

A CASE OF CONGENITAL ICHTHYOSIFORM ERYTHRODERMIA OF BROCQ TREATED BY HYPNOSIS

BY

A. A. MASON, M.B., B.S.

Senior Registrar, Queen Victoria Hospital, East Grinstead

The following case is of especial interest because the aetiology is unknown and the usual prognosis is that the condition is resistant to all forms of treatment. As a rule the course is progressive thickening of the skin from birth, reaching a maximum at about the age of 15 and then remaining static throughout life or deteriorating with secondary complications.

Case Report

The patient was a boy aged 16 who suffered from congenital ichthyosis. The lesion consisted of a black horny layer covering his entire body except his chest, neck, and face. The skin was papilliferous, each papilla projecting 2-6 mm. above the surface, and the papillae were separated from each other by only a very small distance, perhaps 1 mm. The papillae themselves varied in size from small thread-like projections on the abdomen, back, and flexor surfaces of the arms to large warty excrescences 5 mm. across on the feet, thighs, and palms. The small amount of skin which was visible between the papillae was also black, horny, and fissured. To the touch the skin felt as hard as a normal finger-nail, and was so inelastic that any attempt at bending resulted in a crack in the surface, which would then ooze blood-stained serum. In the skin flexures there were fissures which were constantly being reopened by movement and were chronically infected and painful. The ichthyosiform layer, when cut, was of the consistence of cartilage and was anaesthetic for a depth of several millimetres.

The condition varied in severity in different areas of the body, being worst on the hands, feet, thighs, and calves, and least on the upper arms, abdomen, and back. The skin on the face, neck, and chest appeared normal, although, as is shown later, it became papilliferous when transplanted to the palms.

The accompanying photographs show gradation of colour from light to very dark, but it must be emphasized that the light areas are only so in comparison with the grossly thickened and papilliferous darker areas. Even these apparently light areas were pathologically thickened, pigmented, and rigid.

No other evidence of congenital deformities was present. The blood picture and urine were completely normal. The skin of the mucous membranes appeared soft, elastic, and normal.

The patient's parents were alive and well, and there was no family history of ichthyosis. His birth was normal, after an uneventful pregnancy. His schooling was interrupted because the pupils and teachers objected to his smell. The patient himself disliked mixing with other pupils because of his distressing appearance. He was shy and lonely, but a willing worker and quick to respond to any teaching or affection shown to him, which had evidently been very little in the past. He had not had any other illness.

History of Present Disease

According to his mother, the patient was born with a thick skin, which became progressively thicker, darker, and more papilliferous throughout his life. He was seen as an infant at several London hospitals, but treatment was interrupted owing to the war and subsequent evacuation. After the war he received treatment at various hospitals, including

two London teaching hospitals. All treatment was of no avail, as might be expected from the natural history of the disease.

The patient's hands were thickly covered with a rigid horny casing which cracked, fissured, and became chronically infected. This not only rendered them useless for work, but caused him great discomfort. In an attempt to ameliorate this condition, the patient was referred to Mr. F. T. Moore, consulting plastic surgeon to the Queen Victoria Hospital, East Grinstead, who advised the grafting of fresh skin to the affected areas.

Treatment was begun on May 25, 1950, by Mr. Moore, who excised the ichthyosiform skin covering the palmar surfaces of both hands down to the palmar fascia, and covered the raw areas with split skin obtained from the apparently normal chest. At the time of the first dressing 10 days after the operation, the grafted skin was seen to be undergoing some change, and within one month it had itself become indistinguishable from the rest of the affected skin. Two months later a second attempt at grafting the palms was made after a radical diathermy excision. The result of the second operation was the same as the first, with the added complication of severe contractures of the fingers. The patient was seen by Sir Archibald McIndoe and other plastic surgeons, who agreed that further grafting operations were unlikely to be successful, and no alternative plastic procedure was possible.

Treatment by Hypnosis

On February 10, 1951, the patient was hypnotized and, under hypnosis, suggestion was made that the left arm would clear. (The suggestion was limited to the left arm so as to exclude the possibility of spontaneous resolution.)

About five days later the horny layer softened, became friable, and fell off. The skin underneath was slightly erythematous, but normal in texture and colour. From a black and armour-like casing, the skin became pink and soft within a few days. Improvement occurred first in the flexures and areas of friction, and later on the rest of the arm. The erythema faded in a few days. At the end of 10 days the arm was completely clear from shoulder to wrist.

A biopsy of affected skin on the calf was taken and sent for section to Professor H. A. Magnus, of King's College Hospital. This specimen was obtained under hypnosis, no anaesthetic being used.

Section Report.—"The specimen consists of a roughly elliptical piece of warty tissue measuring 1.5 by 0.7 cm. thick. Section shows a massive degree of hyperkeratosis, the average thickness of the keratin layer being 1.5 mm. The keratin is laminated and has produced many fissures in the underlying epidermis, which is atrophic and in places is thinly drawn out into the keratin. The papillae are elongated and narrow (alpine papillae). There are no other changes in the corium, apart from a marked deficiency in elastic fibre, only occasional fragments of which are present. One hair follicle and the somewhat atrophic duct of one sweat gland are present. This histological picture is similar to that described as being present in ichthyosis congenita."

The right arm was treated in the same way (see Figs. 1a and 1b), and ten days later the legs (see Figs. 2a and 2b; 3a and 3b) and trunk were treated.

