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AN ELDERLY WOMAN WITH RECURRENT VESICO-BULLOUS &CRUSTED LESIONS IN FLEXURAL PARTS OF THE BODY: A CASE REPORT ON HAILEY-HAILEY DISEASE IN BANGLADESH

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ABSTARCT

Hailey- Hailey Disease (HHD), a rare genetic skin disorder, is inherited as autosomal dominant pattern. Abnormal epithelial cell adhesion due to altered calcium metabolism is the main pathology of the disease. Persistently recurrent, pruritic, painful, flaccid vesicles, bulla, pustules with crusted and erosive plaques along with maceration and fissuring are the clinically characteristic features of the disease which involve the neck, axilla, flexures and groins. the onset of the disease is common in late tensor early twenties. Characteristic clinical presentation and histopathological findings of "dilapidated brick wall"– are the aids to establish the diagnosis. Here we report a case of a 59-year-old woman with recurrent attack with the features consistent with HHD with positive family history and specific histopathological findings helped us to reach the final diagnosis.

Key Words: Hailey-Hailey Disease, Autosomal dominant disease, Flaccid vesicles, Histopathology, Dilapidated brick wall.

INTRODUCTION

Hailey-Hailey disease (HHD), also known as familial benign chronic pemphigus, was first described by two brothers William Howard and Hugh Edward Hailey in 1939[1]. It is inherited as an autosomal dominant pattern affecting around 1/50000 of people equally both male and female[2]. HHD clinically appear as recurrent, pruritic, painful, vesico-pustules, erosions, fissuring and erythematous plaques with maceration and malodour in the flexural parts of the body. The typical onset of the disease is seen during early twenties to elderly, it can occur at any age[3,4]. The main genetic defect is the alteration of the intracellular calcium gradient in the Golgi complex due to mutation in the ATP2C1 gene located in chromosome 3q22.1 resulting in to acantholysis of the epidermis[5]. Ultraviolet radiation, heat, sweat, friction, bacterial and fungal infections are some factors those can aggravate the disease condition. As the disease is relapsing and remitting in nature, it produces discomfort, physical inactivity, impairs patient's quality of life significantly.

Here we report a case of an elderly patient presented with multiple vesico-bulla and vesico-pustules in different folds of the body with crusting and erosions resembling eczema for 1 year. Late onset of the disease with some deviation from clinical presentation caused diagnostic dilemma and made a challenge for us to diagnose properly and report the case.

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CASE REPORT

A 59-year-old woman presented to outpatient department as she was suffering from multiple, vesicles and bulla, some of them were pustules in flexural parts of the body for last 1 year associated with remission and recurrent summer exhibition and fever for 5 days.The fluid-filled vesicles initially appeared on the folds of the neck, antecubital fossa which gradually spread in axillary, sub-mammary folds and groins bilaterally. The blisters ruptured spontaneously within 1-2 days leaving weeping, eroded and crusted surfaces. The patient had the complaints of severe itching associated with stinging and burning sensation and also malodorous discharge from the lesions which caused discomfort to the patient. At that time after using some topical preparations, oral antihistamines the lesions were resolved. After 1 year, the similar cutaneous features developed with fever. Her brother had same kind of disease in family.

On examination, there were multiple, vesico-bulla and vesico-pustules distributed in folds of neck, antecubital fossa, inframammary and axillary folds, groins. Some of the lesions were ruptured having discharge, erosions and crusting resembling eczema. Nikolsky and Asboe-Hansen signs could not be elicited. Palms, soles, mucous membranes, nails and hair were not involved (Figure 1). Other systemic examinations revealed normal findings. With above mentioned history and clinical findings, we decided to do skin biopsy for histopathology considering the differentials of flexural eczema, HHD, Darier disease and Grover disease.

Outcomes of routine laboratory investigations including complete blood count, liver function test, renal function tests were within normal range. Skin biopsy for histopathology showed epidermal hyperplasia with suprabasilar and intraepidermal clefting. It also revealed acantholoysis of keratinocytes resembling dilapidated brick wall. The dermis showed mild chronic inflammatory infiltrate.In Figure 2sections show skin with an epidermis demonstrating prominent intraepidermal (predominantly suprabasal) acantholysis with formation of multiple clefts/lacunae. The acantholysis was extensive and multilayered, producing the characteristic “dilapidated brick wall” appearance. The basal layer was relatively preserved along the dermo-epidermal junction.The epidermis showed variable acanthosis with surface hyperkeratosis (with focal parakeratosis, if applicable). Dyskeratosis was not a prominent feature.The superficial dermis showed mild perivascular chronic inflammatory infiltrate, with focal exocytosis; no vasculitis or malignant atypia was seen.All these findings of histopathology are consistent with Hailey-Hailey Disease. Thus we reached to final diagnosis.

