Scleredema Diabeticorum: A Case Report and Review of Literature

Donna Tran, DO,* Navid Nami, DO**

*Dermatology Resident, PGY2, Western University of Health Sciences, College Medical Center, Long Beach, CA
**Program Director, Dermatology Residency, Western University of Health Sciences, College Medical Center, Long Beach, CA

Abstract

Scleredema is a rare connective-tissue disorder characterized by diffuse, non-pitting induration of the skin. Scleredema is divided into three types, distinguished by their associations with infection, monoclonal gammopathy, and diabetes mellitus (scleredema diabeticorum). Pathogenesis is largely unknown. We present a case of a 65-year-old man with scleredema diabeticorum and review the literature and treatment modalities.

Introduction

Scleredema, originally described by Buschke in 1902, is an uncommon, fibromucinous connective-tissue disorder characterized by symmetrical, diffuse induration of the skin due to accumulation of collagen and aminoglycans in the dermis. Hands and feet are characteristically spared. Cardiac and other organ involvement is rare, but in severe cases, patients may present with restrictive lung disease. Classically, three variants of scleredema have been described, one associated with infection (usually streptococcal), one with monoclonal gammopathy, and one with diabetes mellitus. We present a case of a 65-year-old male with insulin-dependent diabetes mellitus who presented with a progressive history of thickening of the skin on his back.

Case Report

A 65-year-old man presented with complaints of painless, progressive hardening of his upper back present for years. He denied any associated symptoms. Past medical history was significant for insulin-dependent diabetes mellitus. Physical examination revealed a symmetrical, erythematous, indurated plaque with indistinct borders involving his posterior neck and upper back (Figure 1). No restriction in range of motion of the shoulders and neck was noted.

A histopathologic examination of a punch-biopsy specimen from the skin on his back revealed an intact epidermis with no interface changes. A scant superficial perivascular lymphocytic infiltrate was present (Figure 2). A colloidal-iron stain revealed an increase in dermal mucin (Figure 3). These features supported the diagnosis of scleredema diabeticorum. Our patient was asymptomatic and described no restriction in movement. To our knowledge, his diabetes mellitus was controlled with subcutaneous insulin injections and diet. He opted for close follow-up.

Figure 1. Thickening and erythematous induration of the posterior neck and upper back.

Figure 2. Scant superficial perivascular and lymphocytic infiltrate with sparing of the epidermis.

Figure 3. Colloidal-iron positive with increase in dermal mucin.
Discussion
Scleredema is a sclerotic skin disease that is divided into three types. Type 1 typically affects middle-aged women and is often preceded by an acute febrile disease, most commonly of streptococcal origin. A sudden-onset hardening of the skin of the cervicofacial region with spontaneous resolution in a period of months to years is seen. Type 2 scleredema appears in individuals without diabetes or infection. This type occurs in women and has an insidious and prolonged course. Monoclonal gammopathy, most commonly IgG, with progression to multiple myeloma may be seen. Type 3 occurs primarily in obese, middle-aged men with a long history of diabetes mellitus, as seen in the patient described here. This third type may be referred to as “scleredema diabeticorum.”

Onset of scleredema diabeticorum is insidious, and a progressive, non-pitting induration of the skin involving the posterior neck, back, and shoulders, and less commonly arms, is often seen. Erythema and a peau d’orange appearance may be present, with indistinct demarcation between normal and abnormal skin. Individuals often have a long history of poor metabolic control as well as diabetic complications; however, cases of scleredema diabeticorum in diabetics with controlled glucose have been reported.

The pathogenesis of scleredema diabeticorum is uncertain. Various etiologies have been proposed, including irreversible glycosylation of collagen and resistance to degradation by collagenase, leading to collagen accumulation. Other hypotheses suggest microvascular damage and hypoxia stimulating mucin and collagen synthesis. Diagnosis of scleredema is based on clinical findings of non-pitting induration or stiffness of the skin, which may be of sudden or insidious onset. Histopathologic confirmation is not required. On skin biopsy, the epidermis is usually spared. Thickened collagen bundles within the reticular dermis separated by mucin-containing fenestrations is characteristic. No increase in number of fibroblasts is seen.

Scleredema may be confused with scleroderma, but absence of a history of Raynaud’s phenomenon, calcification, or telangectasia points to scleredema. Scleromyxedema, a generalized primary cutaneous mucinosis that also may have associated monoclonal gammopathy, can be differentiated from scleredema by its diffuse, waxy papules, often in a linear array, in addition to the proliferation of fibroblasts seen histologically.

Scleredema diabeticorum is a relatively underestimated and unrecognized complication of diabetes. The exact prevalence is not known. Cole et al. observed a prevalence of 2.5% in a population of 424 diabetic patients. Satter et al. found a higher prevalence of 14% in a population of 100 hospital-based patients with diabetes mellitus. In most cases, glucose control is the first recommended step in management. In one series of four patients with type 1 diabetes, implantable insulin pump therapy led to improvement of glucose control as well as clinical improvement of the skin in terms of redness, swelling, and induration. Unlike streptococcal-associated scleredema, which often carries a better prognosis and resolution of skin disease after a period of months to years, scleredema diabeticorum often follows an indolent course, and lesions may persist despite good metabolic control. For most patients, diabetes-associated scleredema is a relatively mild disorder, although rarely death may occur when internal organs are involved.

Multiple treatment modalities have been reported as case reports or small series with variable success, including corticosteroids, cyclosporine, methotrexate, potassium para-aminobenzoate, and electron-beam radiation therapy. In a few reported cases, tamoxifen has been effective in improving skin thickening and joint mobility within months of starting treatment. More recently, ultraviolet A-1 phototherapy and psoralen with ultraviolet A (PUVA) has been effective in patients with scleredema diabeticorum. The mechanism of benefit of PUVA on scleredema is suggested to be an increase in collagenase synthesis by fibroblasts and the inhibition of de novo type 1 collagen synthesis. In one case, combining local PUVA and colchicine was reported to have a synergistic benefit in the treatment of scleredema diabeticorum in a 53-year-old woman. At the end of treatment, the patient was able to lie in a supine position without pain or paresthesia, and her epidermal thickness, measured with a 17-MHz probe ultrasound, was decreased by 34.29%, with dermal thickness decreased by 7.03%.

Conclusion
The frequency of scleredema diabeticorum is underestimated. In most cases, scleredema diabeticorum is a self-limited, benign condition. However, severe cases with rapid progression and disabling consequences may occur. Improvement of diabetes control is first-line treatment. Our patient was asymptomatic and opted for close follow-up. His diabetes was controlled with diet management, exercise, and insulin subcutaneous injections.

References

Correspondence: Donna Tran, DO; DonnaDTran@gmail.com