

underlying disease is present, its treatment can lead to improvement or even resolution of the PR lesions.<sup>8</sup>

To our knowledge, this is the first reported case of PR associated with hyperprolactinemia, and we therefore consider its publication important. The finding of this dermatosis should always alert the physician to the possibility of malignancy, systemic diseases, or hormonal disorders.

## Conflicts of Interest

The authors declare that they have no conflicts of interest.

## References

1. Bakry OA, Samaka RM. Photoletter to the editor: Pityriasis rotunda. *J Dermatol Case Rep.* 2012;6:90–2.
2. Friedmann AC, Ameen M, Swale VJ. Familial pityriasis rotunda in black-skinned patients; a first report. *Br J Dermatol.* 2007;156:1365–7.
3. Swift PJ, Saxe N. Pityriasis rotunda in South Africa. A skin disease caused by undernutrition. *Clin Exp Dermatol.* 1985;10:407–12.
4. Batra P, Cheung W, Meehan SA, Pomeranz M. Pityriasis rotunda. *Dermatol Online J.* 2009;15:14.
5. Persechino S, Raffa S, Caperchi C, Visco V, Verga E, Trapolini M, et al. Pityriasis rotunda: Sporadic occurrence in an elderly Caucasian patient with diabetes mellitus. *Eur J Dermatol.* 2009;19:393–4.
6. Makino T, Mizawa M, Seki Y, Hayashi M, Shimizu T. Decreased filaggrin-2 expression in the epidermis in a case of pityriasis rotunda. *Clin Exp Dermatol.* 2015;1–3, <http://dx.doi.org/10.1111/ced.12716> [Epub ahead of print].
7. Grimalt R, Gelmetti C, Brusasco A, Tadini G, Caputo R. Pityriasis rotunda: Report of a familial occurrence and review of the literature. *J Am Acad Dermatol.* 1994;31:866–71.
8. Finch JJ, Olson CL. Hyperpigmented patches on the trunk of a Nigerian woman. Pityriasis rotunda (PR). *Arch Dermatol.* 2008;144:1509–14.

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## Good Response to Doxycycline in Hailey-Hailey Disease<sup>☆</sup>



### Enfermedad de Hailey-Hailey, adecuada respuesta a doxiciclina

*To the Editor:*

Hailey-Hailey disease (HHD) or familial benign chronic pemphigus is a rare skin disorder characterized clinically by vesicles and erosions in the intertriginous areas, mainly the axillae and groin.<sup>1–4</sup> The site of the lesions, the pain, and sometimes their smell have a marked impact on patients' quality of life<sup>5</sup>; this has led to the use of numerous medical and surgical treatments, with variable degrees of success. We present a patient with long-standing, extensive HHD who presented an excellent response to doxycycline.

A 60-year-old man with no past history of interest was referred from another health area for a recurrent dermatosis that had arisen 20 years earlier and affected the skin folds. He presented crusted, exudative, erosive-vesicular lesions in the skin folds, particularly the axillary and inguinal folds (Fig. 1), the cubital fossa, neck, and perineum. The lesions were pruritic and painful and became worse during the summer and with exercise. He had previously been treated with corticosteroids and topical antibiotics with little improvement. The patient reported no family history

of skin disease, though the youngest of his 4 offspring (2 women and 2 men) presented similar but milder lesions on the neck and in the axillae. Biopsy confirmed the diagnosis of HHD (Fig. 2). Direct immunofluorescence was negative. Routine blood tests were normal and antinuclear antibodies and indirect immunofluorescence for anti-intercellular cement substance and antibasement membrane zone antibodies were negative.

