

Section of Dermatology

President C H Whittle MD

Meeting December 21 1961

Cases

Case for Diagnosis
? Acquired Epidermolysis Bullosa
 F Sherry-Dottridge MB

V M, female, aged 24

History: Admitted to hospital fourteen months ago with a history of the sudden appearance of very itchy small blisters round the umbilicus. She had been treated with penicillin powder for two weeks, but the irritation and rash persisted and became generalized. She was hysterical, crying and quarrelling with her mother. Her doctor's letter suggested she would be better apart from her family who were all emotional. Nikolsky's sign was not present. The general appearance suggested dermatitis herpetiformis, but sulphapyridine and later dapsone gave no relief. Prednisone 5 mg t.d.s. gave instant relief and all lesions cleared, but recurred as odd blisters on back, neck, toes, and mouth lesions, &c., wherever she accidentally rubbed or knocked herself. Attempts to decrease the dose to 5 mg b.d. always failed. Numerous milia on hands and neck noticed four months ago. No familial history.

Past history: Seen by psychiatrist for depression in 1957. Not a good mixer and has left several jobs because she said people thought her too slow. August 1959: Treated for ulcerative colitis and anæmia. Diarrhœa cleared on small doses of prednisone 5 mg daily. April 1960: Anæmia persisted in spite of iron by mouth. E.S.R. 40 mm in one hour (Westergren).

Present condition: Odd blisters on fingers, toes, back of hands, gluteal region, chest, following rubbing or injury. Milia on hands and side of neck and hair margin; some nails which were thickened and broken are becoming normal. Patient is more stable in her outlook. Her weight is increasing, with typical moonface. Blood count normal; film shows slight hypochromatosis, but no other abnormality.

The President: Do you accept the diagnosis, and do you think that steroids are going to help in this type of acquired epidermolysis? We had one case three or

four years ago where their effect was doubtful, but that was in an elderly woman (Whittle C H, Leahy J A & Davis R A, 1958, *Brit. J. Derm.* 70, 182).

Dr H T H Wilson: Has the urine been examined for porphyrins?

Dr F Sherry-Dottridge: Yes. Porphyrin was not found.

Dr E J Moynahan: Is she on barbiturates?

Dr Sherry-Dottridge: No.

The President: Do you find that these cases respond to steroids?

Dr G B Dowling: Regarding terminology, is the term 'acquired' quite correct, or are the cases that develop late only late manifestations of a latent disorder that has been present from birth?

Dr Louis Forman: A nursing sister of 44 years, developed blisters of the mouth, hands, feet and the buttocks. The blisters on the hands were provoked by sliding trauma, were sometimes hæmorrhagic, and left milia and obliteration of two finger nails. A very considerable measure of spontaneous recovery occurred over four years. During the last year of observation she was able to work and only had occasional small blisters in the mouth. There was no family history and this was considered to be a case of 'acquired' dystrophic epidermolysis.

The President: Do you accept the idea that the congenital and the acquired are the same disease?

Dr E J Moynahan: It depends upon what you mean by epidermolysis bullosa and what is meant by a congenital as opposed to an acquired defect. Strictly speaking, congenital means present at birth, and such defects may be either inheritable (genetic) or acquired *in utero*, when they originate as a result of developmental error or disease. Epidermolysis may well be due to a defective enzyme system concerned in the production of the cement substance between cells in the epidermis, and as such could be the result of a gene mutation, which is the usual event; alternatively, the enzyme system might become disordered as the result of some epigenetic event such as disease, and may therefore occur long after birth. Greying of the hair, for example, is usually genetically determined but it may be caused by epigenetic events such as exposure to radiation. It is obvious that in almost every event of this kind, either gene or subsequent 'accident' may produce the same result.