SCLEREMA NEONATORUM: RECOVERY WITH CORTISONE

BY

IAN G. WICKES

From Tilbury and Riverside Hospital, Orsett Branch, Essex

(RECEIVED FOR PUBLICATION MAY 17, 1956)

Almost every paper about sclerema refers to the confusion surrounding the subject. This has arisen because the term has unfortunately been used to describe two very different conditions; the salient features of each are set out in the Table.

Thus the confusion is terminological rather than clinical. In reading the literature there is seldom any difficulty in classifying a case by its description as one or other condition, and the same has been my experience in half a dozen cases seen in clinical practice. Occasionally intermediate cases have presented difficulty and some atypical examples, such as the case described by Nicholson and Claireaux (1948) which appeared to be a form of generalized infective cellulitis in an older baby, defy classification. A third condition, characterized by localized pitting oedema in cold premature babies, is sometimes called scleroedema but this is not always easy to differentiate from fat necrosis. Scleroderma with sclerodactyly is a disease of the skin which probably does not occur in infancy.

The generalized disorder in young babies (to which the term sclerema neonatorum will be con-

fined here) was first recorded by Usenbenzius (1722) who described a newborn baby that was intensely cold and rigid. Underwood mentioned the condition in his book in 1784, but described it in full in a later edition (1819) under the heading 'skin bound'. He acknowledged assistance from Dr. Denman in making the following list of pathognomonic symptoms:

1st. The skin is always of a yellowish-white colour, giving the idea of soft wax.

2d. The feel of the skin and flesh is hard and resisting, but not oedematose.

3d. The cellular membrane is fixed in such a manner that the skin will not slide over the subjacent muscles; not even on the back of the hands, where it is usually very loose and pliable.

4th. This stricture often extends over the whole body; but the skin is peculiarly rigid in the parts about the face, and on the extremities.

5th. The child is always cold.

6th. The infant makes a peculiar moaning noise, which is often very feeble; and never cries like other children.

7th. Whatever number of days such children may survive, they always have the appearance of being dying.

TABLE
FEATURES OF TWO FORMS OF SCLEREMA

Synonyms	Acute sclerema Pre-agonal induration Cadaveric induration	Pseudo-sclerema Subcutaneous fat necrosis Subcutaneous fat sclerosis
Onset	Usually first week of life Occasionally later associated with diarrhoea and dehydration	First week of life
Distribution	Generalized (most marked in the legs; palms, soles and genitalia often spared)	Localized (circumscribed areas often symmetrically placed on the back, buttocks or shoulders)
Description	Normal appearing skin tightly bound down to underlying tissues restricting mobility of chest and joints	Discoloured skin overlying swollen plaque with irregular but definite margin; not deeply attached to underlying structures
Temperature control	Poor	Normal
Aetiology	Unknown	Birth trauma, ? infection, ? cold
Prognosis	Bad	Good
Histology	Normal, or slight increase in subcutaneous connective tissue	Gross destruction of normal picture with neutral fat crystals, giant cells, coarse trabeculation and sometimes calcification and cyst formation

Hughes and Hammond (1948), in an excellent review of the subject, collected 16 definite cases from the literature and added three of their own. In 16 the disease began in the first week of life and in this type of case the picture is less likely to be complicated by infections. Only five of the 19 cases survived. Unfortunately their attempt to clarify the nomenclature has not always been heeded. Brain (1953), in a clinical description of a case of fat necrosis under the heading 'sclerema neonatorum', perpetuates the confusion by stating:

'Pre-agonic or cadaveric induration is so obviously an ante-mortem solidification of the fat in a marasmic, moribund infant that it could scarcely be confused with sclerema neonatorum, except in the rare case when the latter disease involves the trunk and limbs, with in consequence, a serious prognosis.'

His predilection for the term 'pre-agonic induration' can hardly be justified now that the disorder has been shown to respond so well to treatment.

