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Extended Hailey-Hailey disease: Clinico-dermoscopic confrontation

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Abstract
Hailey-Hailey disease is an uncommon acantholytic genodermatosis. The extensive form is unusual and poses a difficulty of differential diagnosis with other bullous dermatoses and especially those with an annular arrangement like the case of our patient, in whom the clinical presentation was atypical but the dermoscopic examination had shown usual signs of Hailey-Hailey disease. Hence the interest of our case report.

Keywords: Hailey-Hailey disease, genodermatosis, extended form, dermoscopy, pink, white

Introduction
Described for the first time in 1939 by the brothers Howard and Hugh Hailey, Hailey-Hailey disease (HHD) is a rare genodermatosis. Its clinical presentation is heterogeneous and the differential diagnoses are multiple including other genodermatoses as well as inflammatory and infectious dermatoses. Although the positive diagnosis relies mainly on histology, dermoscopy remains a non-invasive technique that can guide the diagnosis.
We report the case of a 52-year-old patient with clinical, dermoscopic, and histologic evidence of extensive HHD.

Case report
52-year-old man with an erosive and painful skin rash, evolving by flare-up/remission for 4 years. The physical examination revealed erythematous annular plaques with a vesiculo-pustular and crusty border, affecting the trunk and the large folds (Figures 1 and 2).
A dermoscopic examination of a skin lesion revealed irregular pinkish-white areas spaced by pink furrows with white spots in a cloudy pattern. Flaccid vesiculopustules giving a crumpled tissue appearance and polymorphic vessels, including glomerular and linear vessels with peripheral distribution were also observed (Figure 3). Dermoscopy of the nail revealed longitudinal white bands (Figure 4).
Histological examination of a skin biopsy showed acantholysis involving the entire height of the mucous body, giving a shattered brick wall look (Figure 5).
Given these clinical, dermoscopic and histological criteria, the diagnosis of Hailey-Hailey disease was retained.

Discussion
HHD, also called benign familial pemphigus, is a rare autosomal dominantly inherited skin disorder marked by recurring blisters, erosions and crackles in flexural areas [1].
This blistering disease is related to a mutation in a calcium transporter protein in the Golgi apparatus encoded by the ATP2C1 gene on chromosome 3. Disruption of calcium homeostasis in the Golgi apparatus results in loss of cell adhesion of keratinocytes, leading to acantholysis [2].
The estimated incidence of HHD is approximately 1/50,000 [3]. No statistically significant frequency differences were found for gender and ethnicity. In two thirds of all cases, family cases are found. Triggering factors include menstrual periods, pregnancy, skin infections, physical trauma, oversweating and ultraviolet light. Other factors such as friction, patch tests, scabies and non-steroidal anti-inflammatory drugs can also lead to exacerbations [4].

A late onset is usually seen between the third and fourth decades of life. The most common areas affected are the folds such as the axillary, inguinal, perianal and lateral neck areas [3, 4]. The extended form, as in our case, has been seldom reported in the available literature [5]. Several factors have been incriminated, including Koebner's phenomenon and infection [6].

The elementary lesions of the disease are flaccid vesicles and blisters on an erythematous background, with peripheral extension giving an annular pattern. Lesions often result in crusty erosions, painful clefts and bad-smelling vegetations. Over-infections and post-inflammatory hyperpigmentation are common in the affected areas [3].

Dermoscopic features have been recently described and are mainly of interest in atypical forms like ours, where they help rule out other differential diagnoses. It includes irregular pink and white patches that are separated by pink furrows as well as polymorphous vessels [7]. Nail dermoscopy shows longitudinal white bands [8].

Histologically, there is hyperplasia of the epidermis, which usually occurs in more than half the thickness of the epidermis. Partial acantholysis gives a dilapidated brick wall appearance. Dyskeratotic cells are not as common as in Darier's disease. Moreover, parakeratosis and perivascular lymphocytic infiltration in the upper dermis are noted. Immunofluorescence studies are negative [4].

The major differential diagnoses of EHD are inflammatory and autoimmune diseases, especially other acantholytic dermatoses, such as Darier disease, and intertriginous infections.

The disease evolves with periods of remission and relapse. It is a challenging disease to manage which drastically impacts the patient's quality of life. Besides general measures, available therapeutic options include: topical treatments such as corticosteroids, vitamin D analogues, tacrolimus, as well as oral treatments that include antibiotics, corticosteroids, dapsone, retinoids and immunosuppressive therapies. In addition, physical treatments such as botulinum toxin injections, laser, dermabrasion, and surgical removal with secondary grafting may be offered, especially in cases of limited involvement. New systemic treatments have been proposed notably low-dose naltrexone, oral anticholinergics, magnesium chloride, apremilast and more recently dupilumab [3, 9, 10]. But despite all these therapeutic options, the treatment of Hailey Hailey's disease remains a real challenge.

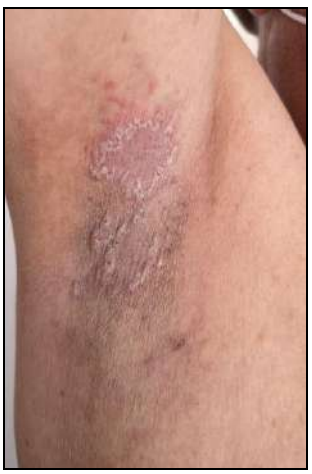

Fig 1: Annular erythematous plaques with a vesiculo-pustular and crusty border on the right axillary fold.

