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Case Report

Pigmented scleredema: An unusual presentation

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Abstract Scleredema adultorum (SA) of Buschke is a rare disorder of idiopathic etiology. Clinically differentiated by diffuse, symmetric, and non-pitting induration of the skin which typically affects the neck, shoulders, trunk, arms, face, and, in rare cases, the buttocks and thighs. Here, we describe a case with scleredema diabeticorum with an uncommonly pigmented appearance.

Key words
Pigmented scleredema; Unusual presentation; Scleredema diabeticorum.

Introduction

Scleredema diabeticorum is a rare connective tissue disorder initially identified by Buschke in the year 1900.[1] In order to differentiate it from scleredema neonatorum, Buschke suggested the term "scleredema adultorum".[2] Clinically, it is distinguished by diffuse, symmetrical, and non-pitting induration of the skin, which typically affects the neck, shoulders, trunk, face, arms, and very infrequently, the buttocks and thighs.[3] There are three known types of scleredema. Type 1 develops after an acute febrile infection, caused by the streptococcus bacteria. Typically, this kind of scleredema disappears over several months. Subjects without diabetes develop type 2 scleredema. Patients with type 1 or type 2 diabetes mellitus develop type 3 scleredema.[1] Here we report a case of scleredema diabeticorum with unusual pigmented presentation, which is a rare occurrence.

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Case report

An 82-year-old male came up with complaints of dark lesions on the trunk and extremities for the past 6 months associated with itching. There was no H/o pain or burning sensation. He has been a known case of diabetes mellitus for the past 10 years. No H/o symmetrical joint pain was present. O/E patient had multiple hyperpigmented plaque present on his back and arms. Few showed induration **(Figure 1a)**. Apart from a raised blood sugar level, his other hematological and biochemical parameters such as a complete blood count, liver, ESR, and renal function tests, thyroid and immunoglobulin profile were within normal limits. Hematoxylin–eosin study of skin biopsy specimen showed superficial dermis with extensive mucin deposits producing separation of collagen fibers **(Figure 1b)**. Hematoxylin-eosin stained section also showed deposition of collagen bundles in the upper part of the dermis **(Figure 1c)**.

SA of Buschke is a mucinous connective tissue disorder that is classified under scleroderma-like diseases.[3] About pathogenesis of scleredema the most reliable theory is that persistent hyperglycemia accumulates in the dermis and causes scleredema by gradually damaging the collagen fibers in connective tissue through a nonenzymatic glycosylation process.[1]

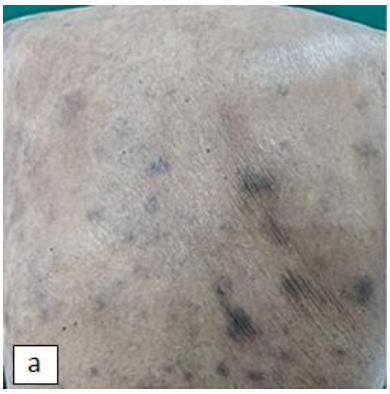

Figure 1a Multiple hyperpigmented plaque present over back, arms.

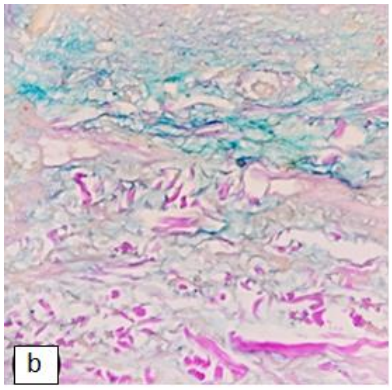

Figure 1b Superficial dermis with extensive mucin deposits.

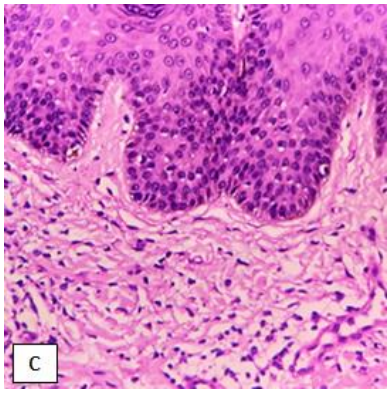

Figure 1c Hematoxylin-eosin stained section (40x view) shows deposition of collagen bundles in the upper part of the dermis.

Another theory links hyperglycemia to the stimulation of fibroblasts and the production of extracellular matrix elements.[3] The link between scleredema and diabetes mellitus is not well understood but it is a well-established fact that scleredema patients with diabetes mellitus will have difficulty in having their diabetes as well as diabetic complications controlled.

Scleredema gives rise to non-pitting hardening of the skin around the shoulders, neck and trunk. Typically, thickening starts in the neck and may spread to the shoulders, upper part of the trunk, and occasionally the face,[4] but in this case, patient had multiple hyper pigmented plaque present on the back, arms. Scleredema is typically suspected based on clinical findings, but a skin biopsy is required to make the actual diagnosis. The biopsy's microscopic characteristics include thickening of the dermis due to increased collagen bundles in deep reticular dermis with clear spaces between them, filled with mucin.[1] In our case, histopathology showed superficial dermis with extensive mucin deposits producing separation of collagen fibers and deep dermis shows thickened collagen bundles. Having blood glucose levels within the normal limits has been the first step in treating scleredema diabeticorum. Differential diagnoses considered were Stiff skin syndrome and morphea. Features suggestive of stiff skin syndrome such as symmetrical joint pain and limited joint mobility were absent. Although morphea can manifest clinically and histologically with indurated plaques and thicker collagen bundles throughout the dermis, mucin is not the primary characteristic.[5] Moreover, dermal-subcutaneous junction frequently exhibits some degree of inflammatory infiltrate. The high level of mucin in our case aroused concerns about the probability of scleredema. Having blood glucose levels within normal limits has been the first step in treating scleredema diabeticorum. Some scleredema patients have also undergone immunosuppressive treatment trials with Methotrexate, corticosteroids, cyclosporine and have given inconsistent results. The glucose level of our patient has been kept under control, and routine blood investigations have been done in view of starting immunosuppressant. However, the patient never returned for follow up.

Conclusion

Scleredema adultorum is an uncommon disorder which can appear in diabetic patients who have poor glycemic control. Scleredema adultorum with hyper pigmented plaque is a very rare occurrence.

Our case supports the hypotheses that diabetic patients presenting with indurated hyperpigmented plaque should raise the suspicion of scleredema adultorum.

Declaration of patient consent The author certify that they have obtained all appropriate patient consent.

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Conflict of interest The author declared no conflict of interest.

Author's contribution

YS: Identification, diagnosis & management of the case, manuscript writing, critical review, has given final approval of the version to be published.

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