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Idiopathic scleredema

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Abstract

Scleredema, which also is known as scleredema adultorum of Buschke, is an uncommon sclerodermiform condition that is characterized by progressive thickening and hardening of the skin due to excessive dermal mucin and collagen deposition. The clinical course is variable, and progression of disease may lead to functional impairment with limitations in mobility. The etiology and pathogenesis are unknown although several well-known associations include streptococcal infection; diabetes mellitus, particularly with metabolic syndrome; and monoclonal gammopathy. We present a case of scleredema in a 52-year-old man with no identifiable associated condition, who experienced improvement with UVB phototherapy.

Case Presentation

PATIENT: 52-year-old-man

DURATION: Two months

DISTRIBUTION: Upper back

HISTORY: A 52-year-old man presented to the Dermatology Clinic at Bellevue Hospital Center for the evaluation of a large mass on his upper back. He first noted the bump on his upper back about two months ago, and it had been enlarging slowly. The mass was asymptomatic; there was no itching or pain. He denied any medical problems aside from pre-diabetes with a recent HgA1c of 5.9 that was being controlled with diet alone. He was otherwise well and did not take any medications. He denied a history of infection prior to development of the mass. Review of systems was negative for sore throat, cough, rhinorrhea, nasal congestion, fatigue, or shortness of breath. There was no history of prolonged glucocorticoid use, travel, or family history of a similar dermatologic condition.

PHYSICAL EXAMINATION: On the upper back, there was a large, non-tender, non-mobile, subcutaneous, firm mass that measured about 23-cm x 9-cm (**Figure 1**). There was no erythema, scale, or other surface changes. Moon facies, striae, and sclerodactyly were not present.

LABORATORY DATA: A complete blood count, basic metabolic panel, thyroid stimulating hormone, and fasting glucose were normal. Total cholesterol level was 212 mg/dL and low-density lipoprotein level was 142 mg/dL. Immunofixation electrophoresis and serum protein electrophoresis were normal. Anti-streptolysin O antibody titers were negative.



Figure 1. On the upper back, there was a large, non-tender, non-mobile, subcutaneous, firm mass.

HISTOPATHOLOGY: Within the deep reticular dermis there are thickened collagen bundles with increased connective tissue mucin as highlighted by a colloidal-iron stain (**Figure 2**).

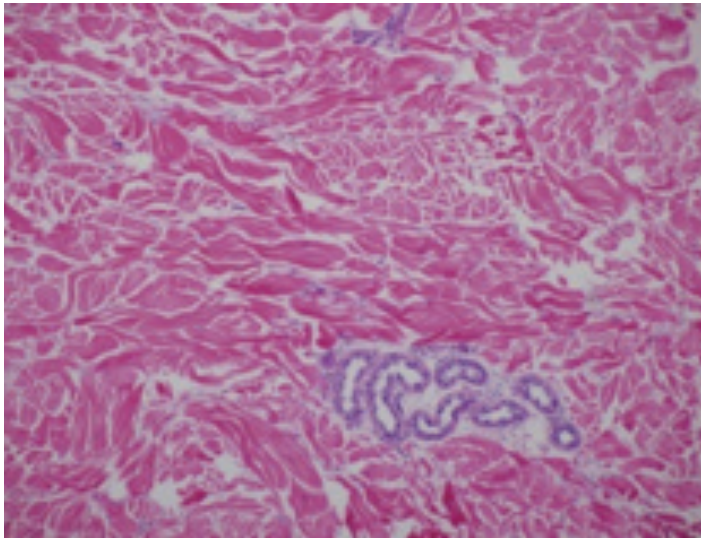


Figure 2. Thickened collagen bundles with increased connective tissue mucin in the deep reticular dermis.

DIAGNOSIS: Scleredema

Discussion

Scleredema, which also is known as scleredema adultorum of Buschke, is an uncommon sclerodermiform condition that is characterized by excessive dermal mucin and collagen deposition. Clinically, it presents as symmetric thickening and non-pitting induration of skin, sometimes with associated erythema or a peau-d'orange appearance. This hardening of the skin may lead to pronounced movement restriction. The upper body, which includes the neck, upper trunk, and upper limbs, is most commonly affected while the hands and feet usually are spared, which is a feature that helps distinguish the condition from scleroderma. Rarely, systemic involvement affects the eyes, tongue, esophagus, and myocardium, which may lead to ptosis, immobility of the tongue, dysphagia, arrhythmias, and pericardial or pleural effusions [1, 2].