Case Report

After a normal pregnancy Mrs. N. gave birth without difficulty to her first baby with the assistance of a midwife at her home. The infant, a boy weighing about 6½ lb., was noticed to have rather stiff legs from the first. He was unable to suck from the breast but expressed breast milk was given with some success from a spoon. His stiffness, immobility and difficulty with feeding increased so that on the fifth day the family doctor (Dr. Kay) was called in. At his request the baby was seen by me on the following morning and immediately admitted with his mother to hospital.

On examination he was a thin, wizened, immobile baby with a weak, hoarse cry and with a subnormal temperature too low to record. He weighed only 5 lb. 2 oz. The skin over the whole body was bound tightly down to the subjacent tissues and could not be picked up from them. The knees and hips were flexed and could not be extended. The cheeks and lips felt woody and the chest expanded poorly. Fine crepitations were audible over both lung fields and the air entry was diminished. In short, the baby seemed certain to die but it was felt that nothing would be lost by the experimental administration of cortisone.

Therapy was started at once (aged 6 days) with cortisone 25 mg. t.d.s. and tetracycline 50 mg. b.d. given by tube. The baby was nursed in an oxygen box and fed with expressed breast milk by tube. Within a few hours of the first dose he seemed a little looser and thereafter steady improvement took place without interruption. The temperature (97° F.) was first recordable on the second day and became normal on the third. The face softened on the second day and the baby was able to suck from the breast for the first time though tube feeding was continued until the third day after admission. The arms and chest became normal by

about the fourth day but slight woodiness of the tissues of the thighs and calves persisted for about one week. Cortisone was reduced from 75 mg. to 50 mg. daily on the sixth day and subsequently the dose was gradually reduced until it was discontinued on the tenth. In all the baby received 570 mg. The daily dose of tetracycline was reduced concurrently and was discontinued at the same time. He was discharged on the twelfth day weighing 6 lb. 3 oz. partly breast fed. Full recovery had apparently taken place and no relapse occurred.

Discussion

At the time when this baby was seen the writer was unaware of the good reports of cortisone and corticotrophin therapy that had appeared in the American journals. The first was by Kendig and Toone (1951) who described a typical case in a premature baby. Treatment with oxygen and penicillin began on the third day but the condition worsened. Cortisone, 10 mg. t.d.s., was begun on the fourth day by mouth; a total of 80 mg. was given over a five-day period. Recovery evidently took place in 36 hours. A very similar case with the same dramatic outcome was reported by Kendall and Ledis (1952) who gave A.C.T.H., 5 mg. six-hourly, by injection. Williams (1953), also unaware of the previous reports, described a case successfully treated with A.C.T.H. but the subsequent progress was marred by superadded infections and the development of bilateral cataracts. Eisenoff, Aaron and Green (1954) reported two successful cases, one similar to those cited above in which the disease began in the first week of life and the other in which a 6-week-old baby developed diarrhoea, dehydration and generalized sclerema four days later. Likewise Buess (1955) reported three recoveries, one being a typical neonatal case and the other two were older infants suffering primarily from diarrhoea and dehydration. In the latter type it is difficult to assess the relative importance of fluids, antibiotics and corticotrophin in aiding recovery. Davis (1955) mentions four recoveries with cortisone or A.C.T.H. but gives details in only one, a newborn baby given intramuscular cortisone, 25 mg. daily.

There are no reports of failure of these hormones in this condition but there are descriptions of two cases in which the response to therapy was not dramatic in the way that is characteristic of the other recoveries. Søndergaard and Nielsen (1954) reported an atypical case of a baby aged 31 days whose indurated areas improved with A.C.T.H. (nine day course, total dosage 80 mg.) but which failed to disappear entirely after a second course (13 days, total dosage 130 mg.) and were still detectable seven months later though they finally resolved but the child was mentally defective. The other case

(Fauser, 1954) is stated to have been unaffected by A.C.T.H. but to have responded dramatically to cortisone; however, from the description fat necrosis seems to have been the condition and perhaps spontaneous recovery is the true explanation of the conflicting therapeutic results.

