

Discrete Papular Lichen Myxoedematosus: A Case Report

Diskret Papüler Liken Miksödematöz: Bir Olgu Sunumu

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ABSTRACT

Lichen myxoedematosus is an uncommon, chronic disorder characterized by lichenoid papules due to dermal mucin deposition. Discrete papular mucinosis is a rare subtype of the localized form. Herein, we report a 49-year-old woman with asymptomatic, white-coloured papules on her upper back. A skin biopsy from a papule on her upper back demonstrated dermal mucin deposition after alcian blue staining.

Key words: exanthema; lichens; scleromyxedema; skin

ÖZET

Liken miksödematöz, dermal müsin depolanmasından dolayı likenoid papüllerle karakterize olan nadir görülen, kronik bir hastalıktır. Diskret papüler müsinoz lokalize formun nadir görülen bir alt tipidir. Burada, sırtının üst kısmında asemptomatik, beyaz renkli papülleri olan 49 yaşında kadın hastayı sunuyoruz. Sırtın üst kısmındaki papülden alınan cilt biyopsisi, Alcian mavisi boyama sonrasında müsin depolanmasını gösterdi.

Anahtar kelimeler: ekzantem; likenler; skleromiksödem; cilt

Introduction

Cutaneous mucinoses are a wide group of disorders characterized by anomalous deposits of mucin in the skin. Lichen myxoedematosus can be classified into several subtypes, depending on the distribution and overall skin involvement. Discrete papular lichen myxoedematosus (DPLM) is an uncommon subtype of primary cutaneous mucinoses. Only five of the cases described in the literature to date showed no relation

with systemic diseases. Herein, we report the first case of a patient with discrete white papules with mucin deposition that appeared localized to the back.

Case Report

A 49-year-old woman was referred to our department with asymptomatic papules of 3 months' duration on the upper back. Physical examination revealed numerous well-defined, white papules, 3–5 mm in diameter (Figure 1). There wasn't any induration or thickening of the skin on the affected site. In addition, there was no history of preceding trauma and the patient was not taking any medication.

Routine laboratory studies were unremarkable. Serum electrophoresis and thyroid function tests were normal. Screening for viral infections remained negative. General physical examination did not reveal any systemic involvement.

A 4-mm punch biopsy was performed from one of the lesions located on the upper part of the back. Histopathological examination showed normal epidermis and widely separated collagen fibers (Figure 2). PAS + Alcian Blue staining showed an increase in blue staining, which indicated an increase in dermal mucin (Figure 3). Based on the clinical, histological and laboratory findings we have diagnosed the discrete papular type of lichen myxoedematosus.

Although topical corticosteroids had been previously administered for three months, they had been ineffective. Because the patient was asymptomatic, we preferred to follow conservatively. During the follow-up of 20 months, we did not observe any evidence of spontaneous resolution or progression.



Figure 1. Well-defined, white papules demonstrated on upper site of the patient's back.

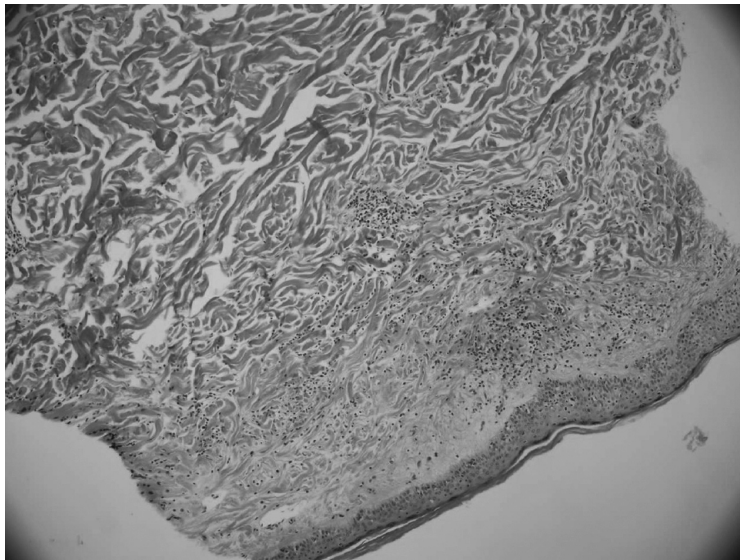


Figure 2. Normal epidermis and widely separated collagen fibers demonstrated in histopathological examination.

