Scleredema of Buschke: a case report

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Abstract

Scleredema of Buschke or scleredema adultorum is a rare sclerotic disorder. It has been reported as an unusual skin complication in diabetic patients with a long history of poor metabolic control. This disease is usually localized in the proximal parts of the trunk including nape of neck, back and shoulder areas. Consequently, the range of movement of the affected areas could be altered. Histologic examination of a skin biopsy shows deposits of collagen and aminoglycans in the dermis. The pathophysiology of scleredema of Buschke and its relationship with diabetes are not clearly identified. Few therapeutic options are proposed without satisfying results. We report an unusual case of a 45-year-old woman with a long history of poorly controlled diabetes type 1 who presented with indurated edema of the trunk. The presumptive diagnosis of scleredema adultorum was confirmed by the presence of typical histological findings and after eliminating other diagnosis of edema.

Key words: Scleredema, Buschke, adultorum

Introduction

Scleredema adultorum of Buschke characterized by thickening and hardening of the skin is an uncommon sclerotic disorder of unknown etiology. It is characterized by thickened collagen bundles and accumulation of acid mucopolysaccharides in the dermis.\(^1\) It affects mostly obese patients with insulin dependent diabetes.\(^2\)

However, as this skin disorder is rather uncommon and develops insidiously, it seems useful to sensitize all clinicians to this diabetic complication by reporting a case observed in our institution.

Case Report

A 45-year-old woman, with diabetes type 1 for the past 14 years, noticed a generalized indurated edema lasting for six months and a progressive limitation of the shoulder motility. The examination of the shoulders, the upper part of the back, the neck and the trunk showed hardness and thickness of the skin with inability of either depressing or pinching the skin (Figure 1). The skin of the upper extremities excluding the hands was also affected in the same way. The BMI was 42.6 kg/m\(^2\). The remainder of the physical examination was unremarkable.

Her diabetes was poorly controlled with HbA1C values ranging between 10 and 11.5% despite insulin treatment. She had preproliferative diabetic retinopathy. Echocardiography, ECG and chest X-ray were normal. The results of laboratory investigations including hepatic, renal and thyroid function tests were normal. Serum protein electrophoresis and immunoserology (antinuclear antibody, extractable nuclear
antigen antibody screen) were also normal. Skin biopsy of the affected area showed marked thickening of the collagen bundles in the dermis and a mild perivascular mononuclear inflammatory infiltrate. These histologic features in conjunction with the clinical picture confirmed the diagnosis of Buschke's scleredema.

A better management of the diabetes was recommended to our patient by improving the insulin therapy. After one year of therapy, the HbA1C was 8% and but there was no improvement of the skin lesions.

Discussion

Scleredema adultorum of Buschke is an uncommon disorder of unknown origin. It has been described as a differential diagnosis of systemic sclerosis. This skin disorder is characterized by thickening and hardening of the dermis affecting the neck, shoulders and the upper part of the back. The face, the arms and the rest of the trunk can be affected rarely. Sometimes, the viscera may be involved like heart, ocular muscles, pharynx, parotid glands, pleurae, peritoneum, spleen or liver. The diagnosis is generally made by physical examination. The skin biopsy should be done to make the final diagnosis. It shows accumulation of collagen within the reticular dermis with deposition of mucin. But the presence of mucin deposits is not obligatory to make the diagnosis.

Patients with type 2 diabetes are predisposed to develop scleroderma of Buschke. Risk factors described in patients with scleroderma of Buschke are: a long duration of diabetes, poorly controlled diabetes, obesity, hypertension and male gender. Frequency of this skin disorder related to diabetes remains underestimated and varies between 2.5 to 14%.

The prognosis of scleroderma of Buschke is generally benign. However, in some cases functional respiratory complications may occur as a consequence of skin induration. Management of scleroderma of Buschke is often difficult. Tight glycemic control and physiotherapy are recommended as a first line treatment.

Immunosuppressive therapy has also been tried e.g. corticosteroids and methotrexate. Ultraviolet A-1 phototherapy and photochemotherapy with PUVA therapy may be advised. But these approaches have been tried on very few patients with inconstant results. Electron beam therapy can be proposed as a successful treatment in patients with long history of diabetes associated scleroderma.

Conclusion

The frequency of scleroderma diabeticorum in diabetic population is probably underestimated. Clinicians should be aware of this skin complication of diabetes which can be easily diagnosed by simple clinical examination. Improvement of diabetes control and physiotherapy remains the first line treatment.


References