Scleredema in a 6-Week-Old Baby

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Scleredema is rare connective tissue disorder which is extremely unusual in infancy. It is for this reason that we present here a 6-week-old baby with the disorder.

Case Report

A 6-week-old male infant, was admitted with the complaints of progressive hardening of the skin all over the body for 15 days and excessive vomiting (upto 3-4 times/day) for a month. The thickening of the skin was noticed first over the thighs and lower abdomen and rapidly progressed over 2-3 days to involve the whole body. There was no preceding history of any infective illness or fever.

On examination, the baby was alert and active, with a weight of 4.5 kg. The eyes appeared protuberant. Mild gynecomastia was evident (*Fig. 1*). The entire skin had a smooth waxy appearance and was firm to woody to feel, non pitting and non-pinchable. The palms and soles were also affected but to a lesser extent, while genitalia were spared. There were no telangiectasia, nodules, ulcers or any other skin lesion. Systemic examination was unremarkable.

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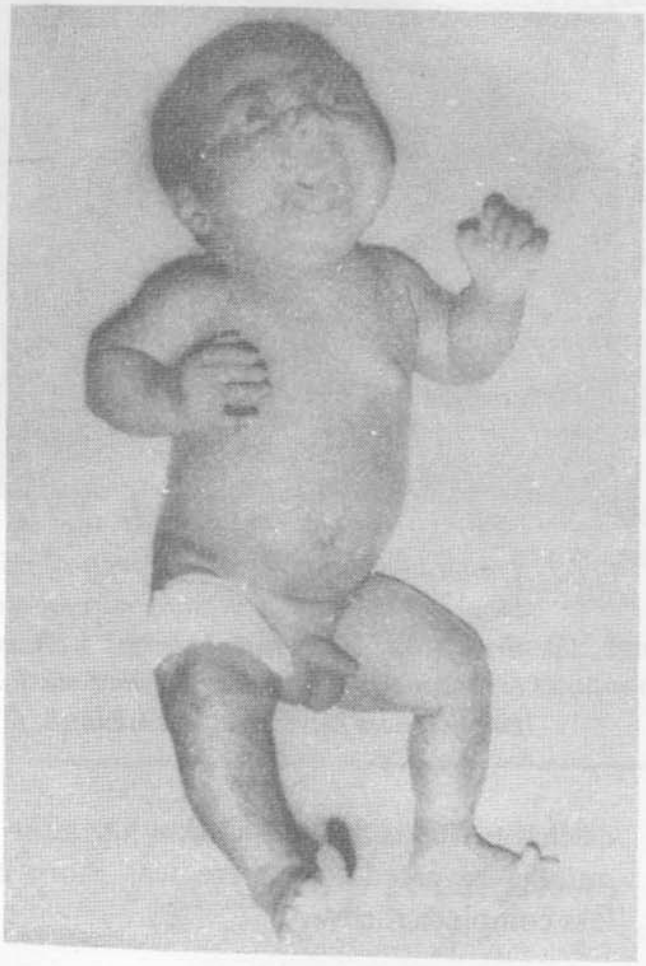

Fig. 1. Photograph of the infant.

Investigations revealed that hemoglobin and blood counts were within normal limits for age. Skin biopsy from the anterior thigh showed a normal epidermis but a thickened dermis with increase in collagenous tissue and slit like empty spaces between collagen bundles suggesting edema in the dermis. The sweat glands appeared normal (*Fig. 2*). Von Geison's staining for the demonstration of collagenous fibres confirmed increased collagenisation in the dermis.

The baby was put on beta-methasone drops for 1 week and then tapered off. He showed slight but definite improvement during this time but the condition then remained stationary till the time of

Fig. 2. Photomicrograph (H & E × 300) of skin biopsy of the patient showing normal epidermis but a thickened dermis with increase in collagenous tissue and slit like empty spaces between collagen bundles.

discharge 15 days later. Follow up after 4 months revealed that there was more or less complete recovery.

Discussion

Since the term scleredema was coined in 1900 less than 300 cases have been reported in the world literature(1). About half of these occurred in the first two decades of life(2). The major interest of this case lies in the fact that it occurred in one so young. To the best of our knowledge this is the youngest patient in the world literature. Pediatric cases have been reported from India also(3,4).

The onset of the disease is preceded in 65-90% of cases (in different series) by an acute febrile illness, usually an infection, and in about 60% of these the disease was probably streptococcal. The acute infection is followed by an asymptomatic period of 2-4 weeks before skin changes appear(2). No history of such a preceding illness was obtained in the present case.

The skin changes of scleredema begin commonly in the posterior neck and extend into the upper trunk, shoulders and proximal arms – "the sweater distribution". In exceptional cases the involvement may extend to the buttocks, abdomen and thighs or even be generalised, only the genitalia being spared(2). This was seen in the present case.

Various systemic pathologies may be associated with scleredema(5). The present patient had frequent vomiting which may or may not have been related to esophageal changes. Vomiting decreased within a week after admission.

The course of scleredema is that of complete resolution. Resolution generally beings 3-6 months after onset and is complete by 2 years(2). Robinow(6) noted that the disease is often less protracted in children. In two children described by him resolution started within a month of onset. Our patient also had a rapid onset of resolution and this was complete within 4 months.

A distinct subset of scleredema is that which occurs in the obese diabetic patient with severe vascular disease. Resolution in this group of patients is rare.

The major differential diagnoses of scleredema at this age include sclerema neonatorum and subcutaneous fat necrosis. Scleroderma has been described in a 2 year old. The course of the illness and the clinical appearance go against these diagnoses in our patient.

In the majority of cases, laboratory parameters are within normal limits, except for raised ASO titres in patients with preceding streptococcal infection(5). Biopsy shows a normal epidermis with markedly thickened dermis. Collagen bundles appear swollen, fragmented and separated from each other by clear spaces

considered by some to be interstitial edema and by others to be a mucopolysaccharide due to affinity for metachromatic stain. The dermal appendages are normal but owing to the thickening of the dermis appear to lie more superficially(5,6). In the mid and upper dermis some workers have found a mild perivascular inflammatory infiltrate. An electron microscopic study showed that the collagen fibrils of diseased skin were thinner than normal and showed a tendency to fractures and there was an excess of interfibrillary cement(5).

There are numerous theories about the etiopathogenesis of this condition. These include an immune mechanism, direct action of a bacterial toxin, and effect of adrenal steroids released due to infection(5). Circulating paraproteins have been found in patients with scleredema. Ohta *et al.* put forward the theory that these may enhance the synthesis of extracellular macromolecules by dermal fibroblasts, thus providing a mechanism for dermal fibrosis(7).

Although there are no controlled trials, therapy has been largely ineffective in this disorder. Bradford *et al.*(8) observed that children receiving steroids seemed to recover relatively quickly. This may be the cause for the rapid improvement in the present case also. Therapy is, however, not generally recommended because of the benign and self limited nature of the disorder.

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Intracranial Tuberculoma with Cutaneous Miliary Tuberculosis

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Tuberculomas constitute upto 15% of space occupying intracranial lesions in India(1). Tuberculomas result from heamtogenous spread from a primary

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