Scleredema diabeticorum – A case report

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ABSTRACT

Scleredema diabeticorum is an uncommon skin disorder which is characterized by stiffness and hardening of the subcutaneous tissues located on the upper back and posterior neck. Patients with this skin disease typically experience pain and stiffness on the neck. Scleredema associated with diabetes mellitus may often go unrecognized. We report a case of a 55-year-old female patient with diabetes mellitus type 1, who presented with skin tightness on her upper back and decreased range of motion. Scleredema diabeticorum is rare and its treatment is difficult and tricky, as no specific regimen has been instituted to treat this skin disease.

Keywords: Diabetes mellitus, hyperglycemia, scleredema diabeticorum

Introduction

The diabetes-associated disease, named scleredema diabeticorum (SD), may develop both in type 1 and type 2 diabetes mellitus patients.¹ It is related with the prolonged exposure to hyperglycemia, obesity, and bad metabolic control.¹ SD consists 20% of scleredema cases.²

Herein, we report a case of SD in a 55-year-old female with long-lasting diabetes mellitus. The purpose of this case report is to trigger the awareness of such an entity in a chronic disease that mainly be managed by general practitioners and illustrate the challenges of treating SD, due to the generally unsuccessful results of currently available treatments.

Case History

A 55-year-old woman presented to our department complaining of skin tightness on her upper back and decreased range of motion. She has had a long history (35 years’ duration) of diabetes mellitus type 1. When she first visited us last year, she had controlled diabetes and good glycemic control (Hb1Ac = 6.5), but diabetes was poorly controlled before her current visit. Recently, she had been diagnosed with hypertension. There were no reported connective tissue disease symptoms, and her endocrinologist had excluded diabetic complications, such as retinopathy, nephropathy, and neuropathy during her early check-up. In her medical history, a respiratory infection that was treated with antibiotics was reported. Her current medications included detemir insulin, premeal aspart insulin, lisinopril, and hydrochlorothiazide. The patient and the institution have granted written permission to the authors to publish this case report.

Physical examination revealed a symmetric, painless induration of the skin located on the upper back and neck [Figures 1 and 2]. On the evaluation of the neck and upper extremities there was...

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How to cite this article: Kyriakou A, Zagalioti SC, Lazaridou E, Patsatsi A. Scleredema diabeticorum – A case report. J Family Med Prim Care 2021;10:1037-9.
a significant decreased range of motion. Heart and lungs auscultation were unremarkable.

Her laboratory evaluation included complete blood count, blood glucose, urinalysis, liver and renal tests, thyroid function test, protein electrophoresis, antistreptolysin O titer, sedimentation rate, and C-reactive protein level that were in normal ranges or negative.

Skin biopsy was obtained from the lesion. Biopsy specimen showed thickening of the dermis with thick, dermal collagen fibers separated by mucin deposits. This histological finding supported our clinical findings to set the diagnosis of SD.

Several treatment modalities have been tried and tested. At first, therapy with cyclosporine (150 mg p.o.s., twice daily) for 3 months appeared unsuccessful, so low-dose methotrexate (15 mg p.o.s., weekly) was the next therapeutic choice. Neither clinical response nor improvement was defined after treatment for 5 months. The patient then received systemic glycocorticosteroids (20 mg prednisolone p.o.s. daily) for a month, with partial improvement. However, due to diabetes mellitus, systemic glycocorticosteroids had to be discontinued. Radiotherapy and especially electron-beam radiation appeared promising and showed partial improvement at last. Overall, the aforementioned therapies used in this case have been proven ineffective. Finally, symptoms showed minimum improvement under treatment with colchicine and doxycycline, but without the desirable outcome.

**Discussion**

Cohn et al. in 1970 were the first that recognized and published scleredema diabeticorum as a syndrome. Scleredema associated with diabetes mellitus is a rare manifestation of diabetes and often stays unrecognized. It typically presents as a diffuse symmetrical tightness of the skin over the neck, shoulders, and rarely affected the upper extremities, causing limitation of the range of motion. It mainly occurs in obese middle-aged men with longstanding insulin-treated diabetes with bad glycemic and metabolic control. It is observed in both types 1 and 2 diabetes mellitus patients and there is no racial predilection.

The exact pathophysiology remains unknown. It seems that the chronic and bad glycemic control plays the main role. It has been hypothesized that a nonenzymatic glycosylation process damages the collagen fibers. In dermis, more collagen and mucin (hyaluronic acid, an acid mucopolysaccharide) is produced by fibroblasts, as there is irreversible glycosylation of collagen and alterations in collagenase activity. Another pathogenesis approach reports that the microvascular damage and hypoxia have the primary role in the excessive genesis of collagen and mucin.

SD can be suspected by the clinical characteristics, but biopsy establishes the definitive diagnosis. Histologically, the skin specimen shows thickening of the dermis with swollen collagen fibers in bundles and deposits of mucin, which can be stained by alcian blue dye.

Because of the infrequent occurrence of this manifestation, there is no standard therapeutic protocol. Although many therapeutic options have been published, the course of SD is usually unpredictable and the disease itself is refractory. Even though, the existence of controlled hyperglycemia over multiple years does not seem to improve the skin disorder, this still remains the first therapeutic step. Treatment options that have been reported to manage SD include topical and systemic treatments, and physical modalities. Topical corticosteroids, local PUVA, or localized electron beam therapy have been used. But the vast majority of reported patients have undergone systemic therapy with systemic steroids, methotrexate, cyclosporine, IV Ig, and colchicine. Physical therapy is recommended to increase range of motion.

Although this is a chronic disease with doubtful responses to many treatments, it is not a life-threatening condition. New
treatments are undeniably needed based on the pathogenesis of SD. Maintaining a strictly glycemic control remains a key factor in preventing complications among primary care patients with any type of diabetes mellitus. Awareness on SD should be raised within physicians and alert them to thoroughly investigate for underlying hyperglycemia or paraproteinemia.

SD is a rare skin manifestation of diabetes. It is a profound concern today, as it is a debilitating chronic disease with unpredictable therapeutic results. A comprehensive composition of publishing data is needed to develop a therapeutic protocol. Finally, as in its pathogenesis bad glycemic control contributes to its genesis, the key for primary prevention of this disease is to manage strict glycemic control.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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