CHRONIC BENIGN PEMPHIGUS OF HAILY & HAILEY

By

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Chronic Benign Pemphigus (C.B.P.) was first described by Haily & Hailey (1939) as a familial benign chronic recurrent vesiculo-bullous disease involving the sides and nape of neck, axillae, inframammary and inguinal areas, histologically characterised by suprabasal lacunae and villi. It is generally accepted that C.B.P. is a genodermatosis, the predisposition to acantholysis being inherited as an irregular dominant trait. Many cases have been reported from the Western countries, but only a few cases have been reported in India (Rajam et al 1959, Kandhari & Gurmohan singh 1963).

A case which came under our observation is described and discussed here.

CASE REPORT

A male labourer aged 35 years reported in December 1964 with the complaint of recurrent itchy vesicular and bullous lesions on the neck of 3 years duration. There was no definite history of seasonal variations, but exposure to sunlight and sweating were stated to trigger the development of new lesions. These new lesions appeared mostly over the healed areas of previous lesions, but sometimes adjacent areas were also involved. There was no family history of similar skin disorder.

Examination revealed impetigo type of lesions–erthymatous patches of vesicular and bullous lesions intermingled with crusted areas over the nape and sides of neck extending as far as suprasternal fossa and a few isolated lesions on cheeks and sides of nose. The vesicles and bullae were flaccid, the new ones contained clear fluid and older ones contained turbid fluid. Nikolsky sign was absent. The healed areas showed slight hyperpigmentation.

Investigations–W.B.C. = 9000/c.mm. N = 58%, L = 38%, M = 4%,
E.S.R. = 18mm. first hr.

Culture of fluid from fresh bulla was sterile. Histopathological examination of a bulla revealed suprabasal lacunae with projecting villi and acantholytic cells. Scanty dermal infiltrate consisted mostly of lymphocytes.

Treatment and course, Wet compresses, topical steriod ointment and antihistmine tablets orally were administered. The lesions healed in about 10 days. The patient was kept under observation for a month during which period there was a relapse. The culture of fluid from a fresh bulla revealed coagulase negative staphylococci. In addition to the above treatment, vitamin A 1 lakh units were given intramuscularly on alternate days. The lesions healed in about 2 weeks and the patient was discharged. The patient reported again after 2 years. During this period of years, he had several minor relapses, but none as extensive as in December 1964. A few impetigo type

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of lesions were found on the nape of neck, The culture of fluid from fresh bulla revealed coagulase positive staphylococci. The lesions healed with wet compresses and topical tetracycline ointment.

DISCUSSION

The diagnosis of C. B. P. was suspected because of itchy impetigo type of lesions, sites, recurrence and sterile culture and confirmed by histopathological examination all features which confirm to the description of the disease by Hailey and Hailey except that there was no familial history. Cases without familial history have also been reported by Rajam et al (1956), Lyles et al (1958) and Kandhari and Gurmohan Singh (1963). Kandhari and Gurmohan Singh (1963) stated that the absence of family history in such cases may be due to genetic mutation. It is also possible that with an irregular dominant gene the abnormality may skip generations and a family history may not be available in all cases.

As regards the seasonsal variations, cases with relapses in summer (Jewell and Key 1957) and in winter (Jewell and Key 1957, Kandhari and Gurmohan Singh 1963) have been reported. Our patient did not notice any definite seasonal variations probably because the temperature variations during different seasons are not marked in this place.

Lowenthal (1959) stated that C. B. P. is a manifestation of pyoderma in a person genetically predisposed to acantholysis. Kandhari and Gurmohan Singh (1963) while conceding that bacteria may play a role in precipitating the lesions, felt that there is insufficient evidence to say that the disease is a manifestation of pyoderma The following are the arguments for and against this view of pyoderma.

FOR	AGAINST
Clinical resemblance to Pyoderma.	Cases without clinical resemblance to pyoderma have been reported (Lyles etal 1958).
Culture of pathogenic organisms.	Sterile culture or culture of non pathogenic organisms.
Gradual improvement with advancing years indicating increasing immunity to organisms.	Relapses and remissions.
Response to antibiotics.	Failure of antibiotics and response to Vitmin A
	Lymphocytic infiltration in the lesions.

In our patient, the clinical picture resembled that of impetigo, culture reports were variable, there were relapses and remissions, and also there was decrease in the severity of lesions with the passage of time and dermal infiltrate was mostly lymphocytic. The fact that culture reports have been variable indicates that besides pyococci. sunlight, sweating and friction may also trigger off new lesions at different times. Decrease in severity of lesions with the passage of time may not necessarity be due to increasing immunity against pyococci, because such an event is also observed in other genodermatoses such as epidermolysis bullosa and ichthyosis. Hence we

agree with Kandhari and Gurmohan Singh (1963) that there is in-sufficient evidence to label, C. B. P. a pyoderma. We would like to state that it is a genodermatosis in which physical, mechanical and bacteriologic factors precipitate the development of clinical lesions, a fact which is borne out by the experimental evidence of Chorzelski (1962).

SUMMARY

A case of C. B. P. without familial history and without seasonal variations is described. The disease was considered to be a genodermatosis in which physical, mechanical & bacteriologic factors precipitate the development of clinical lesions.

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