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Self-healing Cutaneous Mucinosis in an Adult

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Abstract

Observations: Self-healing cutaneous mucinosis is a localized form of primary cutaneous mucinosis characterized by an early age of onset, the presence of plaques and nodules in a typical distribution, and rapid onset followed by spontaneous resolution of the lesions within a period of weeks to months. Histopathologically cutaneous mucinosis is defined by mucin deposits in the dermis. Here we present a clinically and histopathologically proven case of 25-year-old male patient with a 1-year history of papuler eruption on his elbows and dorsum of the feet.

Introduction

Cutaneous mucinosis includes a heterogeneous group of diseases in which acid glycosaminoglycans (mucin) accumulate either diffusely or locally in the skin or within the hair follicle [1]. Self-healing cutaneous mucinosis is a localized form of primer cutaneous mucinosis and firstly described by *Colomb* et al. [2] in 1973. In 1980 *Bonerandi* et al. [3] reported an additional case and referred to two other reported cases with similar features. The authors proposed that this is a distinct form of cutaneous mucinosis characterized by the following: the young age of onset, the peculiar distribution of the eruption on the face, neck, scalp, abdomen, and thighs; the presence of deep nodules on the face and in periarticular regions; an absence of inflammation, dysglobulinemia, endocrinologic changes, or bone marrow plasmocytosis, and an acute onset followed by spontaneous resolution within a few weeks. Fever, arthralgias, muscle tenderness and weakness may be associated.

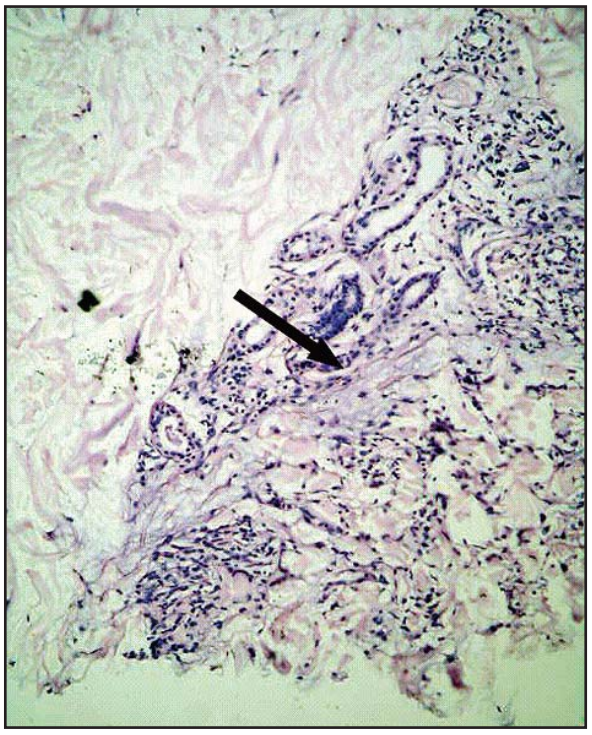

Figure 1. Biopsy specimen shows mucinous material between collagen fibers (hematoxylin-eosin stain)

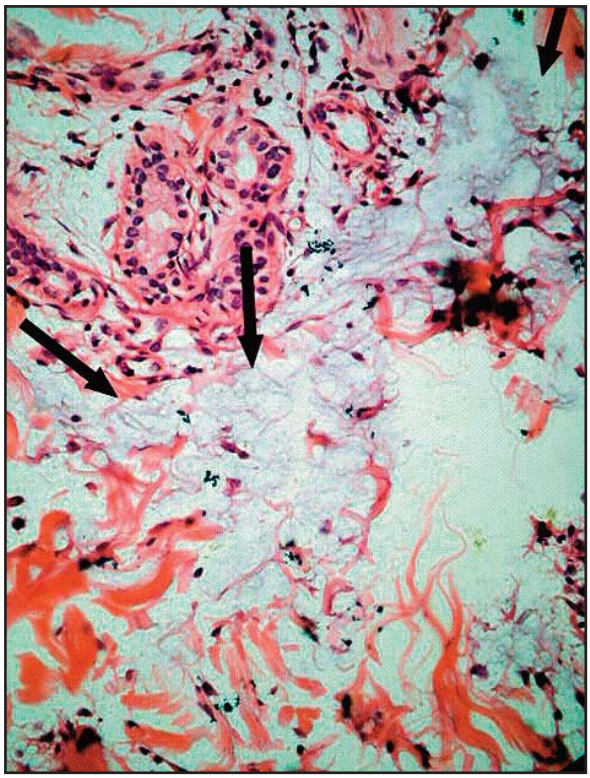

Figure 2. Alcian blue staining highlights presence of an increased dermal mucin (Alcian blue stain pH 2,5)

We report a case of self-healing cutaneous mucinosis in an adult primarily localized around the elbows and dorsal faces of the feet for 1 year.

