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CASE REPORT

A RECURRENT INTERTRIGO: BENIGN FAMILIAL CHRONIC PEMPHIGUS

Niraj Parajuli[1]

[1]Department of dermatology and venereology, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal.

***Correspondence to:** Dr. Niraj Parajuli, Department of dermatology and venereology, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal.*
Email: drnirajparajuli@gmail.com

ABSTRACT

Benign familial chronic pemphigus (BFCP) is an autosomal dominant disorder characterized by recurrent vesicles and plaques mostly over the intertriginous areas. The defect is in the ATP2C1 gene which leads to suprabasilar acantholysis. Pregnancy, physical trauma, excessive sweating, skin infections and exposure to ultraviolet radiation are the important trigger factors. Self-medications or over the counter treatment usually makes a delay in the proper diagnosis.

Key words: acantholysis, benign familial pemphigus, genetic, hailey-hailey, intertrigo.

INTRODUCTION

Benign familial chronic pemphigus (BFCP) or Hailey–Hailey disease (HHD) is an autosomal dominant disorder.[1] It is characterized by recurrent vesicles and plaques mostly over the intertriginous areas. The defect is in the ATP2C1 gene leading to suprabasilar acantholysis.[2]

Pregnancy, physical trauma, excessive sweating, skin infections and exposure to ultraviolet radiation are the important trigger factors.[3]

Treatment include preventive measures like personal hygiene, clothing and lifestyle modification should be opted.[1] Most common treatment modalities include topical corticosteroids as well as topical and oral anti-infective agents.[4]

Most cases of Hailey-hailey disease are being misdiagnosed and treated as fungal intertrigo.

CASE REPORT:

A 33 years old male came to dermatology out-patient department (OPD) with complaints of recurrent crusted plaques on the neck and bilateral axilla for the last several years. Lesions flare-up were noted during the summer months. Only mild pruritus and burning sensation accompany the lesion. Most of the time the lesion would resolve on its own. Sometimes patient would use some over-the-counter medications to get rid of it. No history of fever, sore throat, jaundice or medications prior to the eruption.

Patient claimed that similar problem was also present in one of his sister.

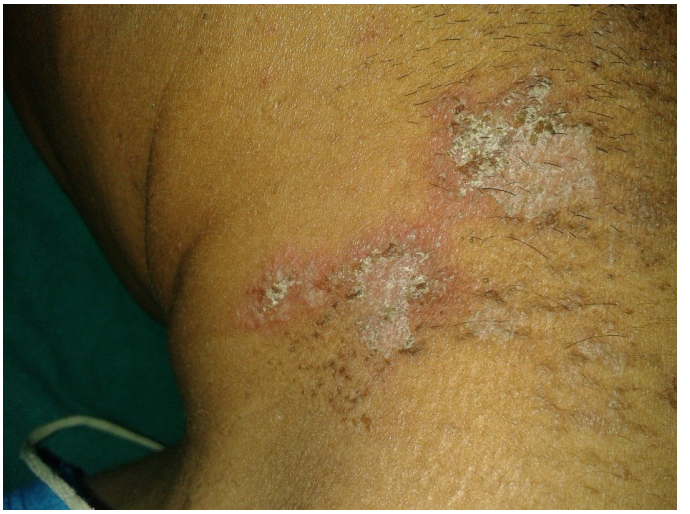
On examination, there were bilaterally symmetrical erythematous crusted plaques on axilla and neck (Figure 1a and 1b). Few studded pustules and vesicles were present over the periphery of the plaques. Nikolsky's sign was negative.

No nails, hairs or mucosal involvement.

A complete blood count, random blood sugar and routine urine analysis were all within normal limits. Skin biopsy was performed from the lesion skin which showed intraepithelial clefting with dilapidated brick appearance (Figure 2a and 2b).

So, a diagnosis of benign familial chronic pemphigus or Hailey- hailey disease was reached. Patient was explained on the nature of the disease and preventive measures.

A combination of steroid-antibiotic cream was prescribed. Patient was advised for regular follow-ups.



