Hailey-Hailey disease

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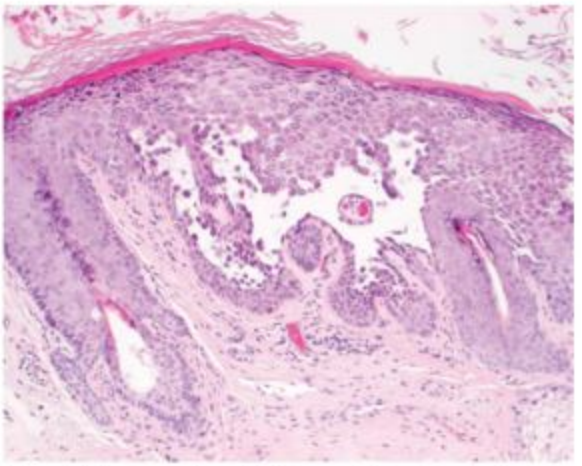

Figure 1. There is acantholysis of a hyperplastic epidermis, showing a suprabasilar cleft or vesicle and a "dilapidated brick wall" appearance.

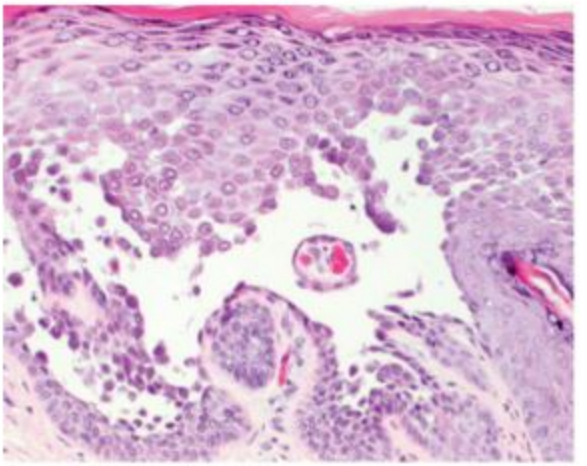

Figure 2. Tombstoning *is the term used to describe the residual basal cells separated from the rest of the epithelium. The cells in the cleft show rounding (corp ronds) of dyskeratotic cells.*

Hailey-Hailey disease is a genodermatosis characterized by incomplete penetrance of an autosomal dominant disorder with defects in the *ATP2C1* gene. Patients present with well-demarcated, erythematous plaques that often show crusting. The plaques may be vegetative, commonly associated with a distinct malodor. There may be associated burning or itching. While intertriginous areas are most commonly affected, the neck is also frequently involved. There is a natural remission and exacerbation cycle, with heat, moisture, and trauma the most common exacerbating factors. The lesions may be managed by corticosteroids or antibiotics, along with retinoids for severe disease.

Histologically, there is epidermal hyperplasia, usually involving more than 50% of the thickness of the epidermis (figure 1). Dyskeratosis and a suprabasal cleft may be seen, creating a so-called "dilapidated brick wall" appearance (figure 2), with dyskeratosis identified in the form of corp ronds and grains. Immunofluorescence studies are negative. The histologic differential diagnosis includes other vesiculo-bullous disorders, such as acantholytic actinic keratosis, acantholytic squamous cell carcinoma, Grover disease, and pemphigus vulgaris, among others. Careful correlation of the clinical findings with the histologic and immunofluorescence findings usually helps to make the diagnostic separation.

Suggested reading

Chiaravalloti A, Payette M. Hailey-Hailey disease and review of management. J Drugs Dermatol 2014;13(10):1254-7.

Dajani ZA, Mutasim DF. Ectopic facial Hailey-Hailey disease. J Am Acad Dermatol 2011;65(1):223-4.

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