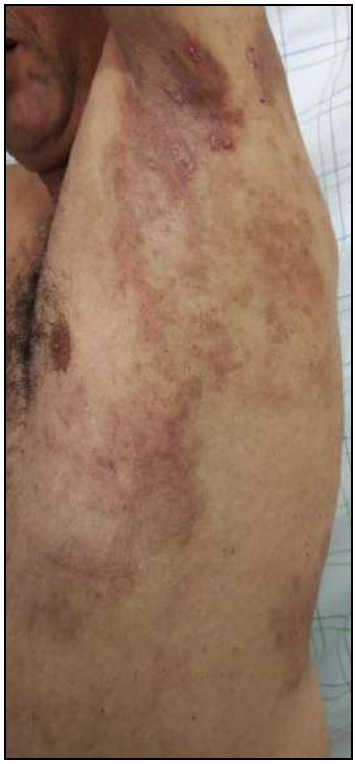

Fig 2: Annular erythematous plaques extended to the trunk.

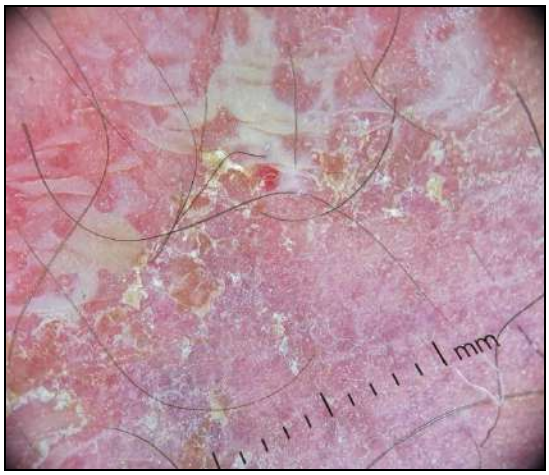

Fig 3: Dermoscopic features of skin lesions showing crumpled tissue, cloudy appearance, thin crusts and polymorphic vascularization.

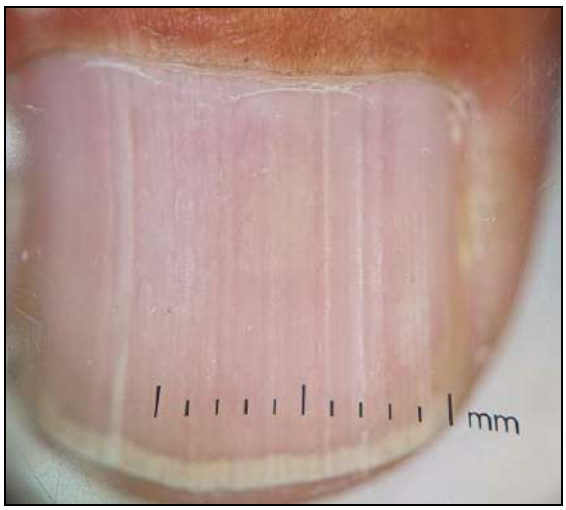

Fig 4: Dermoscopic of the nail showing longitudinal white bands

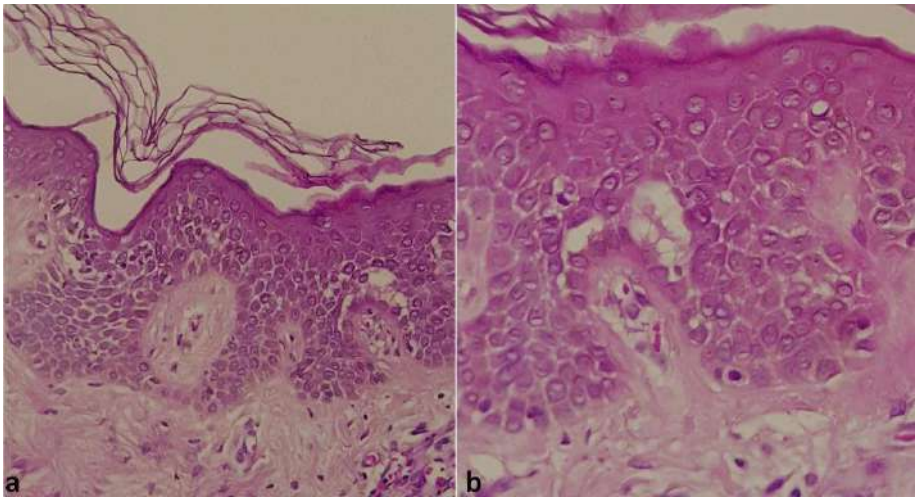

Fig 5: Histological image of a skin biopsy showing acantholysis involving the entire height of the mucosal body giving the appearance of a shattered brick wall. HE x 40 (a) and HE x 100 (b)

Conclusion

Clinical recognition of Hailey-Hailey disease can be easy, but in some cases of unusual presentation such as ours, dermoscopy can be very helpful in making the diagnosis. Furthermore, more studies are needed to come up with a therapeutic consensus to relieve patients whose quality of life is truly impaired.

Conflicts of interest: None disclosed

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Conflict of Interest: Not available

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