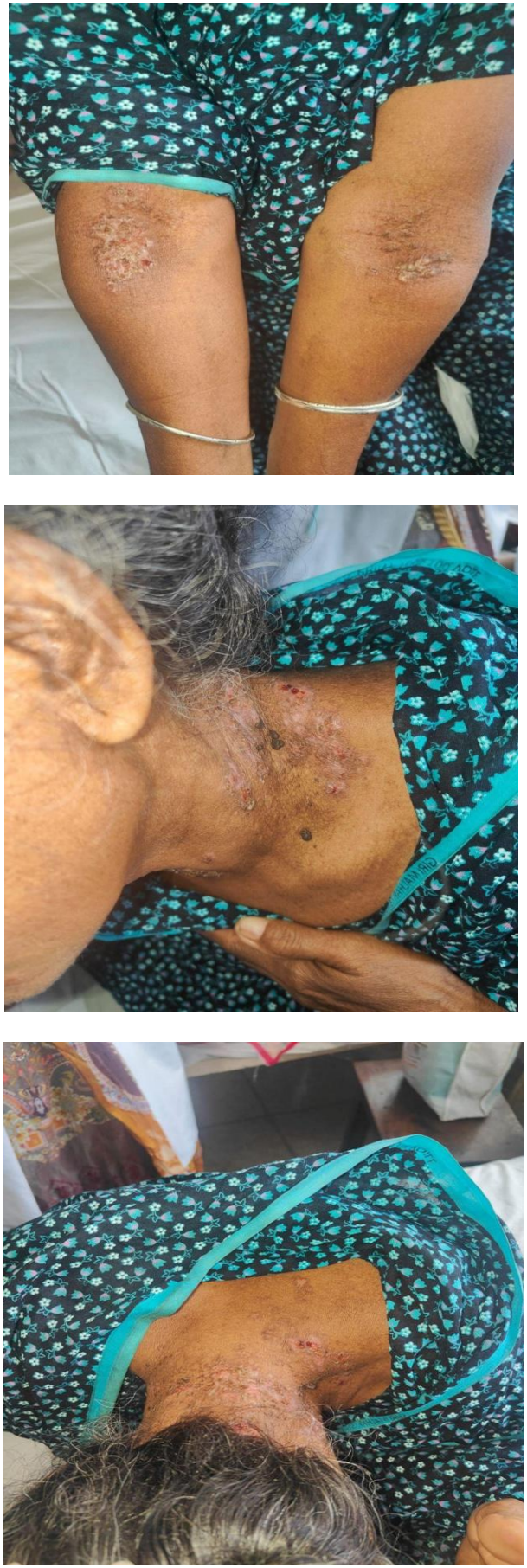

Figure 1: Multiple blisters with crusting and erosions

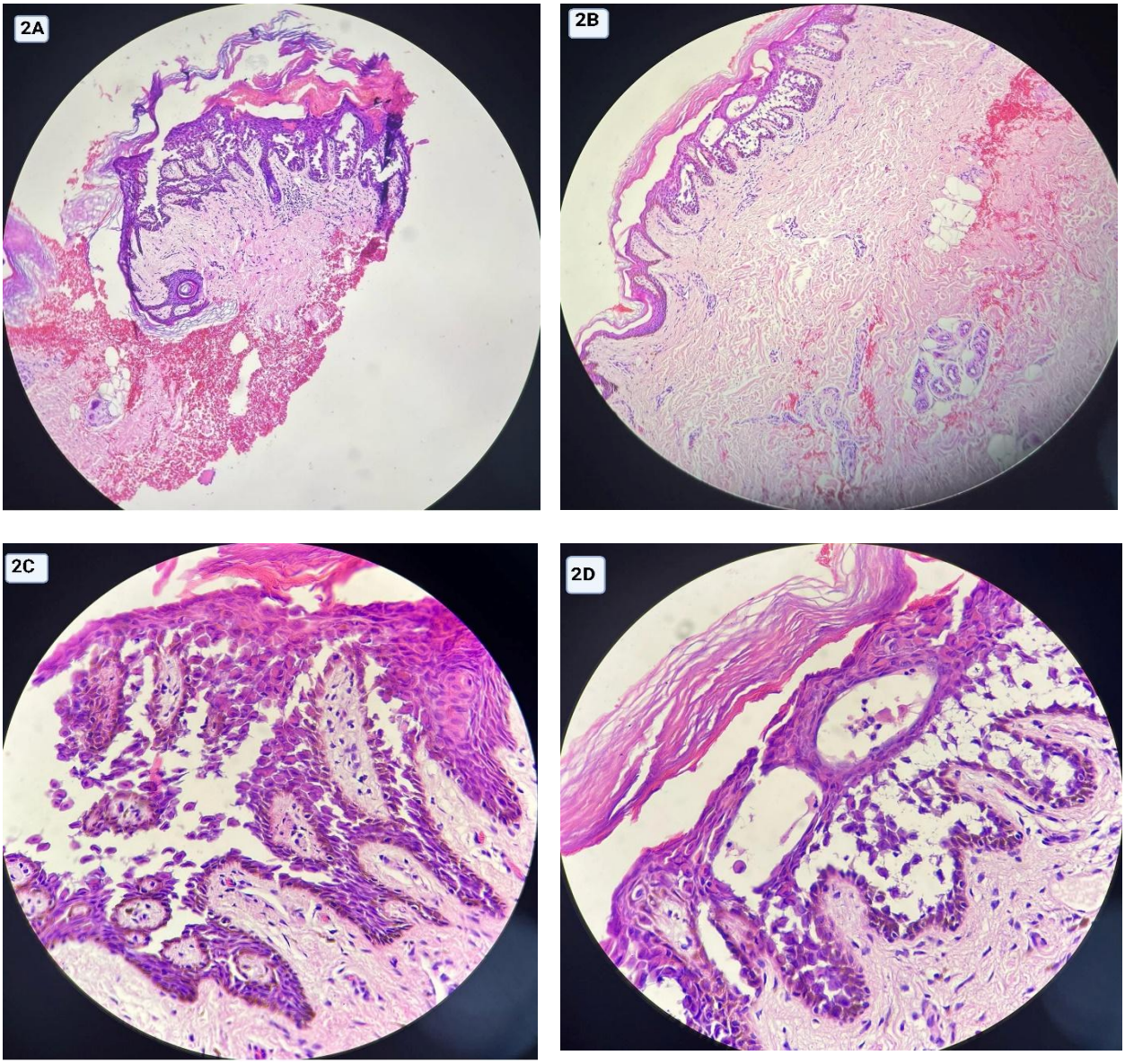

Figure 2 : 2A :Photomicrograph showing suprabasal acantholysis of the epidermis with a characteristic dilapidated brick-wall appearance, consistent with Hailey–Hailey disease (H&E, ×100).2B:Photomicrograph showing suprabasal acantholysis of the epidermis with partial loss of intercellular cohesion producing a dilapidated brick-wall appearance, consistent with Hailey–Hailey disease (H&E, ×100).2C and 2D:Suprabasal acantholysis with dilapidated brick-wall pattern (H&E, ×400).

DISCUSSION

Hailey- Hailey disease or familial benign chronic pemphigus is a rare autosomal dominant genodermatosis named after Hailey brothers in 1939. It occurs due to mutation in the ATP2C1 gene located in chromosome 3 which encodes Golgi-associated Ca^{2+} ATPase resulting in dysfunctional intracellular Ca^{2+} signaling with downstream effects causing impaired keratinocyte adhesion and acantholysis in the stratum spinosum[1]. The approximate incidence of the disease is about 1 in 50000 individuals with no age, gender and racial predilection[7]. Usually it develops in early adulthood, but it can appear at any age[4,7]. Clinically fluid- filled blisters which rupture spontaneously forming weeping erosions, crusting and fissures in an inverse

distribution on the neck, axilla, antecubital fossa, inguinal folds are the characteristic features which can spread to trunk, inframammary, perineal areas[1]. Primarily flaccid vesicles arising on an erythematous macular base which tend to spread peripherally with vesiculation along the advancing serpiginous border[1] and form large, vegetative, malodorous plaques with crusting, erosions and painful fissures as secondary changes[1,5]. Pruritus, stinging, foul smelling discharge with post inflammatory hyperpigmentation impact the quality of life of the patients. There may be longitudinal white lines on finger nails. some triggering factors like skin trauma, friction, sweating, sunburn, bacterial and fungal infection or any other cutaneous disorder can exacerbate the disease condition[8].

