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Hailey Hailey disease: A Presentation out of the Ordinary

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Abstract

Observation: Hailey–Hailey disease (HHD) or benign familial pemphigus is a familial vesiculobullous dermatosis affecting the intertriginous areas with summer aggravation. We report here a rare case of HHD in a 25-year-old woman presenting with persistent crusted erosions on the face for 12 years.

Introduction

Hailey–Hailey disease (HHD) is a blistering dermatosis, inherited as an autosomal dominant trait with incomplete penetrance and without any sex predilection. The onset is often delayed until the second or third decade. Approximately two-thirds of the patients with HHD have a positive family history. Clinically characterized by crusted erosions affecting the axillary, genitocrural and inframammary folds, the lesions begin with vesicles and bullae, which are soon eroded and crusted and spread peripherally with serpiginous borders [1].

A few instances of mucosal lesions of the mouth, the labia majora and the oesophagus have been reported [2]. Lesions are typically symmetric although post-zygotic loss of gene function can result in asymmetric type 1 or segmental type 2 disease [3]. The clinical course is unpredictable and individualized. We report here a case of HHD localized to the face for the rarity of the occurrence.

Case Report

A 25-year-old woman presented to us with a non-pruritic, painful erosion on the left malar area for the duration of 12 years. There was a history of recurrent eruption of small blisters that increased in size gradually, then ruptured forming a crusted erosion. The lesion had a waxing and waning course but never healed completely. She reported worsening of this lesion during summer months. There was no history of photosensitivity. There was no history of similar illness in her family. Her past medical history was unremarkable. She was treated with various topical medications details of which could not be elicited.

Clinical examination revealed a tender irregular erythematous crusted erosion of 2X3 cm size over

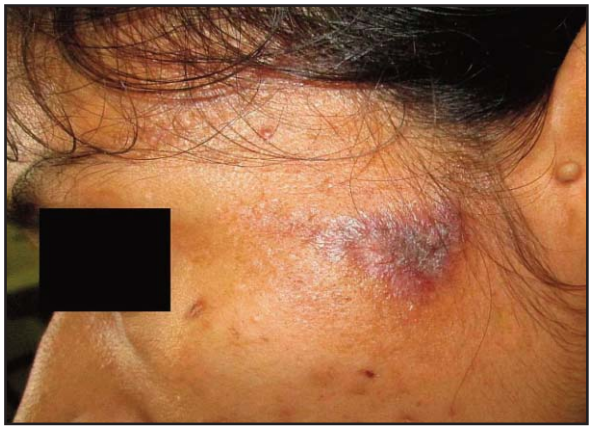

Figure 1. Erythematous crusted erosion on left malar area.

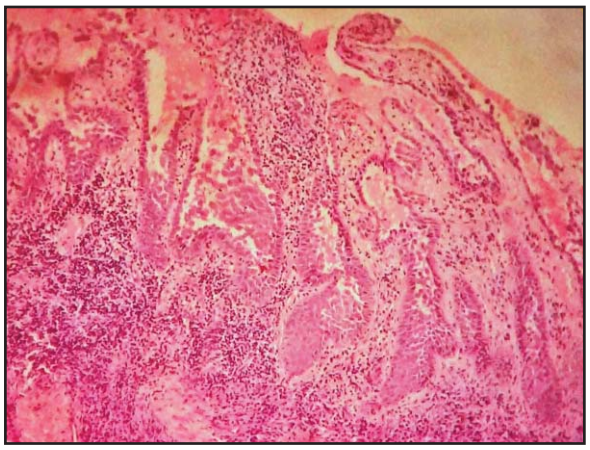

Figure 2. Histopathology showing intraepidermal clefts with partially detached acantholytic cells, appearing as "dilapidated brick wall" and an upper dermal inflammatory infiltrate. (H&E X 100)

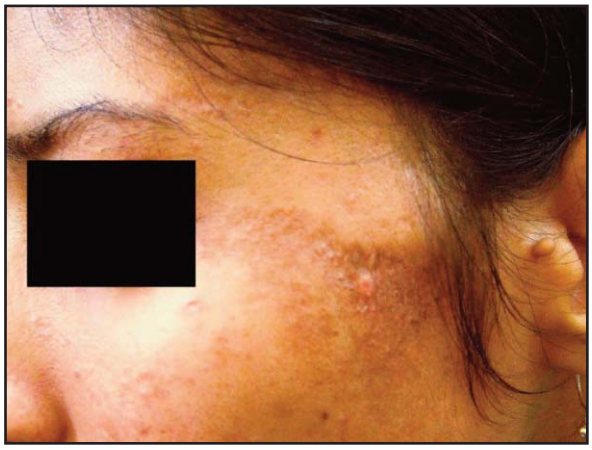

Figure 3. Showing healing of lesion after three weeks.

the left malar region. Other areas of the skin, the hair, nails, and oral mucosa were free. There was no regional lymphadenopathy. Routine laboratory tests including serum chemistry revealed no abnormality. Bacteriological study of the serous fluid had negative results.

Histopathological examination showed intraepidermal cleft with partially detached acantholytic keratinocytes, appearing as "dilapidated brick wall" (**Figure 2**). No dyskeratosis was noted. The epidermal surface showed evidence of crusting and the upper dermis showed a mixed neutrophilic and lymphocytic infiltrate. The subcutis was normal. Direct immunofluorescence was negative.

The patient was treated with oral amoxicillin and topical mupirocin which showed good clinical response within a couple of weeks (**Figure 3**).

Discussion

Hailey–Hailey disease is caused by mutations of ATP2C1 encoding the human secretory pathway Ca/Mn+-ATPase isoform 1 (hSPCA1) on the Golgi membrane [**4**], and insufficiency of it results in acantholysis.

Aggravating factors for HHD include sweating, exposure to UV-B, friction, infection and contact dermatitis. Classical presentation mimics many erosive intertriginous dermatoses such as candidal intertrigo, flexural psoriasis, pemphigus vegetans, seborrheic dermatitis, Darier's disease, glucagonoma, zinc deficiency. Histopathological examination distinguishes HHD from all of them.

Genital lesions may present as verrucous papules [**5**]. White longitudinal bands and ridges on the fingernails have been reported to be of considerable clinical significance [**6**]. Our case did not have any nail changes. Infections and malignant transformation are the two important complications arising out of this disease. Association of primary cutaneous melanoma with HHD has been reported [**7**].

Our differential diagnoses included discoid lupus erythematosus, pemphigus foliaceous, granuloma faciale, Darier's disease, herpes simplex, basal cell carcinoma and pyoderma. Absence of hyperkeratosis as well as dyskeratosis in the form of "corp ronds" and "grains" in the epidermis excluded Darier's disease. Therapeutic response to antibiotic reinforced our diagnosis. Possibility of pyoderma could be excluded by long course of the disease, negative bacteriology and classical histopathological picture of HHD.

The treatment of HHD is difficult. Simple measures include reduction of skin friction and keeping flexures dry. Corticosteroids, administered topically, systemically or both, have shown good response. We treated the case with oral amoxicillin and topical mupirocin with favorable result.

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