INTRAPARTUM TOXIC EPIDERMAL NECROLYSIS

BY

W. P. SWEETNAM, A. J. E. BARLOW, and R. G. DENNISS

From The Huddersfield Royal Infirmary

(RECEIVED FOR PUBLICATION MARCH 18, 1964)

This syndrome was first described by Lyell (1956) in this country and by Lang and Walker (1956) in South Africa and has since been reported with increasing frequency. Beare (1962), in a comprehensive review, collected 35 cases from the published reports and described 10 further cases that he had observed personally. He suggests that it is unlikely that the syndrome could have been seen in the United Kingdom before 1950 because of its dramatic presentation and considerable mortality and considers that it is occurring more often. It is characterized by an increased skin tenderness and erythema affecting the smooth skin. The hairy parts are often not affected. This is followed within 12 hours by a sudden epidermal necrosis giving rise to blister formation and subsequent scaling. The epidermal separation is followed by a rapid healing of the affected areas. Occasionally burn-like scars are left. The condition has been described in both sexes, the youngest being 2 weeks and the oldest 85 years. The present case is of particular interest as the fully-developed lesions were present at birth.

Case Report

This was the mother's ninth pregnancy, the first six producing normal children and the seventh and eighth ending in miscarriages between the 10th and 14th weeks. Pregnancy was uneventful apart from anaemia. There were no clinical infections or known exposure to infectious illnesses. During pregnancy the mother was given some tablets of 'sedin' in the second month. Her anaemia was treated by injection of 'imferon' from the fifth month. She took liquid paraffin and phenolphthalein emulsion regularly three times weekly from the sixth month onwards. At the beginning of labour pentobarbitonum sodium ('nembutal') was given and half an hour before delivery 'pethidine' was administered. All these drugs had been given during previous pregnancies. The second stage took 40 minutes. As the left arm was being delivered anteriorly, the midwife noticed that the skin over the whole outer aspect of the left arm was loose, coming off on her fingers. She described the sensation 'like handling a macerated foetus'. When the child was delivered it was noticed that a similar smaller area was present on the outer side of the left hand and knee.

There were no blisters. The third stage of labour was normal and the membranes complete. The baby, in other ways a normal child of 8 lb. 3 oz. (3723 g.), cried immediately and showed no undue signs of distress. The condition of the baby was seen by the mother immediately after delivery and before division of the cord. The loose skin was removed and the glazed areas dressed with gentian violet (Fig. 1). The areas were healing well by the fourth day. At 14 days the affected areas showed superficial granulations (Fig. 2). The birth weight was regained by the tenth day and the general condition of the baby was excellent. Healing was complete in four weeks leaving scars which have since become keloidal (Fig. 3). The laboratory investigations carried out were necessarily limited but included a blood count and urinalysis, both of which were normal. The Wassermann reaction in mother and child was negative. No antibiotics or steroids were given.

Discussion

The baby was not seen by either author until 12 hours after birth when W.P.S. described the lesions in the notes, 'like a second degree burn one to two inches wide extending from the left shoulder to the wrist'. The condition during delivery was witnessed by two midwives and within a minute or two by the mother. An experienced nursing and medical staff had never seen a similar condition. The close resemblance to a scald is in accordance with Lyell's (1956) original description, and it is probable that the absence of blistering could be attributed to the intrauterine hydrostatic pressure which would be too great to allow blister formation to take place over so large an area. Alternative diagnoses such as 'collodion foetus', epidermolysis bullosa, the Stevens-Johnson syndrome, and other forms of pemphigus were excluded by the localization of the lesions and the subsequent progress in the absence of specific treatment.

Beare (1962) from his analysis concluded that the blistering or skin cleavage occurred within 12 hours of the first skin change, the erythema. In this instance the skin cleavage was present at birth. The first and second stages of labour took about 11 hours. It seems possible that the onset of uterine contrac-

FIG. 1.—Within a few hours of birth: the loose skin cut away and the raw areas painted with gentian violet.

FIG. 2.—At 2 weeks: the affected areas showing granulation tissue.

FIG. 3.—At 2 months: keloidal scarring which resembles the scarring following burns.

tions may have precipitated the syndrome; furthermore, the areas affected, i.e. the outer aspects of the left shoulder and arm and the left knee, were the non-hairy areas most subjected to pressure during the delivery.

Several authors have suggested that drug and possibly bacterial, fungal, or viral sensitivity can be the cause of this syndrome (Ruskin, 1948; Lyell, 1956; Lang and Walker, 1956, 1957; Kennedy, Daves, Henington, and Sternberg, 1957; Gill, 1958; Catto, 1959; Evans, 1959; Browne and Ridge, 1961. Rowell and Thompson (1961) also suggest the possibility of a toxic eruption due to a blood disease or metabolic disorder. There was no evidence in this case of infection in either the mother or child. However, during the last three months of intrauterine life the child was exposed regularly to phenolphthalein and at the beginning of labour to a barbiturate, both common causes of fixed drug reactions. Careful questioning revealed no evidence of any sensitivity reactions in the mother. It is generally accepted that active immunological mechanisms are not operative in the infant before or immediately after birth. It seems very improbable, therefore, that any such reaction could have occurred in the baby. Unfortunately the severity of the lesions made any question of confirming or refuting the

possibility of a drug sensitivity by exposing the baby to phenolphthalein or barbiturates, even in the form of a very limited patch test, quite unjustified.

Summary

A case of toxic epidermal necrolysis with the fully developed syndrome present at birth is described. The differential diagnosis and aetiology is discussed. In this instance a sensitivity reaction seemed unlikely. The aetiology remains obscure.

REFERENCES

- Beare, M. (1962). Toxic epidermal necrolysis. *Arch. Derm.*, **86**, 638.
- Browne, S. G., and Ridge, E. (1961). Toxic epidermal necrolysis. *Brit. med. J.*, **1**, 550.
- Catto, J. V. F. (1959). Toxic epidermal necrolysis occurring in a child. *ibid.*, **2**, 544.
- Evans, C. (1959). Toxic epidermal necrolysis. *ibid.*, **2**, 827.
- Gill, D. (1958). A case of toxic epidermal necrolysis (Lyell). *Brit. J. Derm.*, **70**, 302.
- Kennedy, C. B., Daves, F. H., Henington, V. M., and Sternberg, M. J. (1957). Dermatitis medicamentosa due to antipyrine. *Arch. Derm.*, **75**, 826.
- Lang, R., and Walker, J. (1956). An unusual bullous eruption. *S. Afr. med. J.*, **30**, 97.
- , — (1957). Toxic epidermal necrolysis. A report of four cases. *ibid.*, **31**, 713.
- Lyell, A. (1956). Toxic epidermal necrolysis: an eruption resembling scalding of the skin. *Brit. J. Derm.*, **68**, 355.
- Rowell, N. R., and Thompson, H. (1961). Toxic epidermal necrolysis in a patient with pulmonary aspergillosis. *ibid.*, **73**, 278.
- Ruskin, D. B. (1948). Fulminating dermatitis bullosa medicamentosa due to 'mesantoin'. *J. Amer. med. Ass.*, **137**, 1031.