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Linear Darier Disease

Enfermedad de Darier segmentaria

To the Editor:

Darier disease is an autosomal dominant genodermatosis that usually appears between the ages of 10 and 20 years in seborrheic areas of the trunk and face; it can sometimes present a localized, linear, or zosteriform pattern. The



Figure 1 Patient 1.

presence of these patterns leads to controversy about the most suitable name, as some authors consider it a variant of Darier disease and others consider it an epidermal nevus with special histologic features.

We present 2 cases of linear Darier disease seen in our department. The first case was that of a 42-year-old man with no relevant personal or family history, who had had skin lesions located on the right chest for 25 years. The lesions had remained stable since onset and were asymptomatic, except with exercise, during which the erythema and pruritus increased. The second case was that of a 51-year-old man who was positive for human immunodeficiency virus category A3 and for 17 years had had slightly pruritic skin lesions located on the right abdomen; these lesions had become more apparent in recent months. In both patients we observed several grouped, round papules smaller than 5 mm that were erythematous-orange in color, keratotic, slightly infiltrated, and in some cases excoriated. The lesions had a metameric distribution in the right clavicular fossa and right subpectoral region in case 1 (Figure 1) and on the right abdomen in case 2 (Figure 2). No skin lesions in other areas or mucosal or nail lesions were observed in either of the 2 patients.



Figure 2 Patient 2.

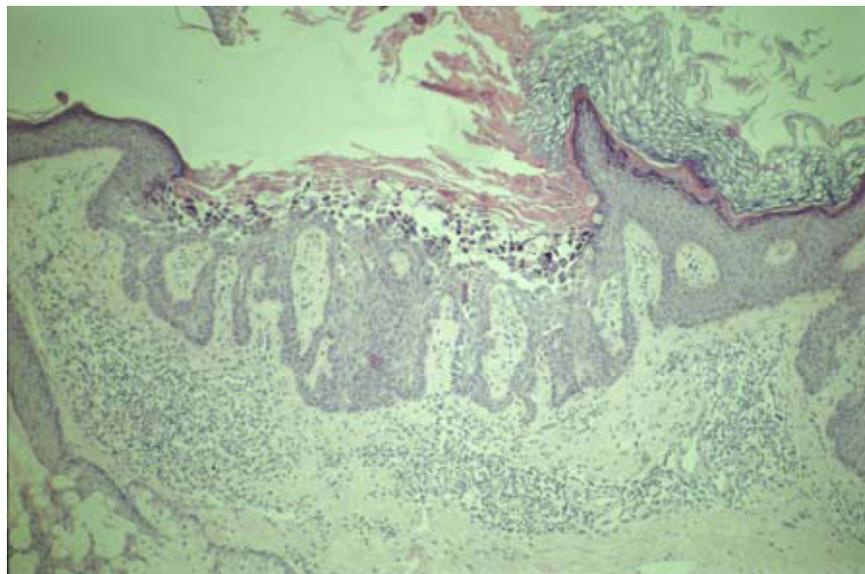


Figure 3 Hematoxylin-eosin, original magnification $\times 10$.

Biopsy of a chest papule showed a suprabasal intraepidermal blister that contained a large number of acantholytic and dyskeratotic cells (Figure 3).

Both patients underwent treatment with topical tazarotene, which was later suspended because it caused skin irritation and led to little improvement.

Darier disease is an autosomal dominant disorder of keratinization. It is caused by a mutation of the *ATP2A2* gene, which leads to premature keratinization of keratinocytes and loss of intercellular adhesion, with the consequent formation of suprabasal clefts. It presents clinically as keratotic papules that are erythematous-orange in color in seborrheic areas of the trunk and face and occasionally in skin folds. There may also be extracutaneous signs of the disease, such as whitish punctate lesions on the mucous membranes and longitudinal nail striations with subungual hyperkeratosis.

Histology (Figure 3) revealed hyperkeratotic and parakeratotic epidermis and the appearance of intraepidermal clefts containing acantholytic and dyskeratotic keratinocytes. This pattern of acantholytic dyskeratosis is not unique to Darier disease but also appears in Grover disease, verrucous dyskeratoma, and papular acantholytic dermatosis of the vulvocrural area. It has also been described as a histologic finding in melanocytic nevi, melanoma, lichen amyloid, granuloma annulare, and pityriasis rubra pilaris.¹

There are 2 clinical variants of this disease differentiated by the location of the lesions: acrokeratosis verruciformis, in which the lesions appear typically on the dorsum of the hands and feet, and linear Darier disease.

