SCLERODERMA DIABETICORUM: A CASE REPORT

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ABSTRACT

Scleroderma diabeticorum is a rare dermatological condition that is found in patients with poorly controlled long-standing diabetes. This is characterized by symmetrical, wooden non-pitting induration of the neck, and the upper parts of the shoulders and back. Aetiology of the scleroderma diabeticorum is not well understood. However, the hyperglycaemia-related fibroblast proliferation and extra-cellular ground substance formation is thought to be the main reason for the associated skin changes. Treatment of this condition is a challenge. Good glycaemia control has demonstrated partial resolution in some individuals. However, some studies have not shown any improvement with good glycaemic control. Phototherapy with UVA-1, immunosuppressive drugs and frequency modulated electromagnetic neural stimulation (FREMS) have been successful to a reasonable extent in some patients.

Running title: Scleroderma diabeticorum

Keywords: Scleroderm diabeticorum, Diabetes, Dermatological manifestations

INTRODUCTION

Scleredema diabeticorum is a rare dermatological condition that is seen in the skin of the neck, and the upper part of shoulders and back, which is characterized by symmetrical, wooded non-pitting indurations. In rare occasions, it can also spread to the face, arms and lower parts of the trunk as well as the visceral organs in the abdomen. However, it never involves the hands and feet. This condition was first described by Buschke in 1902 (1) and there had been large number of cases reported since then.

Depending on the clinical presentation, scleroderma is divided into 3 clinical subtypes. Scleroderma that develops after acute febrile illnesses and recovers within several months is termed type 1. The Type 2 scleroderma develops without any precipitating illnesses and the condition takes a chronic course in these patients. Type 3 scleroderma develops in patients with long-standing type 1 and type 2 diabetes with poor metabolic control and named as scleroderma diabeticorum.

Aetiopathogenesis of scleroderma diabeticorum is not well understood. Some suggest that glucose may be stimulating the fibroblast proliferation and extra-cellular matrix formation, which leads to increased amounts of ground substance formation. In additions, the non-enzymatic glycosylation of collagen fibers can also alter the natural degradation of these components (2). Reversal of this process has been observed in some individuals with good glycaemic control and this observation has been supportive of this hyperglycaemia theory (3, 4). There is a suggestion that long-term diabetes that leads to microvascular damage and chronic hypoxia associated with that also could contribute to this process (5). However, some patients have shown some improvement with immuno-modulating drugs such as cyclosporine suggesting a contribution from an immune-mediated mechanism. However, the lack of T cells in most of these lesions rule out the immune theory as the mechanism for this dermatological condition (6).

Slcreroderma diabeticorum is a rare disease and the treatment of this condition is a challenge. Here we report a case of a patient with long-standing type 2 diabetes mellitus with poor glycaemic control diagnosed to have scleredema diabeticorum.

CASE REPORT

Mrs. Shiromala is a 43-year-old patient with type 2 diabetes who had been taking treatment for the last 13 years. At the age of 28 years, during her third pregnancy, she was diagnosed to have gestational diabetes mellitus and hasn't recovered from diabetes since then. Initially, she has been taking treatment from her general practitioner and several times from a physician in the private sector. However, her glycaemic control had been poor and HbA1C has been above 9% on most occasions.

For the last 3 to 4 years, she had observed a change in the skin in the back of her neck, shoulder and upper back where she noticed a wood like thickening of the skin with discomfort. Her condition was gradually increasing started spreading towards the lower parts of the back. However, this was not associated

with any joint pains or swelling. Except for medication that she had been taking for diabetes and hypercholesteraemia, she had not been on long term medication for any other illness. Since the condition was progressing, she has consulted a dermatologist for specialized treatment. There, she was diagnosed as having sclerodaerma diabeticorum. She was reassured and referred to our diabetes clinic for optimization of glycaemic control.

On examination, she was an average build female with a body mass index (BMI) of 22.4. Her blood pressure was normal and showed no clinical evidence of microvascular complications of diabetes. The skin in the back of her neck and upper back was thickened and had a waxy, indurated appearance (Figure 1). She did not have telangiectasia, joint swelling or joint deformities.

