HAILEY-HAILEY DISEASE – CASE REPORT

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SUMMARY

Hailey-Hailey disease *or* ***Chronic Benign Familial Pemphigus*** *is probably rare in this region and to date there is no case report from Malaysia. I report here a Malay patient with this disease but with no family history and who was believed to be suffering from Atopic Eczema for several years. Dapsone effectively controlled the disease in this patient. The clinical features, histology and the therapy are discussed.*

INTRODUCTION

Benign Familial Pemphigus derives its name due to its histological similarity to Pemphigus Vulgaris. This disease, because of its chronicity, can be confused with many dermatoses particularly, *Pemphigus Vulgaris, Dariers Disease, Impetigo* and *Atopic Eczema* as the distribution of the rash in the flexures and intertrigenous regions of the body is also seen in all the diseases mentioned above.

CASE REPORT

A 50-year-old Malay patient who worked as a painter was seen in April, 1980 with moist, crusted lesions over the antecubital region, sides and back of neck, axilla, groin and back of knee for 15 years, associated with itch and pain.

The lesions started over the antecubital areas as a pruritic rash which became widespread. He could not remember whether there were blisters initially, but noticed that the lesion became exudative and crusted. He was diagnosed as Atopic Eczema but response to topical steroid therapy was negligible. There was no history of chronic skin disease in the family but the patient had Asthma between the age of 10 to 15 years.

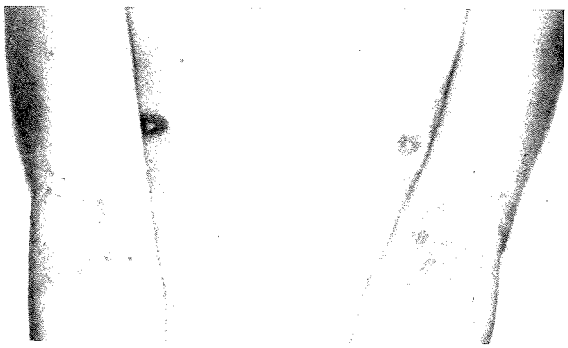

Fig. 1 This shows features of Hailey-Hailey Disease over the front of elbows with irregular, well demarcated 'serpigenous' border.

On examination, the lesions were hyperpigmented with irregular well-demarcated border especially at the antecubital region (Fig. 1). They were moist with fissures, secondary infection and candida. The skin adjacent to the lesions was normal. The lesions at back of the neck was slightly lichenified (Fig. 2 & 3).

In view of the unusual appearance and poor response to the accepted therapy of eczema which was the earlier diagnosis, a differential diagnosis of Chronic Benign Familial Pemphigus was made. A skin biopsy showed prominent intraepidermal separation, predominantly in suprabasal location. There was dyskeratosis, acantholysis and the formation of clefts above the basal cell layer. *Staph.*

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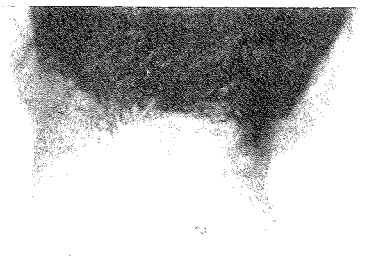

Fig. 2 Lichenified, Papular lesions back of neck.

Fig. 3 Moist Vegetations over the inguinal regions.

aureus was isolated from the lesion which was resistant to Tetracycline and Erythromycin. Moderate lymphocytes were present in the dermis. He was treated with Dapsone 100 mg daily and topical 1% Hydrocortisone. Cloxacillin was given to control infection.

The lesions became dry, infection cleared, itch and pain lessened. Four months later Dapsone was reduced to 50 mg daily as a maintenance dose. No relapse of the lesions noticed since.

DISCUSSION

Benign Familial Pemphigus which is an uncommon disease was described by Hailey brothers in 1939. [1] It derives its name from histological similarity to Pemphigus Vulgaris. Even though it is a familial disease only about 70% of patients have family history of this condition. [2] It is believed that a dominant gene is responsible and that one or more intermediate generations may not have the manifestations.

The aetiology of this disease may be due to a genetically — determined epidermal defect. The fault lies in the synthesis and maturation of *tonofilaments* and *desmosomes* resulting in an in-born tendency to *acantholysis* which can be precipitated by friction, freezing and infection amongst other factors. [3]

The disease presents with lesions on the flexural and intertrigenous areas of the body such as the neck, axilla groin, front of elbow and back of the knees. Occasionally the lesions can be widespread. Mucosal involvement is rare. Groups of small vesicles appear on an erythematous base containing clear fluid which later becomes turbid. The vesicles then rupture leaving erosions which become infected by candida or bacteria. The lesions spread peripherally with an active serpigenous border forming vesicles and crusts. The centre heals with either pigmentation or moist granular vegetation. Healing may take place without scarring. Itching and burning sensation are the common symptoms. Maceration and fissuring at the intertrigenous areas may give rise to pain. The disease usually starts at the second or third decades of life or even later. [5] Both sexes are equally affected.

Histologically Chronic Benign Pemphigus is characterised by suprabasal cleft formation with extensive loss of intercellular bridges — acantholysis. In spite of extensive acantholysis few remaining intercellular bridges still hold the cells of the detached epidermis loosely giving a *"dilapidated brick wall"* appearance. *Villi* — elongated papillae lined by a single layer of basal cells and *corps and ronds* — *Dyskeratotic cells* are also present. This condition has to be differentiated from Dariers disease, and Pemphigus Vulgaris. [6] In Dariers disease the suprabasal separation is usually smaller, acantholysis is less pronounced and dyskeratosis is more obvious. In Pemphigus Vulgaris the acantholysis is extensive, degeneration of the acantholysis cells is noted. Unlike Pemphigus Vulgaris, in Familial Benign Pemphigus there is no circulating antibodies against the epidermis on Indirect Immunoflurescent test and no tissue bound intraepidermal antibodies on Direct Immunoflurescent test.

The course of the disease is that of remission and exacerbation especially during warm condition. Remission can be long but not permanent. [7]

Treatment of this condition is with steroid — antibiotic cream or long term systemic antibiotic

like Tetracycline. [8] However, as in this patient, antibiotic treatment is ineffective and Dapsone controls the disease.

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