Scleredema Diabeticorum

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Abstract

Scleredema diabeticorum is one of the skin disorders associated with diabetes mellitus, characterized by thickening of the deep layers of the dermis, with excessive mucin and collagen deposition, clinically evidenced in hardening of the skin, especially in the upper half of the body. We describe the clinical case of an adult male diabetic who was seen for an indurated cervical lesion which was subsequently diagnosed histopathologically as *scleredema diabeticorum*. The interest in this case lies in the low prevalence of the condition and its association with poor metabolic control of diabetes. **(Acta Med Colomb 2021; 46. DOI: https://doi.org/10.36104/amc.2021.1888).**

Key words: *scleredema, diabetes mellitus, diabetes complications, microangiopathy, mucin*.

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Introduction

Scleredema diabeticorum (SD) is a rare connective tissue disorder which most often affects patients with long-term diabetes mellitus and poor glycemic control. It is also associated with other entities such as paraproteinemia, rheumatological diseases and streptococcal infections. It is characterized by symmetrical, diffuse, non-pitting induration of the skin, with occasional erythema and an orange-peel appearance mainly on the back of the neck, shoulders and upper back. Below we present a diabetes-related case of scleredema confirmed by histopathology.

Clinical case

A 68-year-old man with a 37-year history of type 2 diabetes mellitus and a need for insulin, consulted due to the appearance of a pustular lesion on the back of his neck. Limited folliculitis was found, with notable widespread induration on the posterior cervical region and upper back (Figure 1). The physical exam was remarkable for *acanthosis nigricans* on the neck and axillae, cutaneous stigmata of diabetic vasculopathy and neuropathy, decreased visual acuity and a fundus exam showing grade III diabetic retinopathy. On paraclinical exams, his glycosylated hemoglobin was 13%, with average glucometer readings over 270 mg/dL (preprandial) and 340 mg/dL (postprandial). He received numerous pharmacological treatments for his skin problem, including antimicrobial treatment, without improvement. Therefore, a biopsy was ordered, considering the possibility of *scleredema diabeticorum*. The skin pathology study showed increased thickness of the dermis, abundant thickened collagen fibers and the presence of mucin on toluidine blue staining (Figure 2), thus confirming the diagnosis of sceleredema associated with diabetes mellitus.

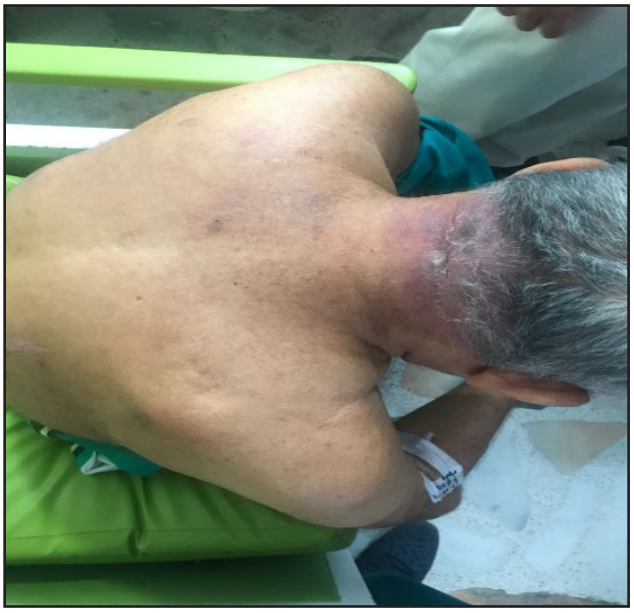

Figure 1. *Area of cervical scleredema*..

