A Case of Idiopathic Scleredema Adultorum of Buschke

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Abstract
Scleredema adultorum of Buschke is a rare, benign disease that results in symmetrical induration and thickening of the skin due to increased collagen and mucin deposition in the dermis. We report the unique case of a patient with no underlying comorbidities who presented with asymptomatic scleredema. Given that this skin condition is difficult to treat and has no standard treatment, this case brings about an important discussion about diagnostic modalities and appropriate management of scleredema adultorum of Buschke.

Introduction
Scleredema adultorum of Buschke, or scleredema, is a connective tissue disorder that results in progressive, symmetrical induration and hardening of the skin, usually of the face, neck, shoulders, upper trunk, and extremities. Histology reveals thick, dermal collagen bundles separated by mucin. It has a female predominance, with a female-to-male ratio of 2:1; however, diabetes-associated scleredema is more common in middle-aged, obese men, with a female-to-male ratio of 1:3. Scleredema can occur in people of all ages but tends to involve adults in their 50s and 60s.

Case Report
A 71-year-old male with a past medical history of coronary artery disease, hypertension, hyperlipidemia, gastroesophageal reflux disease, sleep apnea, chronic back pain, and diverticulosis presented to our dermatology clinic for a routine full-body exam. On examination, his back was mildly indurated with diffuse, flesh-colored atrophic scars with a peau d’orange appearance (Figure 1). The patient stated that in the past, he had always been told they were acne scars, although he did not recall a history of acne. He said the scars were present for approximately 30 years. He denied any symptoms, including sensations of tightness, pain and itch, and said he’d undergone no previous treatments. He denied a history of paraproteinemia, recent streptococcal infections or illness, and diabetes mellitus.

A punch biopsy was performed, revealing mucin deposition between thickened collagen bundles consistent with scleredema adultorum of Buschke (Figure 2). A colloidal iron stain was used to highlight the dermal mucin deposition (Figures 3, 4). A SPEP with immunofixation was performed and showed no monoclonal gammapathy. Free kappa light chains were mildly elevated at 27.09 (normal range 3.30-19.40); however, his kappa/lambda ratio was within normal limits. His recent HgbA1c was 5.7.

Discussion
Scleredema is a rare, benign disease first described as scleredema adultorum of Buschke in 1752. This disease mostly involves the skin and is characterized by acute skin induration with mild sclerosis. Lesions are diffuse and symmetrical and consist of non-pitting edema with ill-defined thickening of the skin. This thickening is secondary to increased collagen and glycosaminoglycans. At times, there is a peau d’orange appearance to the skin. A more severe, progressive form of scleredema with rapid onset, referred to as scleredema fulminant, is life threatening and requires urgent intervention.

Skin manifestations usually begin on the neck and can spread to the shoulders and upper trunk. The disease can also spread to the face, abdomen, and legs, but the hands and feet are spared. Associated symptoms include erythema, pruritus, urticaria, dermatographism, pain, and movement restrictions. Rarer complications include restrictive lung disease, cardiac dysfunction, dysarthria, dysphagia, poor wound healing, and skin infections.

The exact pathophysiology of scleredema is unknown; however, it is believed to involve an increased expression of type I collagen-producing fibroblasts in the skin. Some studies show that factors like infections, inflammation, glucose levels, drugs, genetic mutations, and toxins can stimulate the fibroblasts to overproduce mucin or collagen in the skin. Specific mediators include eicosanoids, growth factors, cytokines, and immunoglobulin paraproteins. Another hypothesis involves the inhibition of collagen degradation by excessive, non-enzymatic glycosylation. The combination of advanced glycation end products and high blood glucose levels leads to collagen cross-linking.

