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Lichen Myxedematosus: Case Report and Review of Literature

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ABSTRACT

Lichen myxedematosus (LM) is an idiopathic cutaneous mucinosis, commonly described as localized scleromyxedema. In contrast to scleromyxedema, there is typically no systemic involvement. Treatment options are limited and spontaneous resolution has been reported.

We present the case of a 66-year-old Hispanic male referred by his primary care physician for evaluation of asymptomatic dark spots on his trunk and extremities present for about one-year. Physical exam revealed smooth, brown hyperpigmented papules coalescing into plaques on the trunk. Multiple well-demarcated oval dark brown plaques measuring 3 cm in size were located on the upper back, periumbilical area, bilateral lower extremities, and buttocks. A diagnosis of lichen myxedematosus was made based on histologic features observed in the dermis.

There are 5 subtypes of LM: a discrete papular form, acral persistent papular mucinosis, self-healing papular mucinosis, papular mucinosis of infancy, and a pure nodular form. Occasional patients with LM have atypical features or features intermediate between scleromyxedema and localized LM. We present a case of atypical LM with mixed features of the different subtypes. Herein we will review the varied clinical presentations of LM and highlight the distinguishing features of scleromyxedema.

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INTRODUCTION

Lichen Myxedematosus (LM), also referred to as papular mucinosis, was first described in 1906 by Dubreuilh. In 1953, Montgomery and Underwood classified LM into subsets which were revised in 2001 by Rongioletti and Rebora. Rongioletti and Rebora categorized LM into three subsets: 1) scleromyxedema, 2) localized, and 3) atypical.[1] LM is characterized by lichenoid papules, nodules, and/or plaques due to dermal mucin deposition with varying degrees of fibrosis in the absence of thyroid disease.[2] We present the case of a patient diagnosed with LM.

CASE REPORT

A 66-year-old Hispanic male presents with a one-year history of asymptomatic dark spots on his trunk and extremities. Physical exam revealed smooth papules coalescing into thin plaques on the trunk (Figure 1). Multiple well-demarcated oval dark brown plaques measuring about 2-3 cm in size were located on the upper back, periumbilical area, bilateral lower extremities and buttocks (Figure 2a, 2b). Lab values were within normal limits and showed a normal electrophoretic pattern. Histology showed increased mucin in the papillary dermis and mildly increased spindle cells and macrophages (Figure 3a, 3b). A diagnosis of LM was made. Halobetasol propionate 0.05% ointment twice daily resulted in improved texture and decreased pigmentation of skin.

FIGURE 1. Hyperpigmented papules coalescing into plaques without epidermal change, smooth on palpation and shiny in appearance distributed on upper back and upper chest.

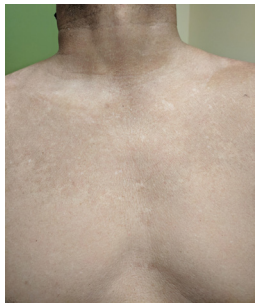


FIGURE 2. Multiple well demarcated oval dark brown plaques measuring ~3 cm in size, located (A) peri-umbilically and (B) on the upper back. There is no epidermal change, but excoriations are present on some of the lesions.

(A) (B)

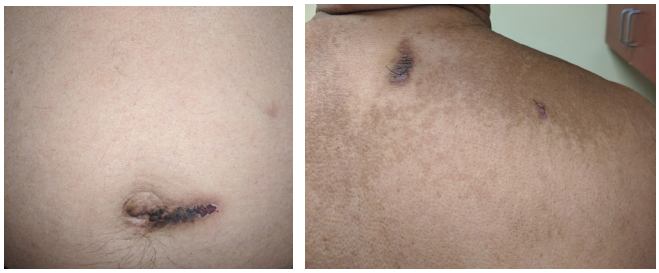
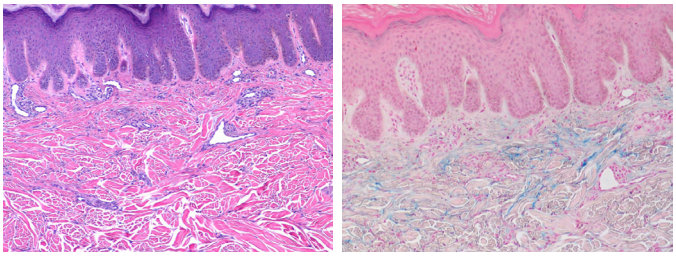


FIGURE 3. Histopathology of lesional skin showing increased mucin in papillary dermis and mildly increased spindle cells and macrophages. (A) Hematoxylin and eosin, 10x (B) Colloidal iron, 10x.

