

Milky-Red Areas: Dermoscopic Insights into Hailey-Hailey disease - A Case Series of 21 Patients

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Introduction

Hailey-Hailey disease, or benign familial pemphigus, is a rare autosomal dominant skin disorder caused by mutations in the ATP2C1 gene [1]. This study analyzed 21 patients diagnosed with Hailey-Hailey disease, focusing on distinctive dermoscopic patterns and complementary imaging findings using line-field confocal optical coherence tomography (LC-OCT), supported by histopathological confirmation.

Case Presentation

All patients in this retrospective study underwent dermoscopic examination of affected skin lesions LC-OCT imaging, and histological evaluation. The diagnosis was confirmed through histological evidence of suprabasal acantholysis and the characteristic “dilapidated brick wall” appearance of the epidermis. A distinctive dermoscopic feature consistently observed in 17 out of the 21 patients was the presence of milky-red areas.

These areas appeared as a homogeneous pinkish-whitish or milky hue, often interspersed with small erosions, crusting, and a patchy scaly surface (Figure 1B–D). This pattern was observed in both intertriginous zones and non-intertriginous areas, such as the trunk and back, highlighting its diagnostic significance across various anatomical sites. LC-OCT imaging corroborated these findings by identifying suprabasal clefts, intercellular splitting, and zones of partial acantholysis resembling a “crumbling brick wall,” hallmarks of Hailey-Hailey disease (Figure 1A). The consistent presence of milky-red areas in dermoscopy helps distinguish Hailey-Hailey disease from other conditions affecting similar anatomical regions. Seborrheic dermatitis is characterized by red dots and branching vessels on a dull red or yellowish background, accompanied by yellow-white scales, which differ in tone and texture from Hailey-Hailey lesions [2]. Extramammary Paget’s disease, particularly in its early stages, presents with porcelain white patches and polymorphic vessels, evolving to include glomerular vessels over time [3]. However, it lacks the

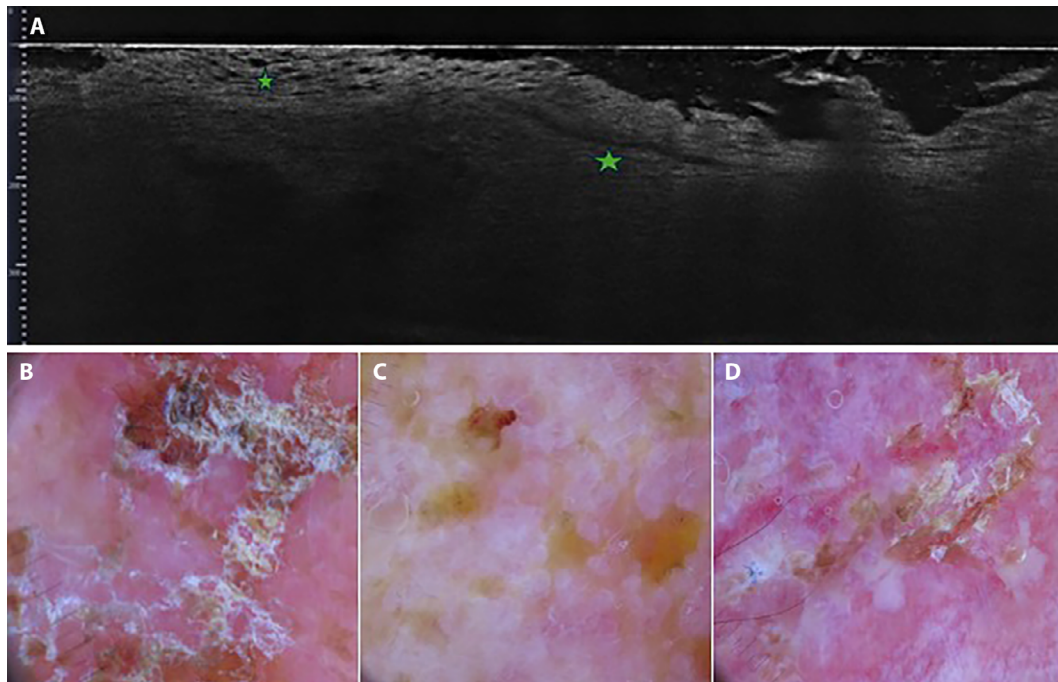


Figure 1. Imaging of Hailey-Hailey disease. (A) LC-OCT vertical section of an affected area: multiple parallel clefting diffuse in the context of the epidermis, along with areas of partial acantholysis resembling a “crumbling brick wall” (green stars). (B) Dermoscopy of neck: homogeneous milky-red areas with adherent whitish scales and overlying yellow-brown crusts. (C) Dermoscopy of axilla: diffuse milky-red background interspersed with small erosions and scattered yellowish crusts. (D) Dermoscopy of back: milky-red areas with thick scales, brownish crusts, and superficial linear erosions.

homogeneous milky-red areas characteristic of Hailey-Hailey disease. Similar dermoscopic patterns have previously been described under different terminologies, such as the ‘pink whitish background’ reported by Oliveira et al. [4] and the ‘pink structureless areas’ described by Kurzeja et al. [5]. Our findings confirm and expand these observations, proposing the term ‘milky-red areas’ to better characterize this distinctive feature of Hailey-Hailey disease.

The inclusion of LC-OCT imaging provides a valuable adjunct to dermoscopy, offering high-resolution, real-time visualization of epidermal and dermal structures. Findings such as intercellular clefting, suprabasal separation, and focal acantholysis closely aligned with histological hallmarks of Hailey-Hailey disease, underscoring the role of LC-OCT in enhancing diagnostic confidence and potentially reducing the need for invasive biopsy procedures in some cases. To date, only one case report, by Di Stefani et al., has described the use of LC-OCT in Hailey-Hailey disease. Our study represents the first series including a large cohort of patients (N=21), thus expanding the current knowledge on LC-OCT features of this condition and confirming its potential role as a non-invasive diagnostic tool.

Conclusion

Milky-red areas represent a consistent and reproducible dermoscopic feature of Hailey-Hailey disease, observed in most

patients and across different anatomical sites. This finding may help distinguish Hailey-Hailey disease from other dermatoses with overlapping clinical presentations. The combination of dermoscopy and LC-OCT provides useful non-invasive support for diagnosis and may reduce the need for skin biopsy in selected cases.

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