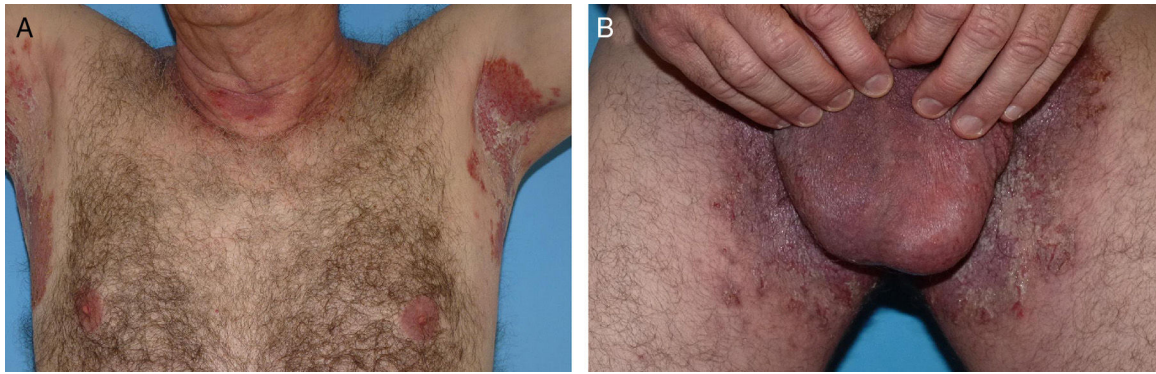
Treatment was prescribed with doxycycline at a dose of 100 mg/d. This was very well tolerated and led to a rapid improvement that was maintained throughout the summer months. The dose was then reduced to 50 mg/d. After 16 months of follow-up on the same dose, the patient remained asymptomatic (Fig. 3).

HHD is a rare chronic genodermatosis of autosomal dominant inheritance. It is caused by mutations in the *ATP2C1* gene on chromosome 3q21-24. Mutations of this gene, which codes for the secretory pathway  $Ca^{++}/Mn^{++}$ -ATPase (SPCA1) of the Golgi apparatus, cause changes in calcium-dependent intracellular signals, producing a loss of cell adhesion in the epidermis, leading to acantholysis.<sup>1–3</sup> The exact mechanism of the changes remains unclear.<sup>2,3</sup>

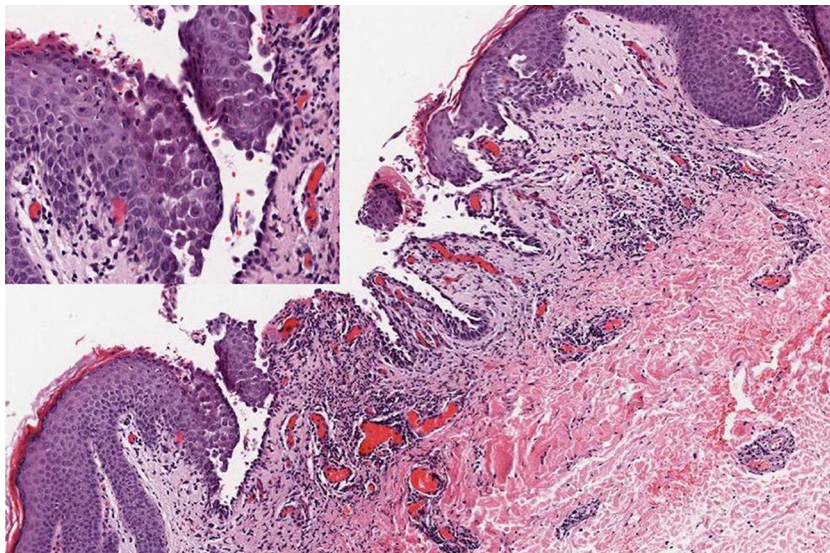
HHD has been treated with varying degrees of success using a variety of treatments aimed at mitigating the inflammation or reducing the triggering factors; treatments reported in the literature include topical and systemic corticosteroids, topical antibiotics, oral retinoids, immunosuppressants such as ciclosporin and methotrexate, dapsone, botulinum toxin, oral glycopyrrolate, dermabrasion, various lasers, and photodynamic therapy.<sup>6</sup>

Tetracycline, doxycycline, and minocycline have been used with success in dermatology, exploiting their nonantimicrobial effects as anti-inflammatory agents (inhibition of leucocyte chemotaxis and activation and regulation of

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**Figure 1** A and B, Erosive, erythematous plaques in the axillary and inguinal skin folds.



**Figure 2** Suprabasal acantholysis with a dilapidated brick wall appearance. Hematoxylin and eosin (H&E), original magnification  $\times 20$ ). Inset: detail of the acantholysis; H&E, original magnification  $\times 40$ .



**Figure 3** A and B, Complete resolution of the lesions in the axillary and inguinal skin folds 16 months after initiating treatment with doxycycline.

inflammatory cytokines in keratinocytes) and their anti-collagenase activity via inhibition of the dermal matrix metalloproteinases. Metalloproteinase 9 and its inhibitor have been implicated in HHD and in Darier disease.<sup>7,8</sup>

The recent publication of 6 cases of HHD with a dramatic response to doxycycline,<sup>7</sup> with ease of access and management, low cost, and few side effects, led us to use this drug. The treatment achieved an excellent response never before experienced by our patient either spontaneously or with other topical treatments (dermal corticosteroids and fusidic acid).

