

Anna Carolina Miola ^{a,*}, Paulo Muller Ramos ^b,
Hélio Amante Miot ^a

^a Department of Dermatology, Faculty of Medicine, Universidade Estadual Paulista, Botucatu, SP, Brazil

^b Department of Dermatology, Faculty of Medicine, Universidade Estadual Paulista, Botucatu, SP, Brazil

* Corresponding author.

E-mail: anna.c.miola@unesp.br (A.C. Miola).

Received 7 October 2021; accepted 9 November 2021;

Available online 7 March 2023

<https://doi.org/10.1016/j.abd.2021.11.013>

0365-0596/ © 2023 Sociedade Brasileira de Dermatologia.

Published by Elsevier España, S.L.U. This is an open access article under the CC BY license (<http://creativecommons.org/licenses/by/4.0/>).

Congenital ulcerated nodule: self-healing Langerhans cell histiocytosis[☆]



Dear Editor,

A full-term male newborn born by vaginal delivery, after an uncomplicated gestation, was observed at our department on the first day of life for a congenital cutaneous lesion. At physical examination, we observed on the left scapular region an 8 mm ulcerated nodule, with a central black crust, a bright elevated pink border, and a peripheral erythema (Fig. 1). Dermoscopy revealed a central reddish-black crust, with a rim of sparse red globules, and a pink border with fine white scale (Fig. 2). No other lesions were evident, and the remaining physical examination had no abnormalities. Eye red reflex and otoacoustic emissions screenings were normal. His family history was unremarkable. A punch biopsy was made.

Histopathology showed an ulcerated lesion with an infiltrate composed of large epithelioid cells with a kidney-shaped nucleus, with epidermotropism (Fig. 3). Mitotic figures were observed. The background infiltrate was composed of lymphocytes, plasma cells and a significant number of eosinophils (Fig. 4). Immunostaining was positive for CD1a (Fig. 5A), langerin (Fig. 5B), S100 protein and CD45, and negative for CD68 and CD34. A presumptive diagnosis of solitary congenital self-healing Langerhans cell histiocytoma was made. Complete Blood Count (CBC), Erythrocyte Sedimentation Rate (ESR), coagulation times, kidney and liver function tests and lactate dehydrogenase were within the normal range for the age. The newborn underwent chest radiography and abdominal ultrasound, which revealed no abnormalities.

The infant maintains regular follow-ups. At 6 months appointment, no systemic involvement was noticed and there was a complete involution of the skin lesion. The overall well-appearing state of the neonate, lack of systemic signs, and spontaneous involution of the lesion, associated with the histopathology and immunohistochemistry findings, were compatible with congenital self-healing histiocytosis.

Discussion

Langerhans cell histiocytosis (LCH) is a rare neoplasm, characterized by a pathological Langerhans cell proliferation.¹ LCH can present as a single organ or multi-systemic involvement, with a wide spectrum of manifestations, ranging from self-resolving skin lesions to disseminated forms.² The prognosis depends on the extent of systemic involvement, with the single-system disease having a good prognosis.²

Congenital self-healing Langerhans cell histiocytosis (CSHLCH) is a rare variant of LCH.³ It usually presents as multiple papules or nodules (from the ‘‘blueberry muffin baby’’ spectrum), but uncommonly a solitary lesion may occur.^{3,4} LC histiocytoma (or solitary CSHLCH) is considered a unimodular or paucinodular CSHLCH variant. It presents as a single reddish nodule at birth or within the first weeks of life, that



Figure 1 Ulcerated lesion with central crust and elevated pink border located on the left scapular region.

[☆] Study conducted at the Department of Dermatology of Centro Hospitalar de Leiria, Leiria, Portugal.

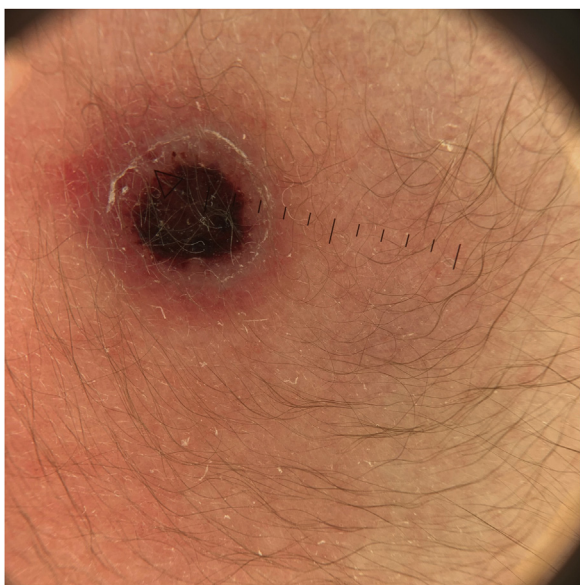


Figure 2 Dermoscopy with central reddish/black crust, a pink border with fine white scale and sparse red globules between the crust and the border.

progresses to crusting and ulceration.⁵ Cutaneous lesions typically regress within a few months.⁶ Neonates are usually healthy, with no systemic involvement.^{5,6} A recent retrospective review of 82 neonates showed complete involution of the lesion in all cases who have not been submitted to excision. Furthermore, the authors observed no progression, recurrence, or multisystemic involvement, with a median follow-up of 15 months.⁶

