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.
Positive reactions to S-100 and CD1a confirm the diagnosis of LCH, but do not allow differentiation between the various clinical forms.\textsuperscript{1-4,6,7} The introduction of immunohistochemistry techniques that detect the presence of Birbeck granules, such as langerin staining, has reduced the need for electron microscope study. However, a negative result with this staining technique, indicating the absence of Birbeck granules, does not invalidate the diagnosis because the granules are only detected in 50% of LCH cases and 10% to 30% of CSHLCH cases.\textsuperscript{1-4,7,8}

The differential diagnosis of CSHLCH includes most importantly cutaneous involvement by more severe forms of LCH, characterized by multisystem disease requiring aggressive treatment. Cutaneous lesions may be the initial manifestation and are similar to those found in CSHLCH; however, the lesions persist or worsen instead of regressing after a few months, and systemic symptoms start to develop.\textsuperscript{4,5,8,10} A solitary nodule with central necrosis, good overall health, and an absence of mucosal or systemic involvement suggest the diagnosis of a self-involuting form; nevertheless, there are no absolute clinical or histologic criteria to predict lesion behavior.\textsuperscript{5,10}

The prognosis of CSHLCH is excellent. No treatment is recommended, although clinical monitoring with imaging and laboratory follow-up are important because multisystem disease has been reported to develop up to 1 year after diagnosis.\textsuperscript{4,5,8} In 2008 Zunino-Goutorbe et al\textsuperscript{1} suggested that monthly clinical follow-up should be carried out for at least 6 months and every 2 to 3 months thereafter up to 2 years and that laboratory workup and imaging tests be performed at the time of diagnosis, that laboratory tests be repeated at 6 months, and that imaging techniques be reserved for cases with clinical manifestations.

References

Nevus Psiloliparus in a Child with Encephalocraniocutaneous Lipomatosis
Nevo psilolíparo en un niño con lipomatosis encefalocraneocutánea

To the Editor:

Nevus psiloliparus—from the Greek psilos (‘hairless’) and liparos (‘fatty’)—is a term coined by Happle to describe a particular adipose tissue nevus that develops in patients with encephalocraniocutaneous lipomatosis (ECCL).\(^1\) The nevus clinically presents as a slightly raised plaque, with well-defined and sometimes irregular borders, and with slightly yellowish or normal coloring.\(^2\) The unilateral lesion is normally located in the frontal or frontoparietal region, and its most characteristic feature is the paucity or absence of hair follicles.\(^2\) Histologically, nevus psiloliparus has 3 main signs: an abundance of nonencapsulated mature adipose tissue, which can produce compression and thinning of the dermis; a paucity or absence of mature hair follicles; and the presence of normal quantities of orphaned arrector pili muscle bundles (that is, arrector pili muscles independent of or unassociated with hair follicles) arranged in a row parallel to the epidermal surface. The sebaceous glands may be normal or enlarged.

We describe a case of nevus psiloliparus associated with characteristic ECCL traits.

The 2-month old baby boy was born to healthy and nonconsanguineous parents following a normal twin pregnancy with caesarean delivery at term. His identical twin brother was entirely healthy. The patient was brought to the dermatology clinic due to the presence of an area of congenital alopecia on the scalp. Physical examination revealed the right parietocipital region to have a skin-colored, well-defined, tear-shaped plaque, with a smooth surface and soft consistency, and with reduced capillary density (Figure 1A). In addition, a yellowish papule with well-defined borders and an elastic consistency was observed on the medial third of the right upper eyelid, and a translucent, yellowish plaque with well-defined borders and with telangiectasia was observed at the corneoscleral junction of the right eye (Figure 1B). These signs were suggestive of choristoma of the bulbar conjunctiva. Examination revealed no other lesions, masses, or organomegaly, and no lymphadenopathies in areas accessible to palpation. Biopsy of the scalp lesion showed a normal epidermis, a thinned dermis with a paucity of hair follicles, bundles of smooth muscle arranged parallel to the skin surface, and a disproportionate amount of nonencapsulated mature adipose tissue in the hypodermis (Figure 2). Nuclear magnetic resonance imaging performed when the infant was 3 months old revealed enlargement

Figure 1  A, Slightly raised and somewhat irregular plaque with well-defined borders, and with a clear reduction in hair follicle density. B, Well-defined, slightly yellowish papule measuring 4 mm in diameter on the medial third of the right upper eyelid, and a translucent, yellowish-pink plaque with superficial telangiectasia extending across the corneoscleral junction.