

Photoinduced lentiginous acantholytic dermatosis. A new and unusual pattern of Grover's disease?

Ignacio Rébora¹, Osvaldo Stringa², Mario Abbruzzese³, Javier Anaya⁴

ABSTRACT

Transient acantholytic dermatosis or Grover's disease, is a dermatosis of unknown aetiology, that typically presents as an erythematous papular eruption with crops of vesicles and crusts over the trunk; in which the most common histopathological finding is the presence of focal acantholysis and dyskeratosis. It affects mostly middle-aged or elderly men. One of the triggering factors most frequently described in the literature is the exposure to sunlight.

Four cases have been recently reported of Grover's disease with a new pattern of clinical presentation, characterized by inflamed papules and crusts following sun exposure, along with lentiginous "freckling". They were three middle aged women and a 60 years old man. The histopathological findings coincided in presenting interpapular ridges elongation associated with focal acantholysis and dyskeratosis. We present a 38 year old female with clinical and histopathological lesions compatible with this new lentiginous pattern of Grover's disease described by Cooper in 2004 (*Dermatol Argent* 2010;16(2):122-125).

Keywords:

transient acantholytic dermatosis, Grover's disease, lentiginous, photosensitivity.

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Introduction

The transient acantholytic dermatosis or Grover's disease, is a skin condition of unknown etiology, characterized by presenting itself as a papular, papuloerotion, or papulovesicular, eruption, polymorphic and pruritic. Usually has a self-limited course of action, although it may persist during weeks or months. It affects mainly men over 40 years old. The etiology is unknown but some triggering factors have been identified, including exposure to sunlight and heat being the most frequently associated, and profuse sweating and obstruction of acrosyringium being mentioned less frequently.^{1,2}

Grover's Disease is characterized for presenting focal acantholysis on a histopathological analysis. It can display under one of the four known patterns or any combination of some of them. The most common one of them is similar to Darier's disease, with suprabasal acantholysis and dyskeratotic cells and lymphocytic infiltrate on dermis. Another pattern is suprabasal acantholysis with blister cleavage that mimics pemphigus vulgaris, or acantholysis in Malpighi stratum similar to that found in Hailey-Hailey disease. The fourth pattern is accompanied by spongiosis. When acantholysis is found inside the spongiotic vesicle, it is considered an important histopathological sign, otherwise it is considered

¹ Chief of Residents of Dermatology

² Deputy Head of the Dermatology Department

³ Medical staff, Department of Dermatology

⁴ Medical dermatopathologist

⁵ Jefe del Servicio de Dermatología

Hospital Universitario Austral. Pilar, Buenos Aires, Argentina.

Correspondence author: Dr. Ignacio Rébora: Mons. Alberti 536 2º B, (1642) San Isidro, Buenos Aires, Rep. Argentina
nachorebora@yahoo.com



PHOTO 1. Erythematous papules lesions, pigmented brown macules papule-crust and lentiginous appearance on the thighs.



PHOTO 2. Erythematous papules lesions, pigmented brown macules papule-crust and lentiginous appearance on the thighs.

pathognomonic of Grover's disease. Other focal epidermal alterations that may accompany the acantholysis are hyperkeratosis, acanthosis and parakeratosis.³⁻⁶

Clinical case

We report the case of a patient of 38 years old, who came for a consult in April 2006 due to presenting an itchy rash after sun exposure on frontal area of thighs, cleavage, external face of forearms and back of hands (**Photo 1**). The patient

had no personal or family history of skin diseases and presented a skin photo-type III. She reported no intake of any drug or photo-sensitizer.

Physical examination showed erythematous-crust papules, of 2 to 3 mm in diameter, some of which were located on brown-pigmented macules of lentiginous aspect. With the same topography, flat pigmented lesions, with net edges, consistent with lentigines, presenting no inflammatory signs (**Photo 2**). There were no other injuries on nails, hair, skin or mucous. The lesions started as asymptomatic brown macules and then, after sun exposure, the rash appeared presenting elevations and serous crusts on top of them.

By dermatoscopy the presence of a fine and regular pigment network at the periphery of the lesions was observed, presenting an erythematous or erythematous-crust center without pigmentation and with some small visible blood vessels. There were some injuries where only the pigmented network could be seen throughout the lesion and some others presented only erythema and crusting, without pigmentation (**Photo 3**).

Photoprotection was prescribed noticing immediate symptomatic improvement and reduction of the erythematous-crust papules, but with persistence of abnormal pigmentation. Later, in the beginning of next summer, in December 2006, the patient consulted again due to the reappearance of papular lesions on top of lenticular lesions and a set of new lesions of the same characteristics and topography. In the subsequent tests the disappearance of the inflammatory component could be observed, but with persistence of lentigines in increasing numbers. The laboratory studies, which included blood count, liver function, kidney function, protein electrophoresis, FAN and porphyrins in blood and urine, gave results within normal parameters. The histopathology performed over a thigh injury, which was clinically characterized as a erythematous-crust papule located over a brown lenticular injury, showed an area with elongation of interpapular epidermal crests, moderated melanin pigmentation without melanocytic proliferation, acantholytic phenomena, focal dyskeratosis and parakeratosis, accompanied in dermis by mild superficial perivascular lymphocytic infiltrates (**Photos 4 and 5**). The direct immunofluorescence study came out negative.