Her baseline HbA1C was 10.2% while on metformin, gliclazide and mixtard 30 insulin and the serum cholesterol level was 148mg/dL while on atorvastatin 20mg at bedtime. Her glyaemic control was optimized by changing over to basal-bolus insulin treatment along with metformin and gliclazide. She was advised to continue with the same dose of atorvastatin for hypercholesterolaemia. Her last HbA1C was 7.6% with current treatment. Even with good glycaemic control, we didn't see any improvement with her skin condition. However, we have not seen any progression of the disease for the last 18 months.

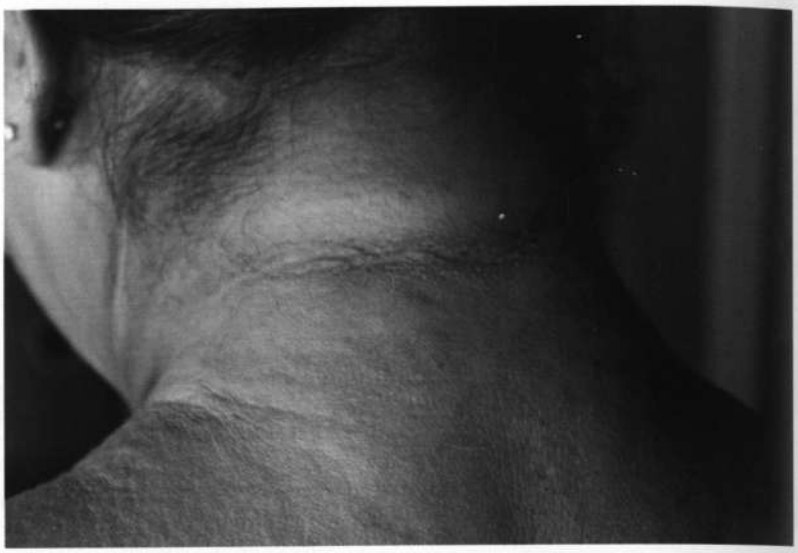

Figure 1. Figure illustrate the waxy indurated thicken skin in the neck and upper back region

DISCUSSION

Scleredema diabeticorum is a disease that is commonly seen in males with a male to female ratio of 10: 1 (7). Patients with chronic type 2 diabetes mellitus are prone to this disease particularly in presence of obesity. The occurrence of this disease in childhood is undiscovered in medical literature.

Scleredema diabeticorum is essentially a clinical diagnosis. However, a skin biopsy can be useful in confirming the clinical diagnosis. Histologically, these lesions are seen as thickened dermis with enlarged collagen bundles in the deep reticular dermis with clear spaces in between, filled with mucin.

Treatment of scleredema diabeticorum has always been a clinical conundrum. Currently, there is no standard effective method of treating this disease. Studies have not shown a significant clinical improvement by improving the glycaemic control (6). However, several studies bear witness to partial improvement of scleroderma with optimal glycaemic control (3, 4).

At present, use of ultraviolet A-1 (UVA-1) phototherapy has gained worldwide popularity. The first case of scleredema diabeticorum successfully treated with UVA-1 was published in 2004. Synthesis of the enzyme, collagenase, which enhances degradation of sclerotic tissue, is found to be stimulated by UVA (8-10). Photochemotherapy with psoralen and ultraviolet A (PUVA) therapy is found to be even more effective than UVA since PUVA has shown to reduce types I and III collagen synthesis levels by increasing collagenase synthesis by fibroblasts and by inhibiting de novo synthesis of collagen mRNA (11, 12).

Immunosuppressive treatment with colchicine, cyclosporine, corticoids, and methotrexate has also shown to be effective in some cases while in some others they haven't been successful (13 -16). Improvement of scleredema diabeticorum has also been reported with electron-beam radiotherapy. However, the effect of this treatment modality has been short lived (17). One author has described some benefit with frequency modulated electromagnetic neural stimulation (FREMS) that is used as treatment modality used in symptomatic diabetic neuropathy (18)

CONCLUSION

Scleredema diabeticorum is a rare cutaneous disorder usually seen in patients with long-standing, poorly controlled type 2 diabetes mellitus. Various treatment modalities have been assessed in the management of the disease but there is no consensus with regard to a standard effective treatment method.

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