Result

The improvement shown by the use of this treatment was as follows:

Region	Before Treatment	After Treatment
Hands	Completely covered	Palms clear. Fingers not greatly improved
Arms	80% covered	95% cleared
Back	Covered, but only lightly	90% "
Buttocks	Heavily covered	60% "
Thighs	Completely and heavily covered	70% "
Legs and feet	Completely and heavily covered	50% "

The accompanying photographs illustrate several of these areas. The table is based only upon clinical estimation of the condition, but in conjunction with the photographs it may serve to give a comprehensive picture of the result up to the time of writing.

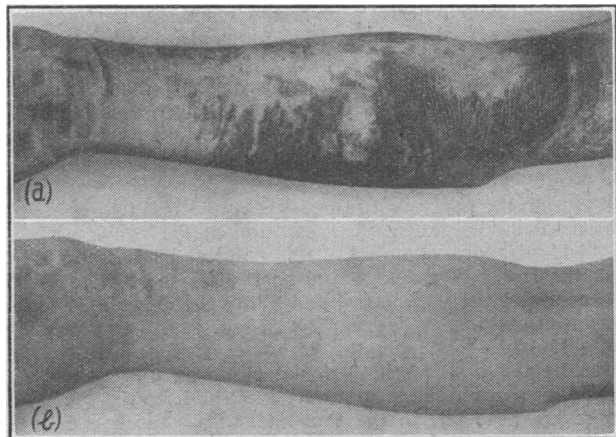


FIG. 1.—Right arm (a) before treatment; (b) eight days after treatment was begun, showing complete regression of ichthyosiform skin. (Left arm shows exactly the same picture.)



FIG. 2.—Legs seen from right (a) before treatment; (b) four weeks after treatment, showing complete regression of ichthyosiform skin in some areas and improvement in others. (The left sides of the legs show the same picture.)

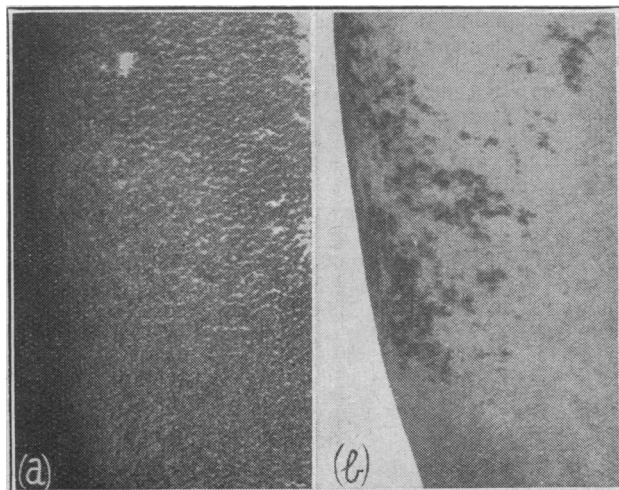


FIG. 3.—Skin of right thigh (a) before treatment; (b) one month after treatment was begun.

The improvement in the patient's mental state has been as dramatic as his organic improvement. Previously his schooling and social contact had been reduced to a minimum, his sensitivity towards his smell and appearance causing him to become lonely and solitary, with a hopeless attitude towards future friendship and employment. Now he has become a happy, normal boy, though still educationally retarded, and is already being employed as an electrician's assistant.

Whereas during the first few weeks clearance of the affected areas was rapid and dramatic, during the last few months there has been no appreciable change. There has, however, been no relapse of the improved areas over a period of one year.

Conclusions

From this response to hypnosis one of two inferences may be drawn. Either there is a hitherto unsuspected psychic factor in the aetiology of the disease or this is a case of a congenital organic condition being affected by a psychological process. A combination of both these factors is of course a third possibility. Whichever is true, the improvement in this case seems to be totally unprecedented, and was effected after the failure of all recognized methods of treatment.

I wish to thank Mr. F. T. Moore for permission to publish his case, and Mr. Gordon Clemetson for the photographs.

FATAL APLASTIC ANAEMIA AFTER CHLORAMPHENICOL TREATMENT

BY

L. A. HAWKINS, M.B., B.Chir., M.R.C.P., D.C.H.

AND

H. LEDERER, M.B., D.P.H.

(From the Royal Infirmary, Doncaster)

Two children living in the same town, but having little else in common, had whooping-cough treated by chloramphenicol in January and February, 1952, jaundice in March, and minor purpuric manifestations in May, ending in mid-June in the death of both from uncontrollable haemorrhages with an aplastic bone marrow. The close parallelism between the two cases led to the only known common factor, chloramphenicol, being suspected as the cause soon after the second patient was admitted to hospital.

A number of deaths from aplastic anaemia after treatment with chloramphenicol have been reported in the United States, and the literature was reviewed in a leading article in the *Journal of July 19, 1952 (p. 136)*. It seems highly probable that the aplastic anaemia in these two children was also due to chloramphenicol, and it is on this supposition that the cases are reported.

Case 1

A girl aged 4 years who was known to have been a contact of whooping-cough began to cough in the last week of January, 1952. On February 6 her doctor made a diagnosis of whooping-cough, and prescribed chloramphenicol palmitate, 250 mg. three times daily. This treatment was continued to a total of 20 g. in about 27 days. The cough persisted, but the child's general condition remained good. On March 16, 39 days after the start of treatment with chloramphenicol (C+39), she was listless and tired and generally unwell, she vomited, and the stools were noted to be pale