The diagnosis of HHD is often delayed as it simulates the other common dermatoses such as eczema, tinea, impetigo, contact dermatitis, intertrigo, pemphigus vulgaris or Darier disease[2,8]. In our case, an elderly patient presented with multiple, recurrent, pruritic, foul smelling, vesico-bullous and vesico-pustular eruptions in the folds of the neck, antecubital fossa, sub-mammary folds, groins for last 1 year which aggravated during hot weather and with rise of body temperature. When the blisters ruptured, eroded surface with crusting overlied the lesions which simulate eczematous dermatitis. So for such type of presentation, skin biopsy for histopathology was done which revealed epidermal hyperplasia with suprabasilar and intraepidermal clefting with acantholoysis of keratinocytes resembling dilapidated brick wall. The dermis showed mild chronic inflammatory infiltrate. These histological features are compatible with the diagnosis of Hailey-Hailey disease.Counselling about the disease process and prognosis was done properly, treatment was given with oral antibiotics, anti-histamines, topical antibiotics and steroids. The patient came for follow upregularly in 3 months interval and her symptoms was relieved partially.

CONCLUSION

Though HHD is a benign genetic disorder, it is difficult to cure completely for the physicians. Despite having characteristic clinical presentations, sometimes it is misdiagnosed due to similarities with other common skin diseases for which the clinicians should keep in mind HHD as a differential diagnosis in their daily practice. Furthermore, the chronic and recalcitrant nature of HHD impacts the patient's quality of life. So, the aim of treatment is to counsel the patients, control symptoms, reduce the recurrence and follow up regularly.

CONFLICT OF INTEREST

There is no conflict of interest.

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