Scleredema diabeticorum – A case report


Abstract

Scleredema, a medical problem first described by Buschke in 1902, is a rare scleromatosis of unknown aetiology, which is characterized by wooden, nonpitting induration of the skin. This skin disorder first affects the neck and face and may symmetrically spread to the shoulders, trunk, arms, and legs. We report a case of a 65 year-old hypertensive and diabetic male who presented with marked thickening of the upper back with obvious peau d’orange changes. Association with other complications of diabetes is not certain and treatment is difficult, though improvement is possible with optimal glycaemic control. Scleredema diabeticorum is rare and is a cause of significant morbidity in diabetes.

Keywords: Scleredema; Diabetes; Hyperglycaemia

1. Introduction

Scleredema, a medical problem first described by Buschke in 1902, is a rare sclerodermatosis of unknown aetiology, which is characterized by wooden, nonpitting induration of the skin. This skin disorder first affects the neck and face and may symmetrically spread to the shoulders, trunk, arms, and legs. We report a case of a 65 year-old hypertensive and diabetic male who presented with marked thickening of the upper back with obvious peau d’orange changes. Association with other complications of diabetes is not certain and treatment is difficult, though improvement is possible with optimal glycaemic control. Scleredema diabeticorum is rare and is a cause of significant morbidity in diabetes.
lymphocytic infiltrate in the dermal lesions seems to rule out a T-cell-mediated aetiologic mechanism (Martin et al., 2011). Thus, there is a need for more studies to focus on scleredema diabeticorum, its features and its associated risk factors that may lead to its development.

In this report, we describe the features of scleredema diabeticorum in a 65 year old diabetic male with long-term hypertension and poor glycaemic control. We also aim to identify other risk factors that may lead to the development of scleredema and to discuss current treatments.

2. Case

A 65 year old civil servant a known hypertensive of 24 years duration was diagnosed diabetic 2 years ago. He initially presented at the A & E with hyperglycaemic emergency having had fever, loss of appetite, gradual alteration of level of consciousness. This was precipitated by a possible urinary tract infection as he had complaint of dysuria, frequency and urgency before his level of consciousness deteriorated. General physical examination revealed a middle aged man with altered level of consciousness, restless, severely dehydrated, with deep sighing breathing, he was not pale and was not jaundiced. He looked obese with a waist circumference of 121 cm. The admitting blood pressure was 110/70 mmHg with a pulse rate of 124 beats/min. The admitting blood sugar was 450 mg/dl. The initial serum electrolyte urea and creatinine showed a slightly elevated urea (10.4 mmol/L) but normal creatinine (112 mg/dl) and a potassium of 4.2 mmol/L, bicarbonate was 13 mmol/L and sodium was 142 mmol/L. He was managed aggressively with fluid replacement due to severe dehydration, Insulin therapy, potassium replacement to correct hypokalaemia due to the four hour potassium of 3.4 mmol/L and antibiotics. He eventually regained consciousness within the first 24 h on admission and commenced oral feeding the second day. Intravenous fluid was stopped at that point and intravenous insulin converted to subcutaneous. Lipid function test done on the fourth day of admission showed a total cholesterol of 7.5 mmol/L, HDL of 0.8 mmol/L, LDL of 4.2 mmol/L and TG of 3.6 mmol/L. Statins (simvastatin) were added to his conventional antihypertensive therapy of 10 mg lisinopril and 25 mg of hydrochlorothiazide which he claimed compliant with. He was discharged after spending 8 days on admission. Following resolution of the hyperglycaemic emergency, his blood sugar control has been suboptimal in the last 2 years despite optimizing diet and oral hypoglycaemic agents. Routine subcutaneous insulin was therefore added to his medication with which he achieved reasonable glycaemic control. Clinical features of long standing hypertension such as thickened arterial walls, cardiac hypertrophy and background hypertensive retinopathy were present. There were no paraesthesia and intermittent claudication. The renal function test

Figure 1. Showing peau d’orange changes in the upper back.

Figure 2. H&E × 6.3 section shows skin tissue with hyperpigmentation of stratum basalis. There is focal lymphocytic infiltration of the reticular dermis.
was normal. There was a wood like thickening of the skin of the upper back with obvious peau d’orange changes in the same region (Fig. 1). Diagnosis of scleredema diabeticorum in a patient with type 2 diabetes with suboptimal glycaemic control was considered and skin biopsy was taken with a 3.5 mm punch to confirm the diagnosis. The histology result is shown below (Figs. 2 and 3). No specific treatment was instituted for the scleredema diabeticorum, but he is being followed up to evaluate the effect of optimizing glycaemic control with the added subcutaneous insulin on the condition.