Histopathologic analysis shows a swollen-appearing dermis with thickened collagen bundles that are separated by prominent mucopolysaccharide deposits but no increase in fibroblasts. The epidermis may be normal or slightly thin, and

there is a decrease in elastic fibers. Unlike with scleromyxedema, there is no increase in the fibroblastic population, and, unlike with scleroderma, the thick collagen bundles that surround the adnexae do not compress or destroy them [1]. The pathogenesis remains unknown, but studies have shown increased protein production, glucosamine incorporation, and collagen synthesis by scleredema fibroblasts from lesional skin as well as stimulation of collagen synthesis in fibroblast cultures by serum from affected patients [3, 4].

Traditionally, scleredema has been categorized into three distinct groups [5]. The first type of scleredema has an acute onset and is associated with a preceding acute respiratory tract infection, which is usually streptococcal. It occurs more commonly in children and tends to resolve spontaneously over the course of several months. The second type is associated with a monoclonal gammopathy, presents more insidiously, and tends to have a chronic progressive course. The third type, which also is known as scleredema diabeticorum, follows a similarly chronic course and is usually found in obese patients with diabetes mellitus. Since its initial description, scleredema also has been associated with a range of other systemic conditions in anecdotal reports, which include insulinoma, secondary hyperparathyroidism, rheumatoid arthritis, Sjögren's syndrome, human immunodeficiency virus infection, and carcinoma of the gallbladder [1, 5].

Owing to its relative rarity, there is limited information regarding clinical course, prognosis, and treatment. A recent multicenter, retrospective study, which is the largest case series published in English literature to date, investigated the characteristics, comorbidities, course, and therapy in 44 patients with scleredema. The study confirmed the classic scleredema patient demographic. Affected adults were primarily in their fifties and sixties, the most common association was diabetes mellitus in 68% of patients, and there was a male predominance among diabetic patients with a ratio of 2.3. Of the diabetic patients, 22.6% had concomitant thyroid disease and 20% obesity. The second most common association was arterial hypertension in 34%, which was followed by monoclonal gammopathy in 11.4%.

There were no cases of post-infectious scleredema although this may be attributable to the lack of pediatric cases in this cohort. In 11.4% of cases, no associated condition could be found, and these were considered idiopathic. In prior published case series, the proportion of patients with diabetes mellitus has ranged from 36.3 to 79.5% while the prevalence of monoclonal gammopathy has ranged from six to 46%. In terms of clinical course and therapeutic response, these were variable, and no relationship was found between response to therapy and associated diseases [2].

There is no standard treatment for scleredema, and results are often variable and unpredictable. As the disease may be asymptomatic and indolent in some patients, treatment may be reserved for those with functional impairment, internal involvement, or underlying systemic conditions. Success has been reported with ultraviolet A (UVA) phototherapy, and this may be considered as initial treatment, owing to its relatively safer side effect profile as compared to systemic medications, such as systemic glucocorticoids and immunosuppressive agents. Physical therapy and, recently, ultrasonic massage also have been reported to be helpful in patients with disease affecting mobility. For those with an associated primary condition, treatment of the underlying disease should always be considered as there have been reports of improvement and even resolution of scleredema after treatment of the associated condition. This has included strict glycemic control in scleredema diabetorum, intravenous immunoglobulin treatment of paraproteinemia-associated scleredema and post-streptococcal scleredema, and bortezomib-based treatment of multiple myeloma associated scleredema [2, 6-9].

Although this patient may be at risk for metabolic syndrome in the future owing to his borderline HgA1c level, his fasting glucose was normal, and his work-up for infection, thyroid disease, and monoclonal gammopathy were negative. Thus, he was determined to have idiopathic scleredema and was initiated on UVB phototherapy with mild improvement noted after five months of therapy.

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