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CASE REPORT

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Scleredema: report of a case

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Abstract We describe a patient with scleredema. Thickened skin was observed in the posterior region of her neck, shoulders, and back. A skin biopsy revealed thickened dermis consisting of numerous thickened collagen bundles. Laboratory data revealed that she was suffering from diabetes mellitus (DM). The skin lesions ameliorated within a period of one month when her DM condition was controlled by oral hypoglycemics. Scleredema is a rare connective tissue disorder; its cutaneous manifestation should be distinguished from scleroderma.

Key words Scleredema · Scleroderma · Diabetes mellitus · Buschke

Introduction

Scleredema is an uncommon dermatosis of unknown etiology, characterized by thickened, indurated skin, sometimes with erythema. Histopathological results reveal thickened dermal collagen with a mild infiltration of mucin in the deeper regions of the dermis.[1–3] We describe a case of scleredema associated with diabetes mellitus.

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Case report

A 41-year-old women was referred to our hospital in 1990 for stiffness of the nucha, shoulders, and back. She had had these symptoms for several years. She had been informed that her urinary sugar was positive 7 years ago, but she did not follow up on her diabetic condition. She had felt general fatigue and thirst since 1993. Her body weight decreased by 7.0 kg within a period of 6 years. She had not had a recent episode of upper respiratory infection. Physical examination revealed her body weight to be 59.5 kg and her height to be 155 cm. Her blood pressure was 110/70 mmHg. Thickened, indurated, edematous, and erythematous skin was observed in the posterior region of her neck, both shoulders, and back (Figs. 1 and 2). The skin lesion was not observed on her face, hands, or fingers. Raynaud's phenomenon was absent. Laboratory examinations revealed WBC 7100/mm^3, RBC 401 $\times$ 10^4/μl, Plate 28.8 $\times$ 10^4/μl, ESR 11 mm/h, CRP (−), total protein 5.9 mg/dl, albumin 3.9 mg/dl, creatinine 0.5 mg/dl, GOT 8 IU/L, GPT 7 IU/L, LDH 282 IU/L, CK 48 IU/L, T-Chol 231 mg/dl, and fasting blood sugar (FBS) 366 mg/dl. Urinalysis revealed glucose 38.0 g/day. Urinary protein and ketone were not demonstrated. HbAl was 13.0% (normal range 2.0–8.0%), and HbAlc 11.0% (2.0–6.0%). Urinary c-peptide was 85.8 μg/day (41–145 μg/day). Autoantibodies, including ANA, RF, anti-dsDNA, anti-SS-A and B, anti-U1 RNP, anti-Scl-70, and anti-centromere antibodies were not demonstrated. The ASK and ASO were negative. Thyroid function tests were within the normal range. Histological examination of the skin revealed markedly thickened dermis consisting of numerous thickened collagen bundles separated by clear spaces, causing fenestration of the collagen. No increase in the number of fibroblasts was observed (Fig. 3). Diabetic retinopathy was demonstrated (Scott II). No abnormal finding was demonstrated by electrocardiogram or chest X-p. We diagnosed that the patient was suffering from scleredema associated with non-insulin-dependent diabetes mellitus (NIDDM) (diabetic scleredema). She was treated with glibenclamide at 2.5 mg/day and a diet of 1400 Cal/day. One month after starting the treatment, when her FBS had

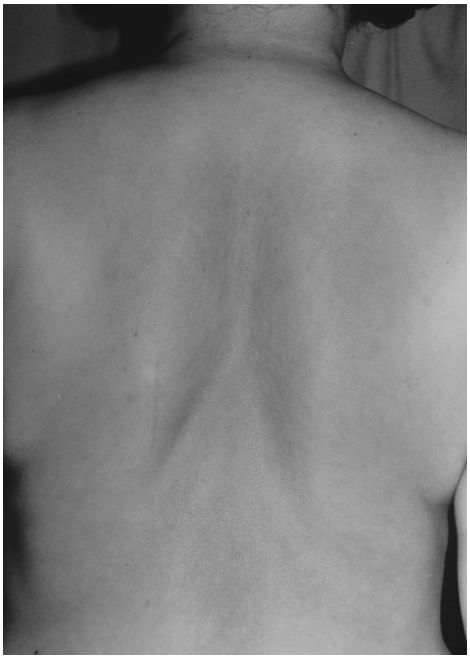

Fig. 1. Thickened, indurated, edematous, and erythematous skin was observed on both shoulders and back