Studies have shown there are three types of scleredema. The classic form, or type I, is self-limiting, lasts anywhere from several months to two years, and is associated with febrile disease. The most common infection associated with type I is streptococcal infection. Other infections include influenza, measles, mumps, chickenpox, and cytomegalovirus. Type II, which is often more chronic, is associated with hypergammaglobulinemia and progression to multiple myeloma. Type II may also be associated with other malignancies, such as insulinoma and carcinoma of the gallbladder.
has an insidious onset without preceding febrile illness.3 Type III scleredema, also known as scleredema diabetorum, is associated with type I or type II diabetes mellitus. There is a higher risk of scleredema in patients with poorly controlled insulin-dependent type II diabetes, dyslipidemia, and thyroid disorders (especially hypothyroidism).3 Underlying diseases rarely associated with scleredema include hyperparathyroidism, connective tissue disease, carcinoid syndrome, pituitary-adrenocortical neoplasms, and human immunodeficiency virus infection.4,5

Diagnosis of scleredema is based primarily on clinical presentation, but a punch biopsy is needed to make a definitive diagnosis. It will show a normal or slightly thinned epidermis.7 The dermis will contain a decreased number of elastic fibers and thick collagen bundles separated by mucopolysaccharide deposits. Fibroblasts will be normal in number and morphology.1 In some cases, the superficial dermis will have a mild, perivascular, chronic inflammatory infiltrate with an increased number of mast cells.7 Adipose tissue may be replaced with collagen bundles in the subcutaneous layer.1 Thickened collagen surrounds the adnexa but shows no compression or destruction, distinguishing it from scleroderma.1 Special staining such as hematoxylin-eosin, colloidal iron, and Alcian blue can be used to visualize these features under magnification.4

Treatment is not necessary for type I scleredema, which is self-limiting and tends to resolve within two years. For types II and III, treatment is needed to prevent worsening of symptoms or systemic involvement. Scleredema is a difficult disease to treat, with no universally accepted treatments. Treating the underlying disease with strict glucose control in scleredema diabetorum is vital. Much research has been done on the effectiveness of different treatments. Out of all the treatment trials, UVA-1 and psoralen plus ultraviolet A (PUVA) light phototherapy had the most successful results. It is believed that phototherapy is effective due to the upregulation of collagenase by fibroblasts that results in degradation of type I and type III collagen fibers as well as collagen mRNA.6,9 The addition of colchicine to PUVA can be beneficial in that colchicine can suppress TGF-beta-induced upregulation of type I collagen mRNA in human dermal fibroblasts.7 Physical therapy can prevent the limitations in range of motion or respiratory issues experienced by some patients.3 Allopurinol has been shown to have antioxidant effects that help in the degradation of thick collagen bundles; it has also been shown to reduce vascular oxidative stress.8 In one case, frequency-modulated electromagnetic neural stimulation (FREMS) was shown to enhance the microvascular blood flow and smooth muscle vasomotor activity in the treatment of scleredema diabetorum.6 Some trials have shown success with systemic corticosteroids, cyclosporine, localized electron beam therapy, IVIG, and high-dose penicillin.3,4 However, these should be reserved for patients with relatively persistent, debilitating disease that fails phototherapy or for patients with multiple myeloma.3

**Conclusion**

Our case is unique not only because the patient was asymptomatic, but also because he had no underlying illnesses associated with his scleredema. Therefore, we are unable to classify it as type I, II, or III. However, studies show that abnormal protein levels may be delayed, with a median period of 2.5 years to 6.9 years between initial diagnosis of scleredema and detection of paraprotein.11 Immunoelectrophoresis studies can be done at regular intervals, and it is recommended that patients follow up indefinitely in order to detect possible monoclonal hypergammaglobulinemia, which can help diagnose type II scleredema. More research and trials need to be done in order to formulate a universally accepted treatment for patients affected by scleredema. Our patient refused any form of treatment. However, we are continuing to follow up with him to monitor for monoclonal gammopathy. We have discussed his results with his primary care physician, who is also closely monitoring him for diabetes. The patient continues to deny any symptoms.

**References**