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Case Presentation

Kathleen Tran, MD, MSc, Kevin P. Boyd, MD, Maria R. Robinson, MD, and Michael Whitlow, MD, PhD

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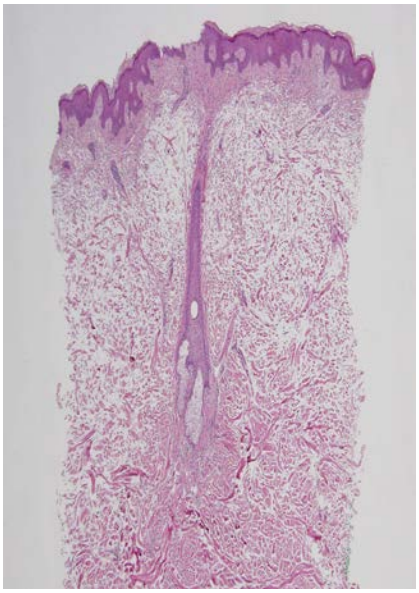
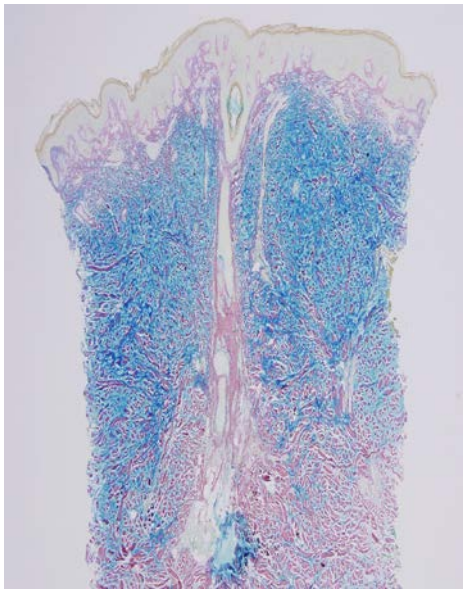
New York University School of Medicine

Abstract

We present a case of scleredema with a leonine facies in a 56-year-old man with a history of poorly controlled diabetes mellitus. The patient initially presented with erythematous, edematous papules and plaques on the face, neck, and upper back.





Case synopsis

A 56-year-old man with a history of long-standing, poorly-controlled diabetes mellitus presented to the Department of Dermatology at the Manhattan Campus of the Veterans Affairs New York Harbor Healthcare System for evaluation of a several-month history of gradually-developing, asymptomatic plaques on the face, neck, and upper back. He denied any illnesses preceding the onset of his skin findings. He denied recent fevers, chills, nausea, vomiting, diarrhea, or weight loss.

Physical Examination: A leonine facies was notable with firm, edematous, erythematous plaques on the forehead and malar aspects of the cheeks. Indurated papules and plaques with a cobblestone appearance also were present on the posterior aspect of the neck and the upper back.

Laboratory Data: A complete blood count and comprehensive metabolic panel were normal, with the exception of a non-fasting blood glucose of 349 mg/dL. A urinalysis showed a glucose level of greater than 1000 mg/dL and no protein. A hemoglobin A1C was 10.7%.

Histopathology: There is prominent separation of collagen fibers throughout the dermis. A colloidal iron stain highlights the presence of abundant mucin.

Diagnosis: Scleredema diabeticorum

Discussion: Scleredema is a rare disorder of connective tissue that was first described by Abraham Buschke in 1902 [1]. It is characterized by gradually developing, painless induration and thickening of the skin usually overlying the face, back, shoulders,

and neck. Morbidity is rare and is usually related to limitation in movement that is secondary to thickening of the skin, although visceral involvement may occur rarely. Three variants of scleredema are classically described [2]. Type 1 most often occurs in middle-aged women and children and is associated with a preceding febrile respiratory illness that is most commonly streptococcal in origin. This variant of scleredema usually resolves after several months to years. Type 2 is not preceded by a febrile illness, is slowly progressive, and is associated with a monoclonal gammopathy [3]. This type tends to persist for years and may carry an increased risk of multiple myeloma [4]. Type 3, which otherwise is known as scleredema diabeticorum, usually occurs in middle-aged men with a history of longstanding diabetes mellitus. This type also tends to persist and there is no clear relationship between prognosis and control of blood glucose levels [5].

Although scleredema may be diagnosed clinically, diagnostic confirmation may be obtained with a tissue biopsy. Histopathologic findings include marked thickening of the reticular dermis, which shows large collagen bundles that are separated by spaces filled with mucin [6]. The number of fibroblasts is not increased and elastic fibers are reduced. A sparse, perivascular, lymphocytic infiltrate is often present. The epidermis usually is uninvolved.

The pathogenesis of scleredema diabeticorum has not been established [5]. Proposed hypotheses include the glycosylation of collagen fibers that leads to altered degradation. Another proposed mechanism implicates hyperglycemia in stimulating fibroblasts and the synthesis of extracellular matrix components.

The differential diagnosis of scleredema includes fibrosing disorders such as scleroderma and scleromyxedema. Clinically, scleredema can be differentiated from scleroderma by the absence of Raynaud's phenomenon, cutaneous telangiectases, and cuticular changes. Scleromyxedema presents with indurated plaques in addition to firm papules that often are arranged linearly. It can be distinguished histopathologically from scleredema based on the proliferation of fibroblasts.

Treatment of scleredema is challenging. In scleredema diabeticorum, it is judicious to advise control of blood glucose as a first step in treatment, although a relationship between glucose control and improvement in scleredema has not been firmly established [5]. In isolated case reports and in case series, phototherapeutic modalities, which include psoralen plus ultraviolet A (PUVA) photochemotherapy [7], ultraviolet A1 phototherapy [8], and narrow-band ultraviolet B phototherapy [9] have shown therapeutic efficacy. In a few patients with disabling scleredema diabeticorum, electron-beam therapy [10] and tamoxifen [11] have been reported to be beneficial.

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