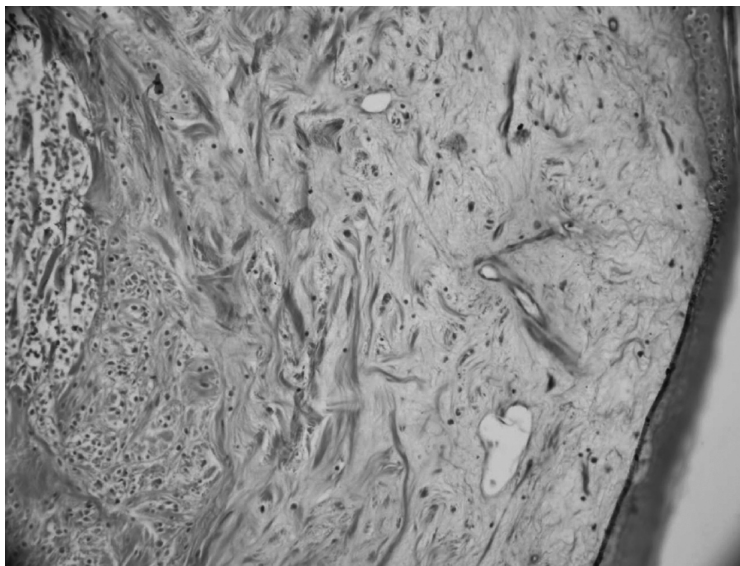


Figure 3. PAS and Alcian Blue staining showed an increase in blue stain indicating an increase in dermal.

Discussion

Cutaneous mucinoses are a wide group of disorders characterized by anomalous deposits of mucin in the skin¹. Lichen myxoedematosus can be classified into several subtypes, depending on the distribution and overall skin involvement. Discrete papular lichen myxoedematosus (DPLM) is an uncommon subtype of primary cutaneous mucinoses².

Papular mucinosis is a rare and chronic disorder which was described by Montgomery and Underwood in 1953³. The cutaneous focal mucinoses are a group of connective tissue disorders characterized by deposition of mucin found in the middle and deeper layers of the dermis, displacing collagen fibers but not involving the dermal papillae or accumulating around blood vessels⁴.

Rongioletti et al. suggested that lichen myxoedematosus included clinic-pathological subsets: a generalized papular and sclerodermoid form with systemic effects (also called scleromyxedema) and localized papular forms without systemic effects. The localized papular subtype was further subdivided into acral persistent papular mucinosis, papular mucinosis of infancy, self-healing papular mucinosis and a discrete papular or nodular form²⁻⁵. We classified our patient as the discrete papular form.

The pathophysiology of cutaneous mucinosis is not well-known. Cytokines, paraproteins, chronic antigenic stimulation, viral infections, and inflammation may contribute to the pathogenesis.

DPLM is a very uncommon type of cutaneous primary mucinosis, which affects both genders equally. There have been only five cases unrelated to systemic diseases reported previously in the literature. Lesions are usually found on the face, neck, trunk or extremities in a symmetrical pattern. Lesions are flesh-colored to erythematous, smooth surfaced and approximately 2–5

mm in diameter. To the best of our knowledge, this is the first case of discrete papular mucinosis with white colored papules ever reported in the literature.

Cutaneous eruptions may be generalized or localized in papular mucinosis. Systemic organ dysfunction does not occur in localized papular mucinosis. There was no organ involvement in our patient.

The treatment of mucinosis is difficult. Spontaneous healing has rarely been observed. As it is limited to the skin and has little or no morbidity, no systematic treatment approach is an acceptable option⁵. Transition to scleromyxedema has not yet been reported.

In conclusion, we report a case of DPLM in an otherwise healthy woman. The results of clinical examinations and laboratory tests were all negative for conditions typically associated with mucin deposition disorders and our patient had no improvement in her lesions during the follow-up of 20 months.

References

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