


## CASE IMAGE

# Lichenoid lesions around the anus: An unusual site of Hailey–Hailey disease

Fatma Hammami  | Khadija Sellami | Rim Chaabouni | Sonia Boudaya | Meriem Amouri | Emna Bahloul | Hamida Turki

Dermatology Department, Hedi Chaker University Hospital, Sfax, Tunisia

**Correspondence**

Fatma Hammami, Department of Dermatology, Hedi Chaker University Hospital, Sfax, Tunisia.

Email: [fatmahammami.medicine@gmail.com](mailto:fatmahammami.medicine@gmail.com)

**Abstract**

The anal region is an unusual site of Hailey–Hailey disease. It manifests with lichenoid lesions with crusted erosions around the anus. It should be differentiated from condylomata acuminata, extramammary Paget disease, and bowenoid papulosis.

**KEYWORDS**

acantholysis, condylomata acuminata, Hailey–Hailey disease, lichenoid lesions, perianal

A 77-year-old woman presented with a 2-year history of anal pruritus. Physical examination revealed a lichenoid lesion around the anus with multiple white papules and erosions (Figure 1). There were no other similar lesions elsewhere on the body. There was no nail, mucosal, or scalp involvement. There was no family history of similar lesions. A biopsy from the perianal region was performed. Histology showed prominent suprabasal keratinocytes acantholysis in the epidermis giving a dilapidated brick wall appearance with hyperacanthosis and focal parakeratosis. Direct immunofluorescence examination was negative. Therefore, the lesions were diagnosed as Hailey–Hailey disease (HHD). The lesions resolved after topical corticosteroid treatment. Relapse occurred after the discontinuation of the treatment.

Hailey–Hailey disease is an autosomal dominant disorder. The disease is caused by a mutation in the calcium-dependent adenosine triphosphatase *ATP2C1* gene resulting in defects in keratinocytes adhesion.<sup>1</sup> Clinically, it manifests with flaccid vesicles or bullae leaving macerated or crusted erosions. The lesions are commonly located in intertriginous areas. Unusual presentations include



**FIGURE 1** White smooth papules and erosions around the anus

This is an open access article under the terms of the [Creative Commons Attribution-NonCommercial-NoDerivs](https://creativecommons.org/licenses/by-nc-nd/4.0/) License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2022 The Authors. *Clinical Case Reports* published by John Wiley & Sons Ltd.

segmental, extensive, mucosal, genitoanal, and photoexposed area involvement.<sup>2</sup> In the anal region, the lesions may be misdiagnosed as condylomata acuminata, extramammary Paget disease, or bowenoid papulosis. Histology shows acantholysis and dyskeratosis. Treatment of perianal HHD can be challenging. Regular defecation, diarrhea, and humidity in the anal region gave rise to a chronic course of HHD. Topical corticosteroids and tacrolimus may be effective in some mild cases of perianal HHD. Other modalities such as surgical resection or ablative laser can be useful in recalcitrant more extensive diseases.

#### AUTHOR CONTRIBUTIONS

Dr Fatma Hammami wrote the manuscript and is the guarantor of the content of the manuscript, included the data and analysis. Dr Emna Bahloul and Dr Khadija Sellami revised it critically. Dr Sonia Boudaya, Dr Meriem Amouri, and Dr Hamida Tuki contributed to the final approval of the version of the manuscript to be submitted.

#### ACKNOWLEDGMENT

None.

#### FUNDING INFORMATION

None.

#### CONFLICTS OF INTEREST

None.

#### ETHICS STATEMENT

The paper is not currently being considered for publication elsewhere.

#### CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

#### DATA AVAILABILITY STATEMENT

None.

#### ORCID

Fatma Hammami  <https://orcid.org/0000-0001-8463-3742>

#### REFERENCES

1. Hu Z, Bonifas JM, Beech J, et al. Mutations in ATP2C1, encoding a calcium pump, cause Hailey-Hailey disease. *Nat Genet.* 2000;24(1):61-65.
2. Vasudevan B, Verma R, Badwal S, Neema S, Mitra D, Sethumadhavan T. Hailey-Hailey disease with skin lesions at unusual sites and a good response to acitretin. *Indian J Dermatol Venereol Leprol.* 2015;81(1):88-91.

**How to cite this article:** Hammami F, Sellami K, Chaabouni R, et al. Lichenoid lesions around the anus: An unusual site of Hailey–Hailey disease. *Clin Case Rep.* 2022;10:e06702. doi:[10.1002/ccr3.6702](https://doi.org/10.1002/ccr3.6702)