The term linear Darier disease is used when the lesions present in a zosteriform pattern. However, some authors prefer the term acantholytic dyskeratotic epidermal nevus, especially when the lesions are linear. In favor of the first name we find the study by O'Malley² of several patients

with linear or zosteriform lesions who also had ipsilateral nail lesions, and the study by Sakuntabhai³ of 2 patients presenting with zosteriform lesions who had *ATP2A2* mutations, revealing the existence of type 1 mosaicism, as described by Happle. Other authors,⁴ in the absence of a genetic study, defend the term acantholytic dyskeratotic epidermal nevus for cases in which other manifestations of Darier disease are absent, there is no relevant family history, and the onset is at birth or in childhood.

In our case, though no relevant family history was noted, we believe the appropriate term is linear Darier disease because the lesions first appeared in early adulthood and worsened with sweating. We also agree with Martínez et al,⁵ who recently reported a similar case in *Actas Dermosifiliográficas* and concluded that the description of the mutation in localized cases supports the use of this term. To date, the transmission of generalized Darier disease to the offspring of a patient with linear Darier disease has not been reported.⁶ This transmission would be possible in cases of gonadal mosaicism but in clinical practice the parents of patients diagnosed with generalized Darier disease are not examined.

Conflict of Interest

The authors declare that they have no conflict of interest.

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Solitary Congenital Self-Healing Histiocytosis (Hashimoto-Pritzker Disease)

Histiocitosis congénita autolimitada (Hashimoto-Pritzker) de presentación solitaria

To the Editor:

Congenital self-healing Langerhans cell histiocytosis (CSHLCH), or Hashimoto-Pritzker disease, is a rare variant of Langerhans cell histiocytosis that was first described in 1973 by Hashimoto and Pritzker.^{1,5} It presents at birth or in the neonatal period.^{1,9} Few case series have been published in the literature.^{1,4,5} However, it is believed that the actual incidence is higher and that many cases go unreported due to spontaneous resolution or the lack of clinical recognition.

We describe the case of a 2-month-old girl who presented a single lesion on the left thigh from birth. The lesion consisted of a desquamating indurated violaceous nodule with central ulceration covered by a serosanguineous scab (Figure 1). The biopsy showed a dermal and hypodermal proliferation of histiocytic cells with oval vesicular nuclei and abundant eosinophilic cytoplasm, accompanied by multinucleated giant cells (Figure 2). The histiocytic cells were positive for S-100, CD1a, and langerin (Figure 3). The laboratory workup and imaging tests ruled out systemic involvement, and the patient was diagnosed with CSHLCH. The lesion involuted spontaneously, and after 16 months of follow-up the patient is free of disease.

Langerhans cell histiocytosis (LCH) refers to a group of disorders characterized by cell proliferation in various organs; the cells are positive for S-100 and CD1a and contain Birbeck granules in the cytoplasm.^{1,2,4,5} In 1987 the Writing Group of the Histiocyte Society grouped 4 conditions together under the heading of LCH, according to clinical presentation: an acute, disseminated form (Letterer-Siwe disease), a multifocal chronic and progressive form (Hand-Schüller-Christian disease), a chronic localized form or eosinophilic granuloma, and a congenital form (Hashimoto-Pritzker disease). In 1997 the group published another

classification in which they recommended that only the term LCH be used and that patient grading be based on the extension of the disease.^{9,10}

CSHLCH is a rare disease characterized by painless reddish-brown papules or nodules that are present at birth or in the early months of life and involute within a few weeks or months.^{1,6} The lesions tend to be multiple and are most commonly distributed on the trunk, face, and scalp.^{4,5} Solitary lesions, first described by Berger et al. in 1986, are rare; since that initial description, only around 30 cases (25% of all reports) have been published.^{1,3,6} Lesions are rarely found in the oral mucosa or internal organs, although a few cases with systemic or mucosal involvement have been described, but these lesions regress along with the cutaneous one.^{1,5,7,9}

Histologically, the lesions are characterized by a superficial and deep dermal infiltrate composed of histiocytic cells with kidney-shaped nuclei and abundant eosinophilic cytoplasm. Inflammatory cells such as lymphocytes or eosinophils and multinucleated giant cells can also be observed.^{2,4,5}



Figure 1 Physical examination of the left thigh: ulcerated desquamating plaque covered by a serosanguineous scab.