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# Scleroderma-like skin changes not involving the hand in a prepubertal male with type I diabetes mellitus

## A case report

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**Abbreviations:** T1DM, type I diabetes mellitus; $HbA_1C$, hemoglobin $A_1C$

To our knowledge there have been no reports of scleroderma-like skin changes, not affecting the hand in prepubertal patients with Type I Diabetes Mellitus (T1DM). We report a prepubertal caucasian male with T1DM, and early morphea-type skin changes of the trunk and extremities, not involving the hand.

### Introduction

Scleroderma-like skin changes as part of "diabetic hand limited joint mobility syndrome" is a clinical entity reported in 10–50% of adolescents and adults with diabetes mellitus.[1] The skin changes may occur early in the disease. The assumption is that vessel and connective tissue alterations as well as the impairment of the immune system and other associated metabolic changes caused by diabetes play an important role.[2] Male and female patients are equally affected and there is no racial predilection.[1] Its frequency appears to be related to duration of diabetes and increasing age, and most studies have failed to show a relationship to glycemic control ($HbA_1c$).[2] Its importance appears to lie in its association with a two- to threefold greater risk of microvascular complications such as retinopathy and nephropathy during the first 15–20 years of diabetes.[1] To our knowledge there is no report of scleroderma-like skin changes, not affecting the hand and joint in a prepubertal patient with T1DM.

### Case Report

Patient is a 9-year-old boy diagnosed with T1DM at the age of 5 years. One year after diagnosis, he developed lipoatrophic skin changes specifically at insulin injection sites on his upper arms and thighs. His glycemic control was complicated by episodes of hypoglycemia and his hemoglobin $A_1C$ ($HbA_1C$) was in the 6.5–7% range. The skin lesions were initially attributed to the insulin type, and he was switched from NPH insulin and regular insulin to Humalog insulin, delivered by an insulin pump. He subsequently developed similar lesions on is abdomen at sites not limited to the insulin infusion (**Fig. 1**).

On physical examination, weight was 37.6 kg (45%) and height 141 cm (20%) and normal pulse and blood pressure. He has painless, indurated, discolored, indentations of the skin of both upper extremities, thighs and lower abdomen, both at the sites of and far from insulin infusion (**Fig. 1**). There was no evidence of hand involvement such as thickening and induration of the skin of the dorsum and proximal interphalangeal joints. There was full range of motion in the joints, no flexion contractures, no trigger finger, Raynaud's phenomenon or telangiectasia. Other laboratory tests included ACA (anti-centromere antibody), ANA (anti-nuclear antibody), Scl-70 and celiac panel were all negative.

Six years later, the lesions continue progressing with the old ones enlarging and new ones developing at sites remote from insulin injections. He had new scleroderma lesions on his shoulder, abdomen and thigh. Recent follow up visit at the age of 17 years and 6 months and again noted to have stable scleroderma like changes on his abdomen and still there is no hand and joint involvement. He has fair diabetes control ($HbA_1C$ 7.8%) and no evidence of nephropathy or retinopathy.

A biopsy from the affected areas revealed attenuated epidermis, dermis expanded with fibrous bands, atrophy of the adnexal structures (**Fig. 2**). There is no evidence of specific injury or inflammation of the subcutaneous fat. All of the above findings are consistent with the diagnosis of sclerodermal-like skin

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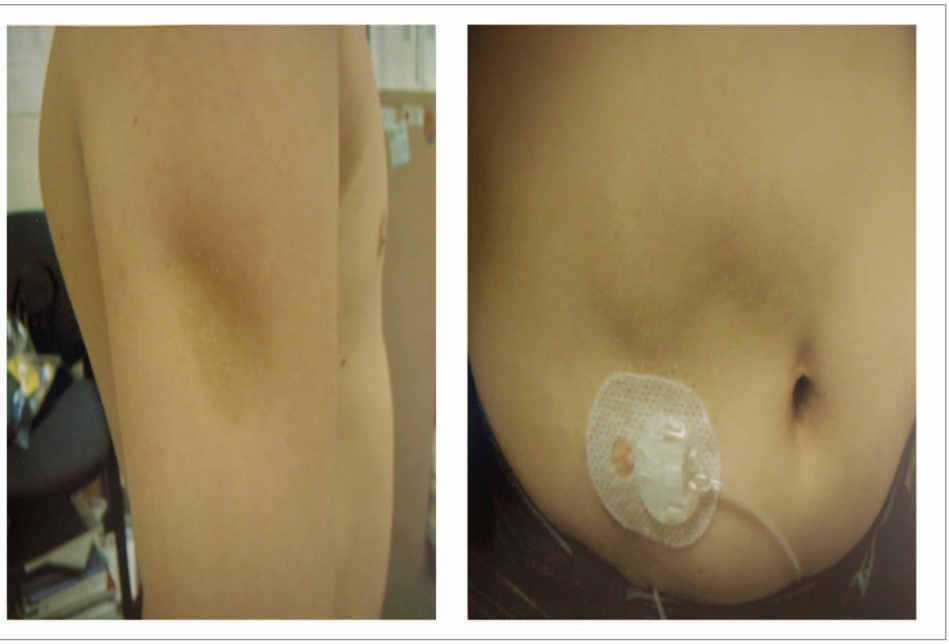