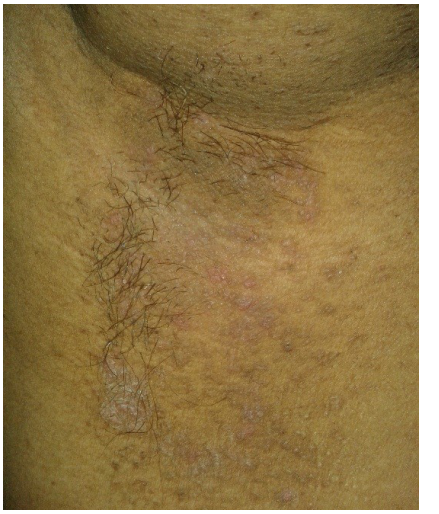


Figure 1a and 1b: Erythematous crusted plaques over nape and right axilla.

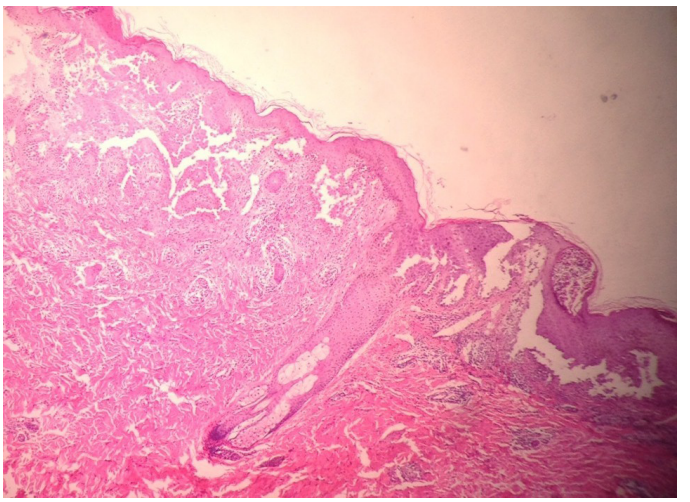


Figure 2a: Histopathology of the skin showing suprabasal acantholysis.
(10x magnification, hematoxylin and eosin stain)

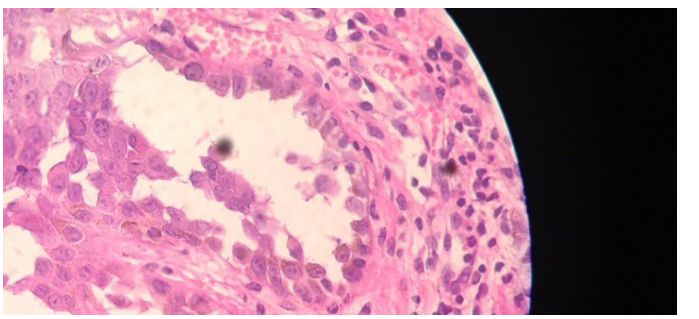


Fig 2b: A typical "dilapidated brick wall" appearance.
(40x magnification, hematoxylin and eosin stain

DISCUSSION & CONCLUSION:

BFCP is a rare chronic recurrent eruption, caused by defect in gene ATP2C2. It is characterized by recurrent crusted plaques or blister over the intertriginous region.[2]

A study conducted by Burge in United Kingdom showed that the most common sites for HHD were flexures, which included axilla, groin, perineum, inflammatory region. Other sites including neck, shoulder, chest

arms and trunk were less frequently involved.[4]

The diagnosis can be usually made by skin biopsy. Histopathologic changes seen include intraepidermal bullae along with suprabasal acantholysis. A partial acantholysis gives a typical appearance of dilapidated brick.[3]

Most of the cases are best treated with topical steroids and topical antimicrobials. But, in some difficult and refractory cases oral antibiotics, excisional procedures as well as botulinum toxin A have shown good results.[5]

Typically, these cases are usually treated as dermatitis or fungal infection by over-the-counter medications and usually takes a long period of time before being properly diagnosed.

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