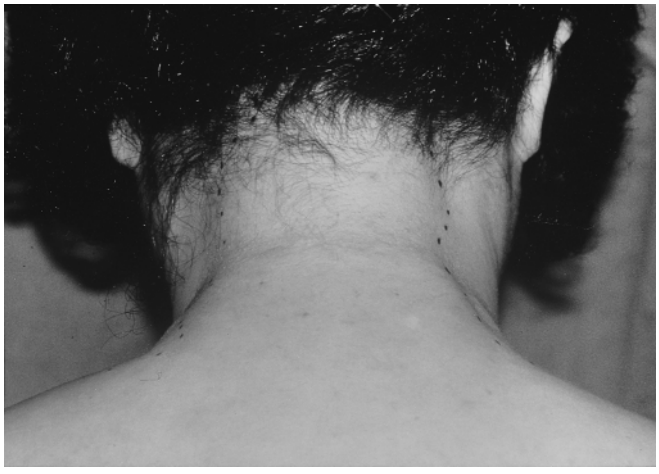

Fig. 2. Thickened and edematous skin at the nucha was demarcated with normal skin. The region is shown by *black dot* markings

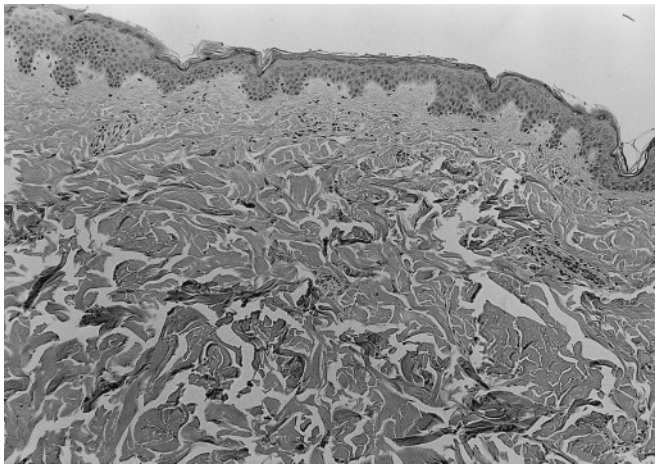

Fig. 3. Microscopic section of skin from the nuchal region, showing numerous thickened collagen bundles in the dermis separated by clear spaces (hematoxylin and eosin, ×100)

decreased to 125 mg/day and urine glucose was within 5 g/day, erythema, pitting edema, and thickened, indurated skin lesions had dramatically improved.

Discussion

Scleredema was originally described by Piffard, although it was recognized as a distinct entity following the publication by Buschke.[1,2] This disorder is characterized by symmetric, painless, edematous induration of the skin of the face, scalp, neck, trunk, and upper extremities. In contrast to systemic sclerosis, the distal extremities are seldom affected. Other manifestations include hydrarthrosis, pleural and pericardial effusions, widespreaded involvement of skeletal or cardiac muscle, and macroglossia. Recent streptococcal infection in childhood or as an adult (scleredema adultorum of Buschke[2]) and diabetes mellitus (DM) (diabetic scleredema[3–6]) are the two most frequent associations. Histochemical studies have revealed swollen collagen bundles and the accumulation of mucopolysaccharide (hyaluronic acid) in the dermis, subcutis, and skeletal muscle. In the dermis, in particular the numerous thickened collagen bundles cause fenestration of the collagen, leaving clear spaces. Increased synthesis and deposition of collagen and hyaluronic acid has been reported.[7] No increase in the number of fibroblasts was observed, although histological findings show that this does occur in scleroderma. This condition either resolves spontaneously after 6–12 months or persists for many years.[8]

The incidence of scleredema associated with DM has been reported in approximately 1.0–2.0% of patients with DM.[9,10] Compared with scleredema associated with streptococcal infection, diabetic scleredema develops insidiously.[11] Most patients with diabetic scleredema are obese, poorly controlled, insulin-dependent adults and are associated with diabetic micro- and macro-angiopathies.[6,12] Our case was diabetic scleredema, which occurred insidiously along with the development of DM.

Although scleredema is not an uncommon disorder to dermatologists or physicians specializing in DM, rheumatologists should be aware of this scleroderma-like connective tissue disorder.

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