Narrowband UV-B Phototherapy in the Treatment of Generalized Hailey-Hailey Disease

Tratamiento de la enfermedad de Hailey-Hailey generalizada con fototerapia UVB de banda estrecha

To the Editor:

Hailey-Hailey disease (HHD) is an autosomal dominant genodermatosis caused by a mutation in the ATP2C1 gene. This gene encodes the hPACA1 protein, which is responsible for calcium homeostasis. The mutation affects desmosome function, resulting in suprabasal acantholysis.1-3 Clinically, it presents with recurrent flares of erythematous and macerated plaques in intertriginous areas, sometimes accompanied by erosions, fissures, and vesicles.4 Lesion superinfection and a foul odor, which can compromise patient quality of life, are observed in some cases.5 Generalized forms are infrequent and can be triggered by infections or drugs.5,6 There are multiple available therapeutic options of variable efficacy.1,7 Here, we describe complete lesion disappearance following narrowband ultraviolet B (NBUVB) phototherapy in a patient with generalized HHD resistant to multiple therapeutic regimens.

The patient was a 41-year-old man with skin phenotype III and a history of histologically confirmed HHD that began at age 28 with involvement of the neck, axillae, and groin.

He was seen for eroded and macerated erythematous plaques located in the skin folds. Over the preceding months the lesions, which caused mild pain and produced a foul odor, had spread to the trunk, upper extremities, and proximal thighs (Figs. 1 and 2).

The lesions spread progressively despite treatment with 1% isoconazole and 2% mupirocin cream, 100 mg minocycline every 12 hours, and 150 mg fluconazole per day for 7 days, in repeated cycles.

In the absence of a clinical response 3 weeks after starting treatment with prednisone (40 mg/day), NBUVB phototherapy was added to the regimen. NBUVB irradiation was carried out using a Waldmann F85/100W-01 cabinet equipped with TL01 lamps. The patient underwent a total of 8 weekly sessions, starting at a dose of 300 mJ/cm² and increasing by 50 mJ/cm² per week. The maximum dose of 500 mJ/cm² was reached after the fifth session, and was maintained until the eighth week. The patient received a cumulative dose of 3500 mJ/cm². During each session he was positioned standing with the lower limbs in abduction and the upper limbs elevated above the level of the shoulders, thus allowing irradiation of the skin folds. No adverse effects were observed during treatment.

Four weeks after starting phototherapy a marked improvement was observed; the lesion extension decreased and the odor subsided. Consequently, the prednisone dose was progressively decreased at a rate of 5 mg per week, and prednisone treatment was ultimately discontinued 1 month after cessation of phototherapy.

After completing the 8 phototherapy sessions the lesions resolved completely, leaving only residual hyperpigmentation (Fig. 3).

Currently, the patient is being treated with 1% clotrimazole and 0.1% betamethasone cream once per day, and has not relapsed over 12 months of follow-up.

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Figure 1  Macerated erythematous plaques.

Conventional treatment of HHD consists of topical and oral corticosteroids, antibiotics and antifungals, and topical calcineurin inhibitors, combined with general measures. Typically, the therapeutic response is both temporary and partial. The use of methotrexate, cyclosporine, thalidomide, retinoids, dapsone, botulinum toxin, dermabrasion, CO$_2$ laser, alexandrite, erbium-doped yttrium aluminum garnet laser, radiotherapy, ultraviolet B (UV-B) phototherapy, and surgical treatment has been described, and photodynamic therapy has been proposed for the treatment of refractory cases.

In the case reported here, the patient presented with generalized HHD that responded poorly to corticosteroids in monotherapy, but responded well when NBUVB phototherapy was added to the treatment regimen. An improvement was observed 1 month after starting phototherapy and the lesions disappeared after 8 sessions over 2 months.

The use of NBUVB phototherapy for the treatment of HHD was first described in 1999 in a woman with psoriasis and concomitant HHD who was treated twice per week for 2 months with a suberythematogenic dose, to which she responded well. Mizuno and coworkers described the case of a woman with disseminated HHD who, after multiple treatment regimens without adequate response, underwent 8 sessions of NBUVB phototherapy using a similar regimen to that of our patient, and showed lesion improvement 2 weeks after completing treatment. The patient relapsed after treatment discontinuation, and once again showed a good response following its reintroduction. By contrast, our patient, who was treated for generalized HHD, showed no signs of recurrence up to 1 year after discontinuation of phototherapy. The lack of relapse over a long follow-up period may be because oral corticosteroid treatment had been initiated before NBUVB phototherapy began, and was

Figure 2  Involvement of the chest, face, and upper arms.
continued, albeit at a lower dose, after completing the phototherapy regimen.

Classically, UV light is considered an aggravating factor of HDD because it stimulates production of proinflammatory cytokines, suppresses ATP2C1 mRNA expression, and promotes acantholysis. Conversely, UV-B phototherapy decreases the number of epidermal T lymphocytes and dendritic cells, thereby attenuating cutaneous inflammation. In our patient, and in other cases described in the literature, phototherapy resulted in clinical improvement, suggesting the involvement of other factors in cases in which lesions worsen.7,11

The present case is of particular interest because the patient in question had generalized HHD that responded poorly to conventional treatments but showed a good therapeutic response to NBUVB phototherapy. Based on these findings, we propose NBUVB phototherapy as a useful therapeutic alternative in patients with these characteristics.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

Acknowledgments

The authors thank the patient for providing written consent to the publication of the images included in this letter.

References


Simultaneous Disappearance of Various Nevi in a Patient with Autoimmune Disorders

Desaparición simultánea de nevus en un paciente con procesos autoinmunes asociados

To the Editor:

During digital follow-up of a 42-year-old woman with atypical nevi using a MoleMax II video dermoscopy system (Derma Medical, London, UK), the patient's melanocytic lesions progressively disappeared and were practically absent by the end of a 3-year period (Fig. 1). She presented with several autoimmune processes, including HLA-B27-positive sacroiliitis, ANA-positive peripheral arthritis, and autoimmune hypothyroidism that was treated with supplemental L-tyrosine. She had no personal or family history of melanoma. There was no evidence of a halo phenomenon or any other reaction around the nevi at any time. Several potential explanations can account for the disappearance of melanocytic nevi. Progressive disappearance in adulthood is a described feature of acquired melanocytic nevi (AMN).

However, the rapid disappearance of AMN in young adults, as in the present case, is unusual. This involution can be caused by certain specific reactions. The halo or Sutton nevus can progressively fade and ultimately disappear, leaving a depigmented halo surrounding the site of the original nevus. Histologically, the condition is characterized by the presence of lymphocytic infiltrate with predominance of CD8+ T lymphocytes. This infiltrate results in progressive destruction of the melanocytes via a mecha-

Figure 1 Clinical and dermoscopic images showing changes in the patient's nevi.