# FAMILIAL BENIGN CHRONIC PEMPHIGUS OF HAILEY AND HAILEY

By

K. C. KANDHARI,* L. K. BHUTANI,** G. H. HAJINI and Y. K. MALHOTRA,***

In 1939 Hailey and Hailey reported 4 cases of what they considered a new entity, and named it familial benign pemphigus. Since then many cases have been reported from different parts under different names but only 2 cases have been reported from India (Rajam et al, 1956; Kandhari and Gurmohan Singh, 1963). The following two cases are being further reported because of comparative rarity of this condition in this part of the world and an usually late onset of the disease in both these cases.

## CASE REPORTS

*Case I.*

V. W. 64 F was admitted in the All India Institute of Medical Sciences, New Delhi on 16th June, 1966 with a history of recurrent crops of itchy vesicular eruptions in axillae, groins, medial surface of thighs and submammary folds for past 15 years with attacks mostly in summer. She had 4 children, all of whom were well. There was no history of similar illness in the family.

Examination revealed an obese patient who had erythematous exuding lesions present in the groins, medial side of thighs, submammary folds and axillae. Mucous membranes were free. Systemic examination revealed only a slight enlargement of liver, which was firm on palpation.

*Laboratory investigations.*

Blood:-Hb 12 gms%; TLC 6300/cmm; DLC-P62, L34, E1, M3; ESR, 5mm/1st hour westergreen.

Urine:-Sugar 0.5% rest normal. Stools-normal. Blood urea-47 mg%; Liver function tests-total proteins: 6.8 gm%, Glob, 3.9% Serum alk. phosphatase-3 kA units, T. T.-5 units; Zn S04, turbidity-11 units; G. T. T revealed a hyperglycemic state, scrapings for fungus-negative. Skin Biopsy-The biopsy showed a large supra-abasal bulla in epidermis with broad and prominent dermal papillae at its base. The epidermal cells showed acantholysis with occasional dyskeratotic cells, The dermis showed oedema, vascular dilatation and perivascular lymphocytic infiltration (Fig 1).

*Case II.*

M. D. 75 yrs old widow admitted on 18th September, 1968 with a history of recurrent attacks of vesicular lesions in axillae, groins, submammary folds, sides of neck and occasionally other parts of trunk for past 23 years. She also complained of generalised itching. Lesions used to appear mostly in summer. Mucous membranes were always free. No family history of similar illness.

*Department of Dermatology & Venerology, All Institute of Medical Sciences, New Delhi-19.

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Local examination revealed flaccid grouped vesicular lesions together with exuding and crusted lesions and typical radiating fissures in axillae, groins, sides of neck and submammary region (Fig. II). A few vesicular lesions were also seen in other parts of body. Mucous membranes were unaffected. Exposed areas of body revealed poikilodermatous changes. She was hypertensive.

An intradermal injection of 0.1 ml of suspension of candida albicans in normal saline resulted in acantholysis.

*Investigations.*

Blood-Hb-11.5 gms; TLC-7500/cmm; DLC-P60, L32, M8; ESR-11mm/1st hour Westergren. Urine and stools-normal. Blood sugar-fasting 65 mg%. and 110 mg%. Blood urea-30 mg%, scrapings for fungus-negative, X-ray chest-nad Skin biopsy-The epidermis showed acantholysis with development of regular intra-epidermal clefts. Small dermal papillae projected into the larger clefts. Several dyskeratotic cells resembling grains were seen inside the clefts. The dermis was infiltrated by moderate number of lymphocytes (Fig. III).

She was treated with systemic tetracyclines, local corticosteroids and antimonilial preparations.

*Comments.*

The unusual feature about these two cases is the onset of disease late in life; the age of onset being 49 and 52 years respectively. Although the disease usually manifests first after puberty and most cases are young adults of either sex (Palmer and Perry 1962; Cullan, 1965), cases have been reported even in 6th (Jewell and Key, 1957) and 7th decade (Hurley and Cornelius, 1967). The first case from India reported by Rajam et al (1956) was a middle-aged female and second by Kandhari and Gurmohan Singh (1963) a boy who developed lesions at $2\frac{1}{2}$ years of age.

The absence of family history in some patients of benign familial pemphigus can be explained on the basis of its inheritance, being autosomal dominant with incomplete penetrance (Cram et al, 1967). Patients without family history have been reported by Rajam et al (1956) Jewell and Kay (1963) and Hurley and Cornelius (1967).

The disease, though an inherited disorder, is precipitated by a number of obviously unrelated factors. Lowenthal (1959) having successfully demonstrated acantholysis after surface inoculation of staph. aureus, was misled to believe that the disorder was an unusual form of pyoderma. Burns et al (1967) demonstrated that acanthoiysis could also be produced by topical application of candida albicans. Chorzelksi (1962) was able to induce typical lesions not only by bacterial inocula ion, but also by physical factors such as freezing, mild burns, chemical insults and allergy to adhesive tape, while Cram et al (1967) could induce acantholysis in such cases by ultra-violet rays.

Attempts at isolation of candida from the lesions were unsuccessful in both of our patients. Artificial production of acantholysis by intradermal injection of candida

in case II should therefore not suggest that C. albicans is the cause of the disease. Again broad spectrum antibiotics administered for long periods were without any effect on the lesions. These facts would tend not to favour an infective (Fungal or pyococcal) etiology for the disease, but would only fortify the thought that different forms of trauma could preciptate acantholysis in a genetically predisposed individual.

## SUMMARY

Two cases of familial benign chronic pemphigus, from India, are reported. Unusual late onset and absence of family history in both cases are discussed.

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