Scleredema adultorum of Buschke over an unusual site associated with pregnancy

C. Sujatha Vinod, H. Ambika, Hariharasubramony Ambika, Nithya Reddy, Jayantha Kumar De

ABSTRACT

Scleredema adultorum of Buschke is characterized by symmetrical, diffuse, non-pitting erythematous swelling of the skin due to accumulation of collagen and mucopolysaccharides in the dermis. Herein we report a case of scleredema over an unusual site in a pregnant woman.

Key words: Pregnancy, scleredema, unusual site

INTRODUCTION

Scleredema adultorum of Buschke is characterized by symmetrical, diffuse, non-pitting erythematous swelling of the skin due to accumulation of collagen and mucopolysaccharides in the dermis. Of unknown etiology, it may be associated with infections, blood dyscrasias, or diabetes mellitus. The swelling and induration of the skin usually begins on the neck and upper back and slowly spreads to the other areas. We herein report a case of scleredema adultorum in a pregnant woman at an unusual site, associated with streptococcal infection.

CASE REPORT

A 24-year-old full-term pregnant woman presented with diffuse swelling and thickening of the skin over the abdomen and thighs of 2 weeks duration. It started over the abdomen and slowly spread to the medial aspect of both thighs. It was associated with dull aching pain over the same sites. Patient gave a history of upper respiratory infection one week prior to onset of symptoms for which she was given a course of antibiotics. There was no history of fever, local rise of temperature, or past history of diabetes mellitus. On examination, her blood pressure was 110/70 mmHg, the skin over the lower abdomen, pubic area, and medial aspect of the both thighs showed a diffuse swelling with induration and peau d’orange appearance. There was no pedal edema or edema of the vulva. Ophthalmic examination was within normal limits. Blood investigations showed hemoglobin 11.2 gm%, total count 8800/mm³, erythrocyte sedimentation rate 28 mm/hr, fasting blood sugar 82 g/dl, and postprandial blood sugar 128 g/dl. Liver and renal function tests were within normal limits. C-reactive protein and antistreptolysin O titer were elevated. Blood culture and throat swab culture showed no growth. Ultrasonography of the affected area showed thickening and hypechogenicity of the skin and subcutaneous fat over the lower abdomen and medial aspect of thighs. Venous doppler of the involved area was unremarkable. Skin biopsy and histopathologic examination revealed mild thinning of the epidermis with thickening of the dermis. Deep dermis showed enlarged bundles of collagen separated by clear spaces causing fenestrations. Alcian blue staining showed increased mucin and thickened collagen in the middle and deep dermis.

From the history, clinical examination, and histopathologic findings, a diagnosis of scleredema adultorum was made and the patient was put on a course of amoxicillin–clavulanic acid for 14 days. Patient delivered a healthy male baby weighing 3.2 kg by normal vaginal delivery. The swelling and induration slowly reduced and subsided completely within 3 months.

DISCUSSION

Although the name Bushcke is associated with scleredema, it was originally described by Pitford in 1876. Its relationship with diabetes mellitus was
established in 1970.[1] It is seen more commonly in females, but the type associated with diabetes is more common in males. Various hypotheses have been put forward regarding the pathogenesis of scleredema, which include obstruction to lymphatic channels by inflammation, streptococcal sensitivity, immune sensitization phenomenon, increased collagen synthesis and mucin deposition due to microvascular damage and resultant hypoxia, and resistance to collagen degradation by collagenases.[2]

Three distinct types have been recognized. The first type is preceded by an infective episode, commonly Group A streptococcal infection in the respiratory tract. After a prodromal symptom of fever and malaise, it starts as hardening of the skin over cervicofacial region, which gradually extends to involve the trunk. This type resolves completely within few months. The second type is associated with blood dyscrasias like multiple myeloma and paraproteinemias. The third type is scleredema diabeticorum which is seen in obese, middle-aged men with long-term type 2 diabetes, having bad metabolic control, and in the presence of specific complications of diabetes mellitus.[3]
The specific histological findings of scleredema are thickness of the dermis due to both enlarged collagen bundles and presence of clear spaces or fenestrations between them filled with mucopolysaccharides. Mucin deposits are more likely to be observed in the dermis by Alcian blue, toluidine blue, or colloidal iron staining.

The differential diagnoses include cellulitis, myxedema, scleroderma, eosinophilic fasciitis, and cutaneous amyloidosis.

Management of scleredema is often difficult. Antibiotics are recommended in those cases associated with streptococcal infections, though they do not appear to shorten or cure skin conditions in scleredema.[4] Other reported therapies include oral or intralvesional steroids, cyclosporine, colchicine, low-dose methotrexate, high-dose penicillin, electron beam therapy, extracorporeal photopheresis, psoralen followed by ultraviolet A Psoralen followed by ultra violet A (PUVA) and intravenous immunoglobulins.[3] Prognosis depends on the etiology. Though in most cases it is self-limited, death due to cardiac and pulmonary involvement has been documented. Involvement of unusual sites like periorbital area and thighs has been reported in the literature.[6,7] Here, we report a rare case of scleredema over the lower abdomen and thighs in a pregnant woman, associated with streptococcal upper respiratory tract infection.

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Cite this article as: Vinod CS, Ambika H, Ambika H, Reddy N, Kumar De J. Scleredema adultorum of Buschke over an unusual site associated with pregnancy. Indian Dermatol Online J 2014;5:466-8.

Source of Support: Nil, Conflict of Interest: None declared.