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Dermoscopic and reflectance confocal microscopic presentation of Hailey-Hailey disease: A case series.

Oliveira A, Arzberger E, Pimentel B, de Sousa VC, Leal-Filipe P. Skin Res Technol. 2017 Aug 7. doi: 10.1111/srt.12394.

ABSTRACT

BACKGROUND/PURPOSE: Hailey-Hailey disease is a rare inherited acantholytic skin disorder characterized by heterogeneous clinical presentation. Its differential diagnosis might be wide, including other genodermatoses, inflammatory, and infectious skin diseases. Although histopathology remains as diagnostic gold standard, noninvasive techniques such as dermoscopy and reflectance confocal microscopy may assist clinical examination. Herein, we aim to further characterize the dermoscopic and reflectance confocal microscopic presentation of Hailey-Hailey disease with histologic correlation. METHODS: Eight patients with Hailey-Hailey disease were consecutively recruited. All patients were examined using dermoscopy and reflectance confocal microscopy. RESULTS: In all cases, dermoscopy enabled the visualization of polymorphous vessels, including glomerular and linear-looped vessels, within a pink-whitish background. Reflectance confocal microscopy revealed wide suprabasilar partial acantholysis and clefting, crusts, dilated papillae with tortuous vessels, and inflammatory cells. Dyskeratosis, uplocated papillae, and adnexal sparing were also observed. CONCLUSION: Although definite diagnosis was obtained by histopathology in all cases, dermoscopy and reflectance confocal microscopy allowed the identification of common features (even in cases with dissimilar clinical presentation) that may support an early diagnosis of Hailey-Hailey disease, and its differentiation from other more frequent skin disorders © 2017 John Wiley & Sons A/S. Published by John Wiley & Sons Ltd. KEYWORDS: Hailey-Hailey disease; acantholysis; dermoscopy; familial benign chronic pemphigus; genodermatosis; reflectance confocal microscopy PMID: 28782140 DOI: 10.1111/srt.12394