

Scleredema in a Child

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Abstract

A rare case of scleredema in a seven year old child is presented. The relevant literature regarding clinical presentation, differential diagnosis and treatment of this disease is also being reviewed.

Key Words

Scleredema, Scleroderma, Scleredema adutorum

Introduction

Scleredema (*Scleredema adutorum*, *Scleredema of Buschke*) is a rare connective tissue disorder. The onset is sudden, with brawny edema of the face and neck that spreads rapidly to involve the thorax and arms in a sweater distribution but usually spares the hand, abdomen and feet. Systemic involvement, which is uncommon, may be marked by thickening of tongue, dysarthria, dysphagia, restriction of eye and joint movements, pleural, pericardial and peritoneal effusions (1-3). The disorder usually follow streptococcal infection, influenza, measles and mumps (1-7). The condition must be differentiated from dermatomyositis and scleroderma as both the conditions are progressive and carry poor prognosis as compared to scleredema.

Case Report

A 7-year female child presented with thickening of skin. It started from face progressing to neck, and then soon moved symmetrically downwards in

continuity involving chest (front and back), shoulders and extensor aspect of upper arms and proximal half of forearm over a period of three weeks. However, both the hands, feet, abdomen and genitalia were spared. The movements of mouth (difficulty in opening mouth) and neck were limited. There was history of fever and sore throat about 3 weeks prior to the episode. There was no history of itching, joint pains, dyspnea, dysphagia and rash. There was no history suggestive of diabetes. There was no significant family history of such ailment. The child had received topical steroids and oral antihistaminics. The child was afebrile, well nourished but had anxious look and mask like facial appearance.

The skin over face, neck, shoulders and extensor aspect of upper arm was involved symmetrically. It was hard, non-pitting and non-tender. It gave a rigid, wooden board like feel and could not be pinched. Involvement

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was diffuse with no sharp line of demarcation between involved and uninvolved skin. Face gave expressionless, mask like appearance with near obliteration of skin folds (especially nasolabial folds and forehead wrinkles) and with inability to smile and cry fully. There were no colour or temperature changes. There were no altered sensations, atrophy, ulceration, hair changes or nodules in the skin. Finger tips were soft without any evidence of scleroderma.

The hemoglobin was 11.2 gm%, TLC - 10,600/mm³ with polymorphs - 60, lymphocytes - 34 and eosinophils - 6 percent. The peripheral blood smear showed normocytic normochromic picture. ESR was 20 mm 1st hour. LFT/RFT were normal. Tuberculin test was negative. Chest skiagram and ECG did not reveal anything abnormal. CRP, rheumatoid factor and LE cells were negative. Most significant positive investigation was the serological evidence of recent streptococcal infection, i.e. elevated ASO titre (more than 400 Todd units). Skin biopsy taken from nape of neck revealed a normal epithelium with thick dermis with swollen collagen bundles separated by an increase in ground substance, and presence of sweat glands. No subcutaneous fatty tissue was discernible in biopsy (Fig. 1).

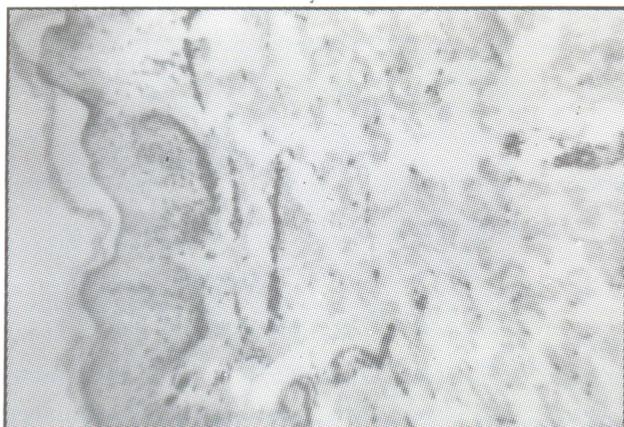


Fig. 1 Skin biopsy showing normal epithelium with thick dermis and swollen collagen bundles separated by an increase in ground substance.

The child was given procaine pencillin 4 lac units for 10 days and then switched on to long acting pencillin (benzathine pencillin) every month for 6 months. Local emollients and oral multivitamins were also given. There was no further progress and she started showing improvement with tendency towards gradual softening of skin symmetrically especially over extremities. By 6 weeks there was significant decrease in hardness and appearance of wrinkles and nasolabial grooves. Repeat ASO titre had come to normal.

Discussion

The name *Scleredema adultorum* is a misnomer since cases also occur in pediatric age group though uncommonly, there is no sclerosis and lastly skin induration does not result from accumulation of edema fluid but hyaluronic acid, an acid mucopolysaccharide and other glycosoaminoglycans in dermis (3, 7, 8). In majority the disorder is restricted to skin. Apart from cutaneous effects of this systemic illness, myocardial damage and cardiac arrhythmias, pleural and pericardial effusions have been also observed (1, 9, 10). Onset in diabetic patients may occur insidiously (11). Control of diabetes in such patients is difficult. Usually in 65-90% of cases, the disease follows an infection such as tonsillitis, pharyngitis, influenza, scarlet fever, measles, mumps, impetigo or cellulitis after an interval of days or weeks; most cases follow a streptococcal infection (1-5, 12). Trauma and tubercular lymphadenitis are also reported to the cause. A case following chickenpox has also been reported (2). In our case also, prior history of sore throat, fever and raised ASO titre made us to think the possible cause as streptococcal infection. There occurs immune sensitization with subsequent antigen antibody reaction possibly at the level of collagen fibril which by some unknown mechanism leads to disturbance of ground substance; probably the enzymes that participate in normal breakdown of hyaluronic acid are inhibited (11, 12). It is benign, self-limiting condition.

active phase of the disease persists for 2-8 weeks: spontaneous and complete resolution usually occurs in 6 month to 2 years. Recurrent attacks are unusual. The disorder must be differentiated from scleroderma, morphea, myxedema, trichnosis, dermatomyositis and subcutaneous fat necrosis (1). Considering the significant serological evidence of recent streptococcal infection we treated the child with pencillin as reported in earlier studies (6). The response to treatment was almost similar. Scleredema, though very rare in children should be kept in mind whenever child presents with thickening of skin.

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