## Conflicts of Interest

The authors declare that they have no conflicts of interest

## References

1. Hu Z, Bonifas JM, Beech J, Bench G, Shigihara T, Ogawa H, et al. Mutations in ATP2C1, encoding a calcium pump, cause Hailey-Hailey disease. *Nat Genet.* 2000;24:61–5.
2. Dhitavat J, Fairclough RJ, Hovnanian A, Burge SM. Calcium pumps and keratinocytes: lessons from Darier's disease and Hailey-Hailey disease. *Br J Dermatol.* 2004;150:821–8.
3. Leinonen PT, Hägg PM, Peltonen S. Reevaluation of the normal epidermal calcium gradient, and analysis of calcium levels and

ATP receptors in Hailey-Hailey and Darier epidermis. *J Invest Dermatol.* 2009;129:1379–87.

4. Burge SM. Hailey-Hailey disease: The clinical features, response to treatment and prognosis. *Br J Dermatol.* 1992;126:275–82.
5. Gisondi P, Sampogna F, Annessi G, Girolomoni G, Abeni D. Severe impairment of quality of life in Hailey-Hailey disease. *Acta Derm Venereol.* 2005;85:132–5.
6. Chiaravalloti A, Payette M. Hailey-Hailey disease and review of management. *J Drugs Dermatol.* 2014;13:1254–7.
7. Le Saché-de Peufeilhoux L, Raynaud E, Bouchardeau A, Fraitag S, Bodemer C. Familial benign chronic pemphigus and doxycycline: A review of 6 cases. *J Eur Acad Dermatol Venereol.* 2014;28:370–3.
8. Kobayashi T, Sakuraoaka K, Hattori S, Hori H, Nagai Y, Tajima S, et al. Immunolocalization of human gelatinase (type IV collagenase, MMP-9) and tissue inhibitor of metalloproteinase 1 in Hailey-Hailey and Darier's diseases. *Dermatology.* 1996;193:110–4.

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## Importance of educational sessions on cardiometabolic comorbidities. Awareness among psoriasis patients



## Importancia de las sesiones educativas sobre comorbilidades cardiometabólicas. Conciencia entre los pacientes con psoriasis

Dear Editor:

There is strong evidence that psoriasis is associated with several cardiometabolic comorbidities, and that patients with psoriasis are at a higher risk of cardiovascular morbidity and mortality.<sup>1–3</sup> Understanding this is of crucial importance, not only for physicians but also for patients, as it can impact prognosis and patient quality of life.<sup>4</sup>

It has been shown that few patients with moderate to severe psoriasis are aware of their increased risk of atherothrombotic disease and metabolic syndrome.<sup>5</sup> Educational sessions are a recognized tool for informing and helping patients to understand the nature and course of their disease and the different treatments available, and can also help them to develop coping strategies.<sup>6,7</sup>

We performed an observational study to evaluate the impact of an educational session designed to promote knowledge among patients with psoriasis about their disease,

lifestyle changes, and management of cardiometabolic comorbidities.

The educational session was held in the psoriasis unit of a Portuguese tertiary hospital. Briefly, it consisted of several oral presentations (30 min each) explaining the nature of psoriasis, introducing the various treatment options, exploring the association between psoriasis and cardiometabolic comorbidities/cardiovascular disease, and underlining the importance of monitoring and treating these. A questionnaire was created for the patients to complete before, immediately after, and 6 months after the session. The questionnaire included demographic information, questions regarding the association between psoriasis and cardiometabolic comorbidities/cardiovascular disease, and assessment of lifestyle and comorbidity management.

Seventy patients participated in the session and 53 completed all 3 questionnaires correctly. The demographic data, characteristics of disease, and treatments received are presented in [Table 1](#). Regarding cardiometabolic comorbidities, 35.8%, 13.2%, and 35.8% of patients had a respective diagnosis of hypertension, diabetes mellitus, and dyslipidemia; 20.8% were obese (body mass index > 30); and 18.9% were active smokers ([Table 1](#)).

The McNemar test was used to assess significant improvements in knowledge between the different time points. A *P* value of less than or equal to .05 was considered statistically significant. A significant increase was observed in the percentage of correctly answered questions about the association between psoriasis and cardiometabolic comorbidities/cardiovascular disease on comparing the answers