3. Histology report (Figs. 2 and 3)

Section shows skin tissue covered by intact epidermis of keratinizing stratified epithelia.

The reticular dermis is greatly thickened with bundles of collagen fibres.

The collagen fibres are broadened and are abnormally separated by clear spaces.

Here was a dense fibrosis of the papillary dermis with scanty infiltration by inflammatory cells mainly lymphocytes. There are few fibroblasts within the reticular dermis.

4. Discussion

Scleredema diabeticorum was recognized in 1970 as a syndrome (Cohn et al., 1970), and presents insidiously with painless, symmetric thickening of the skin of the upper back and neck. Spread to the face, shoulder and anterior torso may occur. The skin retains a non-pitting peau d’orange quality. Identical changes occur with post streptococcal pharyngitis, though this is sudden and the symptom remits over time.

Scleredema diabeticorum has a reported a prevalence of 2.5% in patients with type 2 diabetes with a male preponderance of 10:1 (Cole et al., 1983). It is a disease of long standing diabetes, associated with obesity. Most patients have type 2 DM. It has not been reported in children.

The aetiology of scleredema diabeticorum is unclear. Glycation of skin collagen and skin hypoxia due to microangiopathy may be key factors. Microscopic examination shows a thickened dermis with large swollen collagen bundles, particularly type 1 collagen, separated by ground substance and wide clear spaces. There is no inflammation of the dermis and no increase in the number of dermal fibroblasts, features which distinguish scleredema from scleroderma. There may be increased numbers of mast cells and variable expression of glycosaminoglycans. Type 1 subunits of procollagen 1 and 111 messenger RNA (mRNA) and fibronectin mRNA were elevated in fibroblasts of cultured scleredema skin (Cole et al., 1983).

Treatment for scleredema diabeticorum is usually unsuccessful. Case reports describe treatment with radiotherapy, low dose methotrexate and prostaglandin E1, 4, 5 (Seyger et al., 1999). Glycaemic control with use of oral hypoglycaemics is usually very difficult.

A study by Leung and Chong (1998) found that 92% of twelve Chinese adult patients with scleredema have hypertension requiring medical treatment. The reason for this very significant association is not very clear but the fact that diabetes and hypertension are known risks for each other and the attendant dyslipidaemia that mostly complicates long standing diabetes mellitus may be an important factor. There is therefore a need for future study to look into these three entities and clearly define the exact aetiology of the associations.

Martin et al. (2011) reported about a 53 year-old white male with type 2 diabetes mellitus and scleredema who was given PUVA treatment and physiotherapy with the amelioration of mobility and acquiring some elasticity of
the upper back. The authors noted that good metabolic control seems not to preclude the development of scleredema in diabetics and that PUVA treatment and physiotherapy are therapeutic options that seem to be of some help (Martin et al., 2011).

A study by Thumpimukvatana et al. in 2010 has described two cases of scleredema diabeticorum with substantial clinical improvement from a course of medium dose (60 J/cm^2) ultraviolet A1 radiation therapy (Thumpimukvatana et al., 2010). Furthermore a study by Kokpol et al. in 2012 reported a case of scleredema diabeticorum that was successfully treated by combining local PUVA treatment with colchicine (Kokpol et al., 2012). The results were similar to those of Yuksek et al. who reported a case of scleredema that improved markedly with low-dose broad-band UVA plus colchicine treatment (Yuksek et al., 2010).

Konemann et al. in 2004 described the case of a 58-year-old patient with scleredema of the neck and upper trunk who was treated twice within 6 months by electron-beam radiation therapy. The authors concluded that regardless of the possible mechanisms in pathogenesis and treatment of scleredema adultorum Buschke, the application of ionizing radiation is an important, effective and well-tolerated therapy option in the treatment of severe cases and may be considered as the first-line treatment of this disease (Konemann et al., 2004).

5. Conclusion

Scleredema diabeticorum is a rare cutaneous disorder seen in long standing type 2 DM. It has been shown in this case report that this disorder is characterized by thickening and induration of upper back. There are various treatments however, evidence is still lacking as to whether they are truly effective or not, given the smaller population studied.

Conflict of interest

None.

References