Discussion

Scleredema is a rare skin condition that usually occurs in association with diabetes mellitus, infections or monoclonal gammopathy. It is characterized by the development of a

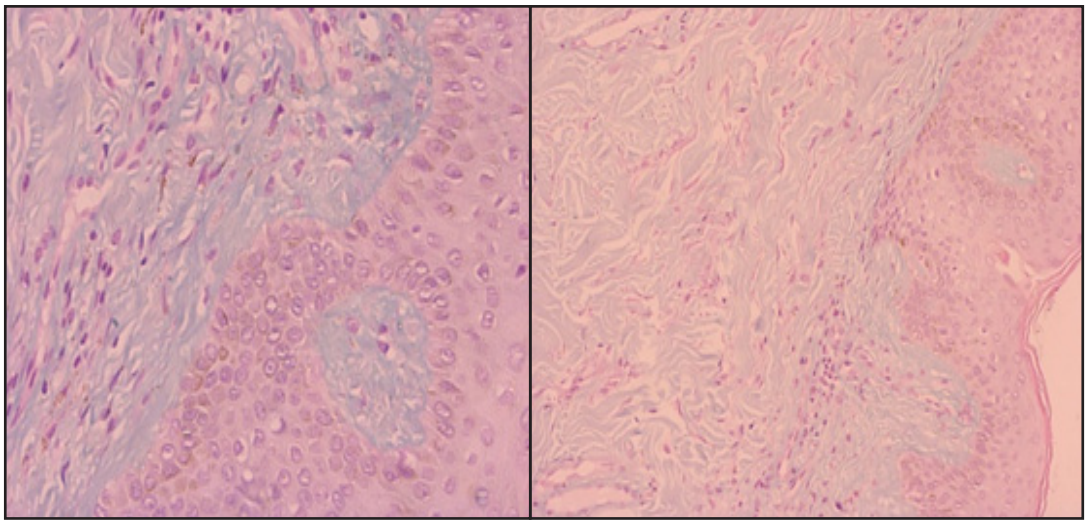

Figure 2. *Toluidine blue staining showing mucin deposits, collagen fiber thickening and dermal thickening.*

diffuse, symmetrical area of induration mainly affecting the upper body (head, shoulders, neck and upper limbs) (1, 2). Its clinical course is variable and is directly related to the situations which cause it. It may be acute or subacute (two to six weeks), occur with infections (usually streptococcal), and generally occurs in people with poorly controlled diabetes mellitus (2, 3).

Scleredema associated with diabetes mellitus has an unknown prevalence, with substantive mentions in reports or case series, reiterating that it mainly affects adults, and predominantly males (2, 3) with obesity, a hallmark of these patients (3). Its cause is unknown; however, it is related to long-term illness, morbid obesity and treatment with high doses of insulin. It is also associated with poor glycemic control and acanthosis nigricans (3, 4). Hypotheses have been made about its etiopathogenesis, with prolonged hyperglycemic states leading to the irreversible formation of glycosylated collagen complexes, and abnormal enzymatic activity of collagenases resulting in excess accumulation of collagen and mucin in the reticular dermis (4). Profibrotic abnormalities secondary to hypoxia and microvascular damage have also been suggested (5).

The diagnosis is clinical, but must be confirmed through skin pathology tests showing a thickened dermis (up to three times thicker), along with a reduction in eccrine glands; a normal number and appearance of fibroblasts; thickened collagen fibers separated by an accumulation of mucopolysaccharides, especially hyaluronic acid; and amorphous material, which are visible with Alcian blue and toluidine blue staining (6). The clinical course of *scleredema diabeticorum* is variable and, although in most patients it is persistent, cases of clinical improvement have been seen (basically, decreased lesion size) once adequate diabetic control is achieved (7).

The differential diagnosis of this disease should mainly include scleroderma and scleromyxedema, the first characterized by Raynaud's phenomenon in the extremities and the presence of telangiectasias, atrophy and calcifications (8); and the second being a type of mucinosis characterized by lichenoid papules and diffuse induration of the skin. The diagnosis should also be differentiated from other diseases like trichinosis, dermatomyositis, primary systemic amyloidosis and eosinophilic fasciitis (9).

A specific effective treatment for *scleredema diabeticorum* has not yet been established, but weight loss and strict diabetic control are recommended which, although they will not solve the problem, may help halt or partially improve the progression of the skin lesion (12). The various proposed treatments, with inconsistent results, include immunosuppressants (like cyclosporine and methotrexate) in scleredema associated with myeloma, and photochemotherapy with psoralen and ultraviolet A radiation (PUVA) (10, 11).

Scleredema should be considered a marker of poor glucose control and the progression of microvascular complications of the disease, being an important clinical sign in the day-to-day assessment of patients with diabetes mellitus, especially those with insulin resistance.

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