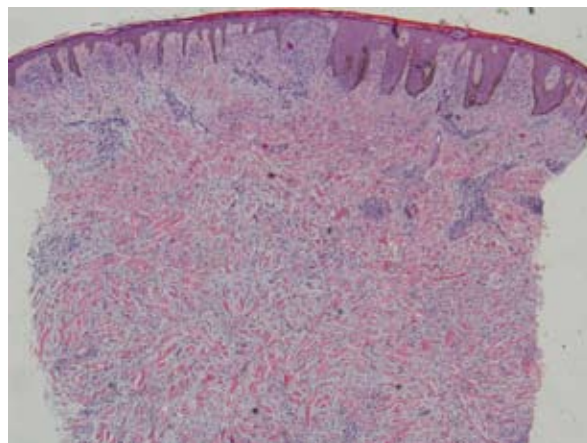


Figure 2A.

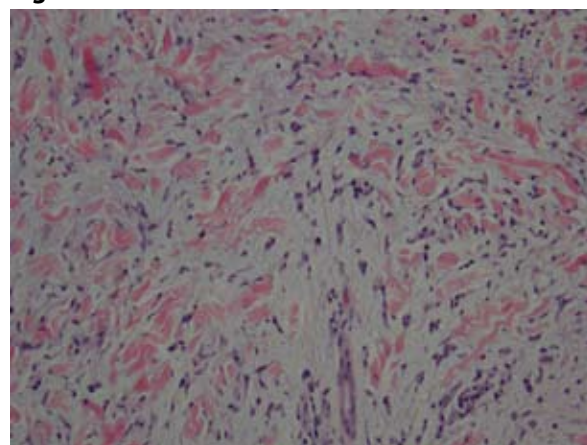


Figure 2B

Figure 2. Mucin deposition in the superficial to mid-reticular dermis with a moderate proliferation of fibroblasts (H&E $\times 40$ (A), $\times 200$ (B)).

renal panel, and serum protein electrophoresis, was within normal limits. Human immunodeficiency virus (HIV), hepatitis B, and hepatitis C antibodies were negative.

The patient was treated with fluocinonide 0.05% ointment twice daily and intralesional triamcinolone 5-7.5 mg/ml monthly for 3 months resulting in resolution of most of the lesions with significant flattening of the remaining ones (Figure 3).

Lichen myxedematosus (LM) is an idiopathic cutaneous mucinosis divided into 2 clinicopathologic subsets: scleromyxedema and localized papular LM (1,2).

Scleromyxedema is a severe, progressive, symmetric eruption of closely-set, often linear, 2-3 mm, firm, waxy papules with surrounding induration favoring the head, neck, upper trunk, forearms, hands, and thighs. It is almost always associated with a paraproteinemia and may have systemic involvement, including musculoskeletal, neurologic, pulmonary, gastrointestinal, renal, and cardiovascular manifestations. Histologically, there is diffuse mucin deposition in the upper and mid reticular dermis, increased collagen deposition, and a marked proliferation of fibroblasts (1,2).

Localized papular LM presents as firm, skin-colored to red-brown papules or nodules involving the trunk and extremities without sclerotic features, paraproteinemia, or systemic involvement (1). Histologically, it is characterized by focal to diffuse mucin deposition in the upper and mid reticular dermis, a variable proliferation of fibroblasts, and an absent to mild increase in collagen (1,3). It is further subdivided into 4 subtypes: discrete papular LM, a symmetric eruption of few to hundreds of 2-5 mm papules, sometimes coalescing into nodules or plaques (2-4); acral persistent papular mucinosis, exclusively involving the dorsal aspects of the hands and distal forearms (2); cutaneous mucinosis of infancy, affecting the trunk and upper arms, especially the elbows, of infants (2); and nodular LM, presenting as multiple 3 mm to 2 cm nodules with a mild or absent papular component (3,5,6).

LM may be associated with HIV and hepatitis C infections, toxic oil syndrome, and L-tryptophan-associated eosinophilia-myalgia syndrome. Additionally, atypical forms exist presenting as scleromyxedema without monoclonal gammopathy and localized papular LM with monoclonal gammopathy or systemic symptoms (1).

Successful therapy for discrete papular LM with intralesional steroids combined with flurandrenolide tape has been reported (7). Topical steroids alone



Figure 3. Lesions after treatment with topical and intralesional steroids.

have been variably successful (3,8). Tacrolimus ointment cleared 2 patients (8) and pimecrolimus cream provided symptomatic relief in 1 patient (3). Our patient is improving with topical and intralesional steroids.

We present this case to raise awareness of the unusual segmental presentation of discrete papular LM. Discrete papular LM typically presents symmetrically and has not been described in a segmental distribution. To our knowledge, this is the first case of segmental discrete papular lichen myxedematosus.

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