In sum, therefore, including the present report, since 1950 there have been seven dramatic recoveries in uncomplicated cases of generalized sclerema which was present at or developed a few days after birth, three recoveries in older babies suffering also from severe diarrhoea, five cases which, for various reasons, are difficult to assess and classify, and no failures.

It seems very probable that cortisone and corticotrophin have been mainly responsible for this excellent recovery rate in a hitherto very dangerous condition. Admittedly nearly all the cases also received antibiotics which no doubt played their part but nevertheless the improvement seemed to coincide with the hormonal therapy. Furthermore, in the period when these hormones have been available but not given deaths have still been recorded (Grotts, 1951; Rodriguez Fuentes, 1955). As already stated, Hughes and Hammond collected four reports of recoveries and added one of their own but in none could the response be described as dramatic with the possible exception of one treated by hot baths (Allyn and Marek, 1933) which recovered in ten days whereas in others the disease was protracted to several weeks. Other treatments claimed to have been successful include thyroid administration (Bourne, 1922) and blood transfusions (Lund, 1950).

The dosage of A.C.T.H. has been fairly uniform and this has been the most usual form of therapy. Cortisone has been given by injection in one case and by mouth in two. Kendig and Toone gave

only 80 mg. in all to a baby weighing 3 lb. 12 oz. whereas the total dosage in my case (weight 5 lb. 2 oz.) was 570 mg. No doubt this was unnecessarily large but the situation was desperate and no harmful effects were observed.

Summary

A case of generalized sclerema neonatorum is described which responded dramatically to oral cortisone.

In recent years there have been six other similar case reports of rapid recovery with A.C.T.H. or cortisone.

No failures with this form of therapy have been reported.

The term 'pre-agonal induration' is no longer appropriate.

My thanks are due to Sisters Hume and Holdsworth and to the ward nurses for their skilful care of the baby.

REFERENCES

- Allyn, W. E. and Marek, J. J. (1933). *Arch. Pediat.*, **50**, 59.
 Bourne, E. (1922). *Lancet*, **1**, 368.
 Brain, R. T. (1953). *Brit. J. Derm.*, **65**, 373.
 Buess, J. E. (1955). *J. Pediat.*, **46**, 224.
 Davis, D. W. (1955). *Neb. St. med. J.*, **40**, 243.
 Eisenoff, H. M., Aaron, H. A. and Green, F. C. (1954). *J. Amer. med. Ass.*, **155**, 905.
 Fauser, K. C. M. (1954). *Ned. T. Geneesk.*, **98**, 549.
 Grotts, B. F. (1951). *Arch. Pediat.*, **68**, 585.
 Hughes, W. E. and Hammond, M. L. (1948). *J. Pediat.*, **32**, 676.
 Kendall, N. and Ledis, S. (1952). *Amer. J. Dis. Child.*, **83**, 52.
 Kendig, E. L. and Toone, E. C. (1951). *Ibid.*, **81**, 771.
 Lund, O. (1950). *Nord. Med.*, **43**, 467.
 Nicholson, D. N. and Claireaux, A. E. (1948). *Archives of Disease in Childhood*, **23**, 290.
 Rodriguez Fuentes, C. (1955). *Rev. cubana Pediat.*, **27**, 293.
 Söndergaard, G. and Nielsen, J. P. (1954). *Acta paediat. (Uppsala)*, **43**, 289.
 Underwood, M. (1784). *A Treatise on the Diseases of Children*. London.
 — (1819). *Ibid.*, 7th ed.
 Usenbenzius, J. A. (1722). *Academiae Caesaræ-Leopoldinæ Carolinæ Naturæ Curiosorum Ephemerides*, Centuria, IX, p. 62.
 Williams, C. G. (1953). *S. Afr. med. J.*, **27**, 78.