Scleredema adultorum of Buschke associated with hypothyroidism and liver cirrhosis

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Introduction

Scleredema adultorum of Buschke is a rare connective tissue disorder, involving the skin. Literature has cited three main causes of scleredema: type 1—an antecedent infection,[1,2] type 2—blood dyscrasia,[3] and type 3—diabetes mellitus.[4] Additional causes include hyperparathyroidism,[5] Sjögren syndrome and rheumatoid arthritis,[6,7] carcinoid tumor,[8] and AIDS and HIV-related lipodystrophy syndrome.[9] Scleredema involves the skin, is usually self-limiting, and is benign with rare systemic manifestations.

The histopathologic examination shows a normal epidermis, with a thickened reticular dermis and amplified spaces—due to deposition of mucin among large collagen bundles. There is a characteristic absence of fibroblast proliferation.

Case Report

Here, we present a case, a 61-year-old male, who is a known case of compensated liver cirrhosis with a past history of being treated for autoimmune thyrotoxicosis, who presented with complaints of alopecia, skin tightening, dry skin, pruritus, and woody indurated plaques on the skin of the upper back, shoulder, and arms. Skin biopsy of the arm revealed the characteristic features of scleredema. He was extensively evaluated for known literature-cited causes of scleredema, and the work up revealed a negative result. He was also found to be hypothyroid on presentation. Hence, we present a case of scleredema occurring in a patient with hypothyroidism and chronic liver disease, which to our knowledge is being described for the first time in literature.
with focally thickened collagen bundles, and a focal increase in mucin—suggestive of scleredema [Figure 2a and b].

The other possibilities considered were scleromyxedema, scleroderma, morphea, and eosinophilic fasciitis.

He had a history of hypothyroidism, but the characteristic absence of fibroblast proliferation in the dermis ruled out the possibility of scleromyxedema. The absence of a history of Raynaud’s phenomenon, sparing of fingers and lungs, negative antinuclear antibody, and absence of collagen accumulation on histopathology of skin biopsy made the possibility of scleroderma/systemic sclerosis unlikely. Multifocal lesions and the absence of lymphocytic infiltrates (plasma cells and eosinophils) ruled out the possibility of morphea and eosinophilic fasciitis.

Hence, the patient was evaluated for the possible etiology for scleredema. He had no prior history of fever, infection, or upper respiratory tract infection and the ASO titer was normal. Blood sugar levels were within normal limits, normal fasting, and postprandial blood sugars with an HbA1c of 5.2%. Serum electrophoresis revealed a normal study. Calcium, phosphorus, and parathyroid hormone levels were normal. Inflammatory markers such as ESR and CRP were normal. Rheumatoid factor and ANA were negative. He had no symptoms suggestive of carcinoid tumors and retroviral status were negative.

However, his blood evaluation revealed severe hypothyroidism with a TSH value more than 100 mIU/L, for which he was started on thyroxine.

He was evaluated for his cause for hypothyroidism. He had a past history of autoimmune thyrotoxicosis, 6 years back. Upon evaluation then, he had low TSH, high thyroid hormone levels, and high anti-TPO antibody levels. A thyroid uptake study revealed a toxic diffuse goiter. He was treated with neomercaptole and propranolol. He was advised surgical thyroidectomy; however, he did not undergo the procedure. He was advised treatment for 2 years, after which he discontinued treatment and was lost to follow-up. During the current presentation, he was found to be hypothyroid. He also had elevated anti-TPO antibodies, lactate dehydrogenase level, elevated transaminases, and creatinine phosphokinase, secondary to hypothyroidism. His USG neck showed atrophied thyroid glands. Therefore, the possibility of hypothyroidism secondary to autoimmune thyroiditis was considered. He was started on thyroxine for the same.

An ultrasonography of the abdomen showed features of parenchymal liver disease with dilated portal vein, with a normal spleen. Fibroscan revealed a median stiffness of 14.3 kPa, suggestive of liver cirrhosis. A/G ratio was normal, and viral hepatitis profile was negative for A, B, C, and E. Upper GI scopy revealed no evidence of esophageal constriction, varices, but showed erosive gastritis. He also had associated thrombocytopenia. He was conservatively managed for the same.

The patient was treated with topical steroid cream—betamethasone plus fusidic acid cream and moisturizing cream. There was symptomatic relief with liberal amounts of steroid cream over a 2-month period.

His liver cirrhosis was closely monitored, not requiring medication as it was a compensated cirrhosis, and hypothyroidism was treated with oral thyroxine.

Since he showed improvement with topical steroid therapy, and the skin lesions were not disabling for the patient, other modalities of treatment were not explored.

### Discussion

Scleroderma adultorum of Buschke is a rare disorder of the connective tissue, involving the skin. On physical examination, it characteristically shows widespread woody induration of the skin, mostly involving the posterior neck, upper back, and chest, with sparing of the distal extremities, unlike systemic sclerosis, where extremities are predominantly involved.

Histopathological examination shows a normal epidermis, thickened reticular dermis demonstrating marked thickening of collagen bundles, separated from one another by mucin, and absence of fibroblast proliferation.

Many modalities of treatment have been proposed, such as physical therapy, phototherapy with UVa1, and immunosuppressive medications such as methotrexate, cyclosporine, and systemic glucocorticoids.

Here, we present a patient with characteristic skin lesions and a typical histopathological diagnosis of scleredema.
Known literature cited causes of scleredema were ruled out using the following investigations [Table 1].

He was also found to be hypothyroid and a case of compensated liver cirrhosis.

The skin biopsy was histopathologically differentiated from scleromyxedema by a characteristic absence of fibroblast proliferation in the dermis. It was differentiated from myxoedema based on the location of lesion and characteristic thickened collagen fibrils, unlike myxedema, where mucin deposition will be sparse, collagen fibrils not thickened and there will be a presence of fibroblast proliferation between collagen bundles.

The patient was treated for his underlying comorbidities—with thyroxine and topical steroid cream–betamethasone plus fusidic acid cream and moisturizing cream. He was symptomatically relieved with 2 months of therapy with liberal amounts of steroid cream. Since the skin lesions were not disabling for the patient and he showed some symptomatic improvement, other modalities of treatment were not explored.

Hence, this is a case of scleredema occurring in a patient with hypothyroidism and compensated liver cirrhosis. Other cases of scleredema in literature had a specific etiology, which were not found in this patient, hence making this association noteworthy.

Conclusion

Autoimmune hypothyroidism and chronic parenchymal liver disease is an extremely rare association with scleredema adultorum of Buschke, which to our knowledge, is being described for the first time in literature.

Table 1: Investigations done to rule out literature cited causes

| Literature cited cause          | Investigation done to rule out the etiology                                                                 |
|--------------------------------|-------------------------------------------------------------------------------------------------------------|
| Streptococcal infection        | He had no prior history of fever, infection, or upper respiratory tract infection and the ASO titer was normal |
| Blood dyscrasia-monoclonal gammopathy | Normal serum electrophoresis                                                                              |
| Diabetes mellitus              | Normal blood sugar values and HbA1c                                                                        |
| Hyperparathyroidism            | Normal serum calcium, phosphorous, and parathyroid hormone levels                                          |
| Sjogren’s syndrome, Rheumatoid arthritis | Negative rheumatoid factor, ANA, and normal ESR and CRP levels                                              |
| HIV and AIDS                   | Negative HIV by ELISA                                                                                    |

Declaration of patient consent

The authors certify that appropriate patient consent was obtained.

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Conflicts of interest

There are no conflicts of interest.

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