Case Report

A 25-year-old male patient with small asymptomatic papular eruption around his elbows was admitted to our clinic. Past medical history, family history, and review of systems were unremarkable. He had no known allergies and was currently receiving no medications. Physical examination disclosed multiple flesh-colored, 1 to 2 mm grouped papules around his elbows and dorsum of the feet. The lesions firstly occured on the dorsum of the feet and 6 months later around his elbows.

The results of a complete blood cell count, sedimentation rate, liver and kidney function studies, cholesterol and triglyceride levels, chest and bone x-ray films, electrocardiogram, immunoglobulin studies, antinuclear antibody, rheumatoid factor, serum protein electrophoresis and thyroid function studies were negative or normal.

A biopsy specimen was taken from the elbow. Hematoxylin and Eosin-stained sections showed focal deposits of a mucinous substance among the collagen fibers in the upper dermis (**Figure 1**). Around the mucinous substance fibroblasts and mast cells are increased in number. Alcian blue was positive for mucin (**Figure 2**).

Discussion

The cutaneous mucinoses are a heterogeneous group of disorders in which an abnormal amount of mucin accumulates in the skin either diffusely or focally and may be classified as primary, in which mucin deposition is the main histologic feature resulting in clinically distinctive lesions and secondary, in which mucin simply represents an additional finding [**4**]. Primary dermal mucinosis (lichen myxedematosus - papular mucinosis) includes two clinicopathologic subsets: a generalized papular and sclerodermoid form (scleromyxedema) with a monoclonal gammopathy and systemic manifestations and a localized papular form which does not run a disabling course [**5**].

Self-healing cutaneous mucinosis is one of the 5 subtypes of localized papular form and is used to describe patients without paraproteinemia or thyroid dysfunction whose lesions resolve spontaneously without sequelae in a period ranging from a few weeks to months [**5, 6**].

The cutaneous mucinoses are defined histopathologically by the presence of an increased amount of dermal mucin. The 2 major subtypes of papular mucinosis have distinct histopathologic features. In scleromyxedema, there is a diffuse deposition of mucin in the papillary and midreticular dermis associated with increased collagen deposition and proliferation of fibroblasts [**7, 8, 9**]. However, in localized papular mucinosis mucin deposition may be either focal or diffuse and fibrosis is not marked despite the increasing number of stellate fibroblasts [**8, 10**].

The pathogenesis of the cutaneous mucinoses remains unknown, but is hypothesized to arise from a yet undetermined trigger which causes fibroblast proliferation and mucin production [**11**]. The role of paraproteins and also the other serum factors in the pathogenesis is unclear yet [**12, 13, 14**]. It is considered a reactive response to some chronic antigenic stimulation, such as inflammation or viral infection, or products of tumors combined

with chemotherapeutic agents. No etiologic factor could be found in our patient.

To our knowledge, only six cases of adult self-healing papular mucinosis have been reported in the English-language literature. These patients had heterogeneous clinical presentations that ranged from a single isolated hand lesion, to a patient with a distinct cutaneous eruption with arthralgias, to a patient with polyclonal gammopathy [**15, 16, 17**]. Most of them resolved in a period of 7 months to 1 year. In our case; the distribution of papular lesions around the elbows and dorsum of the feet was usual. Histological examination of the papular lesions in our patient showed focal dermal mucin deposits identical to those described in previous cases. Our patient had no associated symptoms considering systemic involvement. We have not observed spontaneous resolution in our patient, yet. His clinical follow-up has been continuing for 1 year.

Although self-healing papular mucinosis in adults has no clinical predictive markers in contrast to juvenile variant, the disease is distressing and patient's life is extremely affected. Because of its uniformly favorable course, early diagnosis of the disease is very important in predicting prognosis and avoiding unnecessary interventions and anxiety.

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