(A) (B)



DISCUSSION

The first subset of LM, scleromyxedema, has four defining criteria: 1) generalized papular and sclerodermoid eruption, 2) mucin deposition, fibroblast proliferation and fibrosis, 3) monoclonal gammopathy, and 4) absence of thyroid disease. Scleromyxedema is equally present in both sexes with a mean age at diagnosis of 59.[2,3] It is characterized by widespread, symmetric, 2-3 mm firm, waxy, closely spaced papules in a linear pattern. The surrounding skin is indurated and may display erythema, edema, red-brown discoloration and pruritus. Scleromyxedema involves the face and neck, unlike localized LM. The microscopic triad of scleromyxedema is 1) interstitial mucin, 2) increased collagen deposition in the upper reticular dermis, and 3) marked proliferation of irregularly arranged fibroblasts. As a result, patients experience skin thickening, stiffening, and decreased mobility of the joints. A superficial lymphocytic and plasmocytic infiltrate is also often present. IgG λ paraproteinemia is seen in 83.2% of cases.[2] Characteristic signs of scleromyxedema are "leonine facies" due to deep longitudinal furrowing of the glabella and papular thickening of the ears, "shar pei sign" due to deep furrowing on the back, and "doughnut sign" due to skin thickening causing an elevated rim with a central depression on the extended proximal interphalangeal joint.[2,3] Scleromyxedema may be differentiated from systemic sclerosis by cutaneous findings of telangiectasias, nailfold changes, or calcinosis, which are present in the latter.[2,3] Extracutaneous manifestations of scleromyxedema include myositis, arthralgias, Raynaud's syndrome, Sjogren's disease, central nervous system disturbances, carpal tunnel syndrome, dysphagia, macular edema, ectropion, and lagophthalmos. Mucin deposition in pulmonary and coronary vessels has led to restrictive and obstructive lung disease, and hypertension, atherosclerosis, and myocardial infarction, respectively. Dermatoneuro syndrome is a common cause of death in scleromyxedema. Symptoms include fever, confusion, dysarthria, lethargy, convulsions, and coma. CT is normal and brain biopsy shows gliosis and demyelination, rather than mucin deposition.[2,4] Death has also resulted from myeloid leukemia, Hodgkin lymphoma, and myocardial insufficiency.[4] The etiology of scleromyxedema is unknown. Fibroblast overstimulation of IL-1, TNF-a, and TGF-beta has been proposed.[5] There is no definitive treatment due to its poorly understood pathogenesis. Intravenous immunoglobulin has proven more effective than corticosteroids for both cutaneous and extracutaneous manifestations.[4] Spontaneous resolution is also possible, even after 15 years.[2,4]

The second subset of LM, localized, has three defining criteria: 1) papular or nodular/plaque eruption, 2) mucin deposition with variable fibroblast proliferation, and 3) absence of both monoclonal gammopathy and thyroid disease. Localized LM is further divided into 5 subtypes: 1) discrete papular, 2) acral persistent papular mucinosis, 3) self-healing papular mucinosis, 4) papular mucinosis of infancy, and 5) nodular form.[2,13] The discrete papular type presents with symmetric, 2-5mm firm, smooth, waxy or flesh colored papules involving the trunk and limbs. The face is spared, and skin is not indurated. Fourteen cases of HIV preceding LM have been reported, 13 discrete and one acral persistent. Discrete LM is also associated with Hepatitis C.[1,2,6] It has been proposed that HIV stimulates mucin and collagen production in LM.[5] Acral persistent papular mucinosis is more common in females (4.7:1) and presents with multiple ivory to flesh colored papules exclusively on the dorsal hands, extensor surface of the wrists, and occasionally the distal forearms.[2] Self-healing papular mucinosis is divided into juvenile and adult variants. In the juvenile form, mucin in periarticular nodules leads to arthralgias, weakness, and fever.[2,7,13] These symptoms are rare in the adult variant. Histologically, there is diffuse mucin deposition in the upper reticular dermis.[13] The self-healing subtype resolves spontaneously within weeks to months.[2,7] Papular mucinosis of infancy is often considered the pediatric equivalent of the discrete or acral subtypes, and presents with firm, opalescent papules on the upper arms, and trunk.[2] In childhood, localized LM must be differentiated from mucinous nevus, a neoplastic hamartoma. Mucinous nevus presents as mucinous lesions with nevoid features in a unilateral, linear or dermatomal pattern. Histologically, band-like mucin deposits are seen in the

papillary dermis.[13] Lastly, the nodular form, also known as atypical tuberous myxedema of Jadassohn Dosseker, presents with multiple nodules on the limbs and trunk with absent or mild papular eruption.[2,8] Treatment is not required for asymptomatic, localized LM.[6] Interestingly, a case study of two morbidly obese women demonstrated a correlation of weight loss with complete clinical and histopathologic resolution of cutaneous lesions.[9]

The third subset of LM includes atypical or intermediate forms that do not meet criteria for scleromyxedema or localized LM. This subset includes 1) scleromyxedema without monoclonal gammopathy, 2) localized LM with monoclonal gammopathy and/or systemic symptoms, 3) localized LM with mixed features of the five subtypes, and 4) not well specified cases. The course of atypical forms is unpredictable.[2,9]

CONCLUSION

This case is unusual because of the multiple morphologies seen clinically, which is a rare presentation. It is important to distinguish the chronic, disabling course of scleromyxedema, associated with systemic and fatal manifestations, from the treatment-independent localized form of LM.[2] Topical application of corticosteroids, pimecrolimus, or tacrolimus may be of some benefit, however, spontaneous resolution is most common, even in the setting of HIV infection.[2] With the revised diagnostic criteria, mean time from primary symptoms to diagnosis has decreased from 41.6 to 10 months.[4] Further research is needed to adequately diagnose and treat LM.

DISCLOSURES

The authors declare that they have no relevant conflicts of interest to disclose.

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