**Figure 1.** Patient at age 11 years 8 months with significant scleroderma-like skin changes.

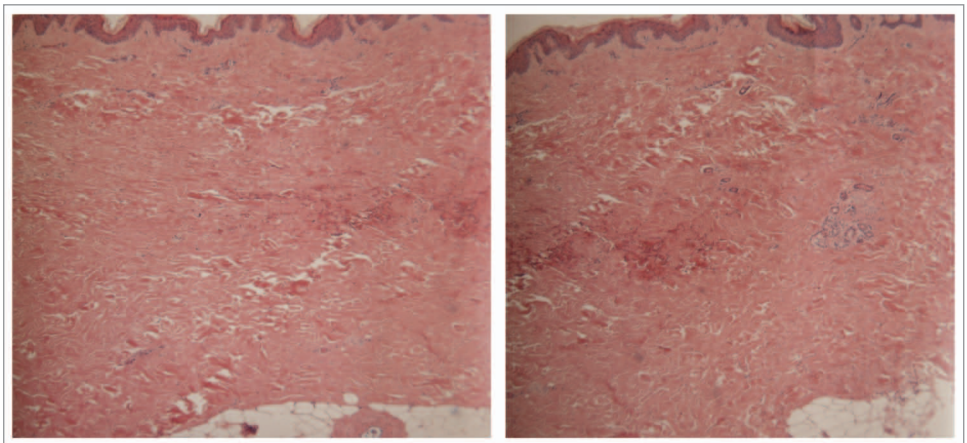

**Figure 2.** Skin biopsy reveals the diagnosis

changes. Mucin stained had been performed on the skin biopsy specimen. The histologic features suggestive of a "scleroderma-like" disorder. However, histologic features are not diagnostic of scleroderma."

## Discussion

This is the first case report of a prepubertal male with T1DM and extensive scleroderma-like skin changes without evidence of the diabetic hand and joint syndrome whether this finding could be a variant of milder form of diabetic hand syndrome was unclear. This presentation is not consistent with linear scleroderma or diffuse morphic especially since the lesion originated in the areas of the insulin injections. In those patients with diabetic hand syndrome, the development of skin manifestation is influenced by the duration of diabetes and association with development of diabetic microvascular complications. Defective collagen formation may be due to the accumulation of advanced glycosylation end products has been assumed as the underlying pathogenic process.[1] In one study by Garza-Elizondo et al. it as noted that patients had scleroderma-like skin changes involving the PIP only and the juvenile patients had diabetes for 6 or more years and the difference in the disease duration between those with hand changes and those without was significant.[4] Therefore, most reports of scleroderma-like changes are noted in the hands of patients with T1DM. This skin changed initially was thought to be a lipohypothropy related to insulin injection. However the pathology report showed the change in the dermis not subcutaneous defect. There is also a case report of sclerderma and type I diabetes.[5] However the pathogenesis of these two co-existing conditions was unclear. It has been thought that interferons (IFNs) could play a major role. IFNs are well-known immunomodulators and inhibitors of collagen production. Beside their immunomodulatory action, IFNs can also be linked to autoimmune diseases.[6]

It is noted that factors such as stratum corneum adhesion and accelerated aging of the skin may be implicated in the development of ichtyosiform skin changes in Type I DM patients and it could be explained by structural changes in skin proteins due to advance glycosylations.[3] There is strong correlation between ichthyoiform skin changes and diabetic retinopathy suggesting microvascular involvement in the pathogenesis of skin changes.[3,7] Further observation will be needed in order to evaluate whether these lesions will progress to the full spectrum of the Limited Joint Mobility syndrome and represent an early risk marker for the development of diabetes complications.

### Disclosure of Potential Conflicts of Interest

No potential conflicts of interest were disclosed.

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P.P. is currently on the speakers bureau for EMD Serono, Genetech, Inc., Endo phamaceutical and Novo Nordisk and

serves as a consultant/advisor to EMD Serono. A.R. is currently on the speakers bureau for Amgen, Abbott and Pfizer and serves as a consultant/advisor to Amgen, Abbott, Pfizer, Merck and Novatis.

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