Commentary

In 2004, Cooper et al., reported two cases of papulovesicular rashes, peeling and itchy with associated lentigines lesions, located in photoexposed areas which appeared after sun exposure, on two middle-aged women (39 and 42 years old).⁷ The histopathological findings were consistent with Grover's disease, with focal acantholysis, dyskeratosis and parakeratosis; and on pigmented lesions, elongation of interpapular

crests with increasing number of melanocytes but without any acantholysis. In the case discussed here we found, on the same lesion, acantholysis, dyskeratosis and parakeratosis in the center with elongation of the network of crests and increase of melanin pigment in the periphery of the lesion.

Subsequently, in 2005, Girard et al. reported two other cases, a 60 year old man and a woman of 43 years old.⁸ They described the appearance of lentiginous in injuries which preceded for months the papular eruption, which also appears after sun exposure. One of the two cases was treated with acitretin 0.5 mg/kg/day with topical calcipotriol, resulting in the disappearance of papules and persistence of lentiginous. In our case we observed a similar response when photo-protection of the injuries, although with recurrent papular lesions when exposed to sunlight again.

Dr. Lucio Bakos, from Brazil, during an exposition (Apertura 2007 Symposium "Clinical and Therapeutic Ateneo X" of the Argentina Society of Dermatology in Buenos Aires, Argentina, in March 2007) described two other cases corresponding to this photosensitive lentiginous acantholytic dermatosis, interpreted at that time, as a lentiginous variant of Grover's disease. We believe that this lentiginous acantholytic and photosensitive rash may represent a new and unusual clinical variant of Grover's disease or to a completely new entity. Grover's disease or transient acantholytic dermatosis, correspond to an acquired dermatosis of unknown cause, clinically characterized by the appearance of erythematous-vesicular or erythematous-crusting papules on stem and root of members, usually associated with exposure to sunlight, heat or sweating. It mainly affects elder men, although the cases described by Cooper and Girard corresponded to three women and one man between 39 and 60 years old; and ours corresponds also to a middle-aged woman. It is usually transient and self-curing once the triggering factor is removed, as it was observed in our case. The histopathological findings of focal acantholysis with dyskeratosis coincides with the most common pattern found Grover's disease, and the elongation of the interpapular crests, with lentiginous lesions described by Cooper and Girard.

We consider as differential diagnosis Darier's disease, for presenting dyskeratotic acantholysis histopathological analysis, being exacerbated by sunlight and erythematous-crusting lesions, although injuries did not show the keratotic characteristic clinical appearance of Darier's disease nor the predominance of seborrheic areas was observed. Moreover, our patient lacked a disease family history and began at a relatively late age onset, which usually happens in cases of Darier's disease. In addition, the histopathology showed that areas of acantholysis presented less dyskeratosis usually seen in Darier's disease; and the elongation and hyperpigmentation of the interpapular crests is not usually seen in this disease.

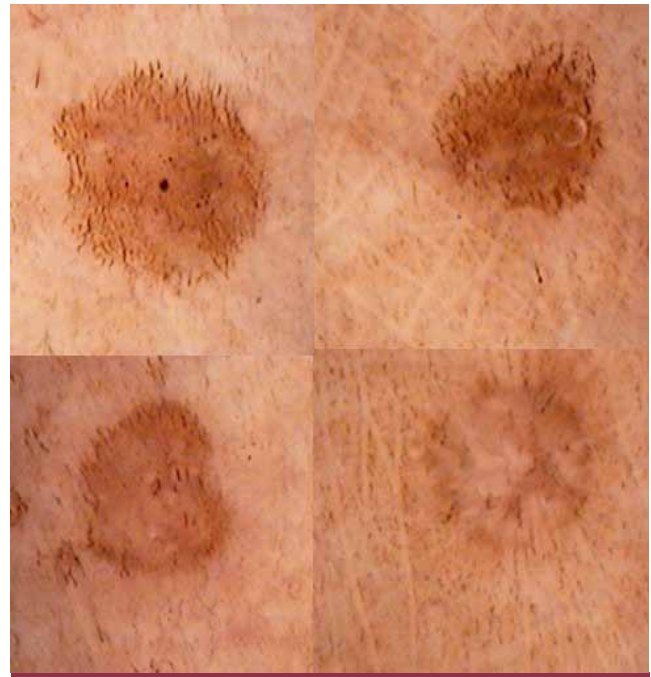


PHOTO 3. Dermoscopy: fine pigmented network, regular on the periphery of the lesions, with erythematous or erythematous-crusting center without pigmentation and with some small blood vessels.



PHOTO 4. Dermoscopy: fine pigmented network, regular on the periphery of the lesions, with erythematous or erythematous-crusting center without pigmentation and with some small blood vessels.

