SCLEREDEMA WITH HAEMATURIA AND A RAISED ANTISTREPTOLYSIN TITRE—RECOVERY WITH CORTISONE

CASE REPORT

BY

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Scleredema, named scleriosis by Piffard (1876) and scleredema adultorum by Buschke (1902), is a rare disease characterized by stiffness of the upper layers of the skin commencing usually around the neck and following an acute infection. It differs chiefly from the acute diffuse form of scleroderma by having an excellent prognosis, the skin lesions usually disappearing completely. The condition was given the name scleredema adultorum (Buschke, 1902) to distinguish it from the scleremas of the newborn, but the name is perhaps unfortunate, as in the larger groups of case records analysed (Epstein, 1932; O'Leary, Waisman and Harrison, 1940) the largest numbers of patients were under the age of 10 years, two cases being reported at 2 months. The cause of the condition is unknown, but a history of previous acute infection is common. Laboratory investigations on previously reported cases have revealed nothing of significance.

The present case is reported because of the recurrence of haematuria with the onset of the skin manifestations, and the association of the clinical disease with an extremely high antistreptolysin in the child's serum.

Case Report

A boy aged 5½ years came under our observation in June 1960, with a history of aural discharge four weeks previously, subsequent malaise persisting up to the time of our examination. There had been frank blood in the urine, together with slight discomfort on micturition and diminished urine output two days before this examination. He appeared reasonably fit and had no pain or fever. There was a reddened throat and easily palpable lymph nodes in both anterior and posterior triangles of the neck. No abnormal physical signs were detected in the heart, lungs or abdomen. There was no oedema and the blood pressure was 86/58 mm. Hg. No hardening of the skin was detected at this examination. Urine microscopies of June 26, 1960, showed four to six red cells per high-

power field of centrifuged deposit. Albumin—less than 10 mg./100 ml.

The patient was admitted to hospital 10 days later as a possible case of acute nephritis, in view of the mother's report that the eyes had seemed puffy for the previous few hours. In other respects his general condition had been improving.

It was only after admission that the abnormal hardness of the skin over the face, neck, front and back chest, abdomen and arms was detected. The clinical impression was that of diffuse slight non-pitting oedema. The hands, feet, legs and lower part of the back presented no hardness and no distinct margin separated the affected and unaffected areas of skin. Indentation of the affected skin on pressure was greatly reduced. The skin could not be picked up between finger and thumb. The lower eyelid, even with considerable downward traction, could only be slightly everted. Normal creasing of the face was obliterated. The affected areas of skin were smooth and the feel was that of waxwork.

In the out-patient department, 10 days previously, the cervical glands had been easily palpable, whereas this was quite impossible after admission owing to the hardness of the overlying skin. There was no other abnormal clinical finding, the boy appearing reasonably well and lively. Fundi normal. Surprisingly there was no definite disability and muscle power seemed normal.

The following investigations were carried out.

Investigations. Antistreptolysin titre on first examination was over 1,200. This was the highest titre that we had seen in the laboratory, over 200 such investigations having been carried out annually and routinely for many years. The titre remained at this level for three weeks, subsequently falling over six months to a level of 200. Urine microscopy: the first urine examined contained fresh red cells in what are probably abnormal numbers, but no cellular casts were seen. Further examinations showed no haematuria.

Throat swabs, after admission, revealed Lancefield Group A, beta haemolytic streptococcus. E.S.R. was 46 mm. per hour on admission and subsequently falling.

Fig. 1.—Photomicrographs of collagen of the dermis from the case with scleredema (a) and from a similar site in the skin from a normal child of the same age (b). (Masson's Trichrome $\times 458$.)

C-reactive protein was negative. Serum cholesterol was 108 mg./100 ml.; albumin, 4.2 g./100 ml.; globulin, 3.4 g./100 ml.; calcium, 10.1 mg./100 ml.; phosphorus, 5.2 mg./100 ml.; alkaline phosphatase, 13.5 units (Jenner and Kay). Transaminase was 47 units; G.O. transaminase, 53 units. Blood counts showed leucocytes 12,000/c.mm.; polymorphs 65%; lymphocytes 35%. Mantoux was 1/1,000, negative. Chest radiograph was negative.

Biopsy of the affected skin from the child's left arm was carried out a fortnight after admission. To the naked eye, the skin appeared normal. Microscopically, no inflammatory change was seen, but there was a marked change in the collagen fibres. The individual collagen fibres were grossly thickened and had a clear inflated fibrillary appearance.

Fig. 1a shows the collagen fibres in the dermis from the case, compared with Fig. 1b which shows collagen fibres from a similar location from a child of the same age. The fibres in the child with scleredema have a fine feather-like appearance as if the collagen molecules are separated by the adsorption of the water. The interstitial fluid did not stain for mucin or amyloid (Weidman, 1932; Niebauer, 1960).

Progress. No improvement was observed until four weeks after day of admission to hospital. It was then appreciated that the hardness did not extend quite so far down the trunk and that the lower eyelid was a little more mobile to traction of the examining finger. Improvement continued and 12 weeks after admission the unusual hardness had practically disappeared and would

not have been detected unless specially searched for. A course of prednisolone had been given commencing two weeks after admission in doses of 40, 30, 15, 10, 5 and 2.5 mg. a day for two, two, two, two, two and one day respectively.

Discussion

The texture of the skin and the clinical course in this child were typical of scleredema adultorum. The microscopy findings of an apparent increased hydration with gross swelling of collagen strands was described in scleredema by Freund (1930). We found no perivascular changes as described by O'Leary *et al.* (1940). They describe the infiltrating cells as lymphocytes and fibroblasts, but Epstein (1932) illustrates microscopic sections from a case in which he describes the presence of such a cellular exudate which we find difficult to identify. The variability of histological findings that he and others note is not surprising. What is probably of more importance is that the lesion we have seen is apparently identical with the very earliest stage of scleroderma (Musso, 1954, 1959), which suggests to us that the distinction between the two conditions is of more importance clinically than pathologically, and scleredema may well be an early reversible form of scleroderma.

The particular interest of the present case lies in the association of the extremely high antistreptolysin

level. This level is considerably higher than is usually encountered in such streptococcal diseases as acute rheumatism, acute nephritis or erythema nodosum. This abnormally high antistreptolysin titre suggests that the child was reacting in an abnormal way to a streptococcal infection.

We have not noted any reports on the antistreptolysin titre level in cases of scleredema, and the present case suggests that such examination should be carried out.

This child was first diagnosed as acute nephritis. There had been frank blood in the urine and initial suppression of urine, and the first urine examined contained more red blood cells than we find normally by the clinical method carried out in this hospital. In the absence of a rise in the blood pressure and of cellular casts in the urine, this does not of itself constitute a diagnosis of nephritis, but does suggest that there is some renal lesion.

The child recovered completely with no residual skin or renal lesions, and this recovery was associated with steroid therapy.

Summary

A case of scleredema was described in a boy of 5½ years. The antistreptolysin titre was extremely high and there was frank haematuria at the onset of the disease. A possible relation between streptococcal infection and the scleredema is postulated.

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