Benign familial pemphigus or Hailey-Hailey disease was considered as a differential histological diagnosis, but our patient has no family record of this condition; convex areas are affected and not fold areas or of friction, and histopathologically no pattern for acantholysis in "collapsed wall" in the Malpighian stratum was found, which is typical of Hailey-Hailey disease.

Finally, we consider the differential diagnosis for pemphigus vulgaris for the observation of suprabasal acantholysis in the histopathology and an exacerbation after sun exposure; but no injuries were blisters or erosions, the sign of Nickolsky was

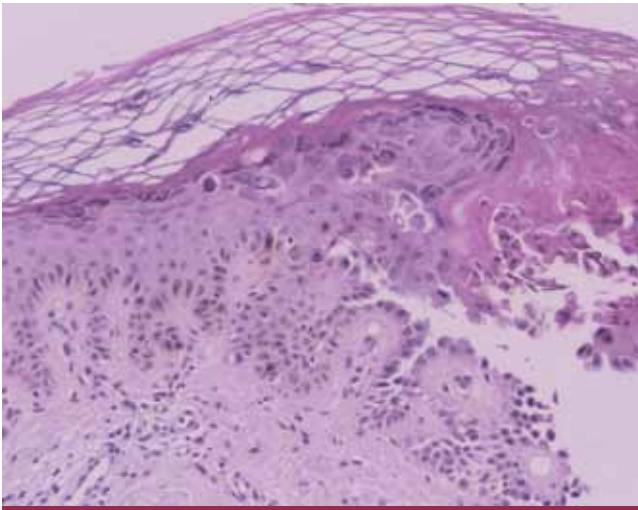


PHOTO 5. Details of the elongation of interpapillary crests, suprabasal acantholysis and dyskeratosis.

negative, no mucosal lesions were found and always had an overall good condition. Furthermore, suprabasal acantholysis was not only with rows of "tombstone-like" appearance, but was also present in spinosum stratum and pemphigus vulgaris showed no dyskeratosis, and immunofluorescence was negative. It was considered the possibility that residual lenticular lesions constituted post-inflammatory hyperpigmentation, but the histological finding of crests elongation and increased epidermal melanin level, rather than presenting melanophages, dismisses this possibility.

A special mention in the differential diagnosis deserves Galli-Galli disease, an acantholytic variant of Dowling-Degos disease. The latter is a rare genodermatosis, autosomal dominant, which is characterized by reticulate pigmentation that predominantly affects folds and flexures, associated with acne-like pimples and facial scars. Histopathological analysis shows elongation and hyperpigmentation of interpapillary crests.⁹ The acantholytic variant was described in 1982 by Bardach et al. and since then a total of 9 cases have been reported. Recently, Shabrawi-Cael et al. reported two cases which were interpreted as odd Galli-Galli disease variants.¹⁰

Both were mature females, with peeling erythematous papules, which worsened during summer months and associated to lentiginous macules, that instead of comprising folds, affected the trunk and the root of the members, and which the histopathology showed elongated interpapillary crests with acantholysis and dyskeratosis. It is likely that these two patients and ours had been affected by the same disease, as well as those previously reported as a variant of Grover's disease with lentiginous and photosensitivity. We believe that with time and further communications of this rare condition more knowledge will help to clarify and determine if it this belongs to a lentiginous variant of Grover's acantholytic dermatosis or to a new entity described by Cooper in 2004.

Bibliografía

1. Grover RW. Transient acantholytic dermatosis. *Arch Dermatol* 1970;101:426-434.
2. Chalet M, Grover RW, Ackerman AB. Transient acantholytic dermatosis: a reevaluation. *Arch Dermatol* 1977;113:431-435.
3. Davis MD, Dinnen AM, Landa N, Gibson LE. Grover's disease: clinicopathologic review of 72 cases. *Mayo Clin Proc* 1999; 74:229-234.
4. Parsons JM. Transient acantholytic dermatosis (Grover's disease): a global perspective. *J Am Acad Dermatol* 1996;35:653-666.
5. Quirk CJ, Heenan P. Grover's disease: 34 years on. *Australas J Dermatol* 2004;45:83-88.
6. Scheinfeld N, Mones J. Seasonal variation of transient acantholytic dyskeratosis (Grover's disease). *J Am Acad Dermatol* 2006;55:263-268.
7. Cooper SM, Dhittvat J, Millard P, Burge S. Extensive Grover's-like eruption with lentiginous freckling: report of two cases. *Br J Dermatol* 2004; 150:350-352.
8. Girard C, Durand L, Guillot B, Guilhou JJ, et al. Persistent acantholytic dermatosis and extensive lentiginous "freckling": a new entity? *Br J Dermatol* 2005;153:217-218.
9. Braun-Falco M, Volgger W, Borelli S, Ring J, et al. Galli-Galli disease: an unrecognized entity or an acantholytic variant of Dowling-Degos disease? *J Am Acad Dermatol* 2001; 45:760-763.
10. Shabrawi-Caelen L, Rüten A, Kerl H. The expanding spectrum of Galli-Galli disease. *J Am Acad Dermatol* 2007;56: s86-s91.