Histopathology is characterized by a dense dermal infiltrate composed of large cells with abundant eosinophilic

cytoplasm and round-oval or kidney-shaped nucleus, accompanied by an infiltrate comprising lymphocytes and eosinophils.^{4,6} Histopathological differences have been between solitary CSHLCH and multiple papular or nodular forms. In the former, there is a deeper infiltrate of the whole dermis and often extends to superficial subcutis, superficial ulceration or crusting, frequent parakeratosis and necrosis, and little or no epidermotropism. In solitary CSHLCH, there are large-sized cells, sometimes with "ground-glass" cytoplasm and numerous mitoses.⁵ Diagnosis is confirmed by positive immunostaining for CD1a, S100, and langerin, markers of Langerhans cells. Electron microscopy reveals the characteristic Birbeck granules,⁶ but usually is not needed. There is some variability in dermoscopy reports of LCH, due to the clinical spectrum and different lesion stages. Reddish-lilac color of nodular lesions, with peripheral telangiectasias, violaceous lacunae and clods, and whitish areas may be present.^{7,8} However, dermoscopy descriptions are sparse in the literature.

There are no specific guidelines for the management and follow-up of solitary CSHLCH. Initial work-up should include a complete physical examination (exclude other skin or mucosal lesions, lymphadenopathy, or hepatosplenomegaly), laboratory analysis (CBC, ESR, C-reactive protein, coagulation studies, hepatic and renal function tests, blood and urine osmolarities) and imaging exams (abdominal ultrasound, chest X-Ray, complete skeletal radiographs).⁹

In summary, solitary CSHLCH diagnosis should be raised in the presence of a single ulcerated congenital lesion and usually portends a good prognosis. CSHLCH diagnosis is made in retrospect after work-up and follow-up of these patients,¹⁰ showing a spontaneous resolution and absence of systemic involvement.

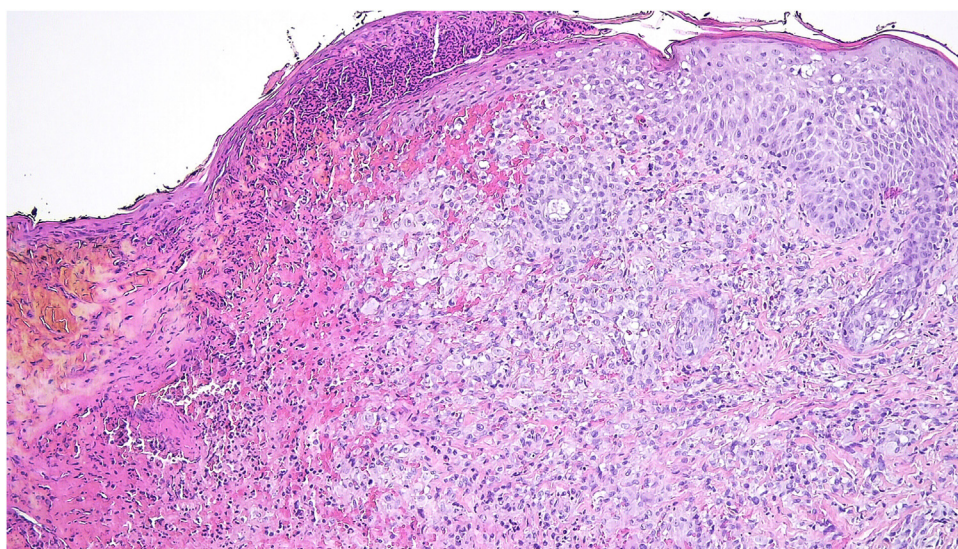


Figure 3 Ulcerated lesion with an infiltrate composed of epithelioid cells with kidney-shaped nucleus, with epidermotropism (Hematoxylin & eosin, $\times 100$).

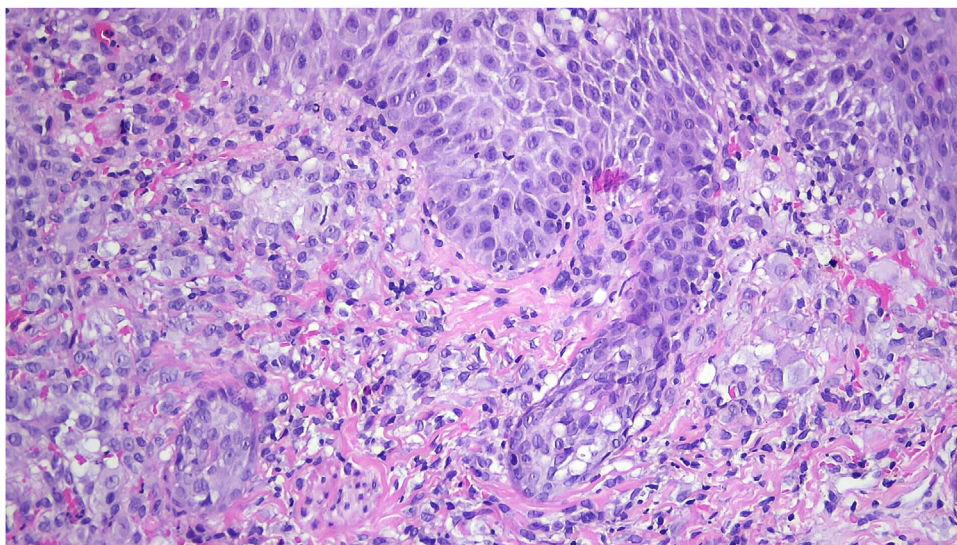


Figure 4 Infiltrate composed of epithelioid cells with kidney-shaped nucleus, accompanied by lymphocytes, plasma cells and a significant number of eosinophils (Hematoxylin & eosin, $\times 200$).

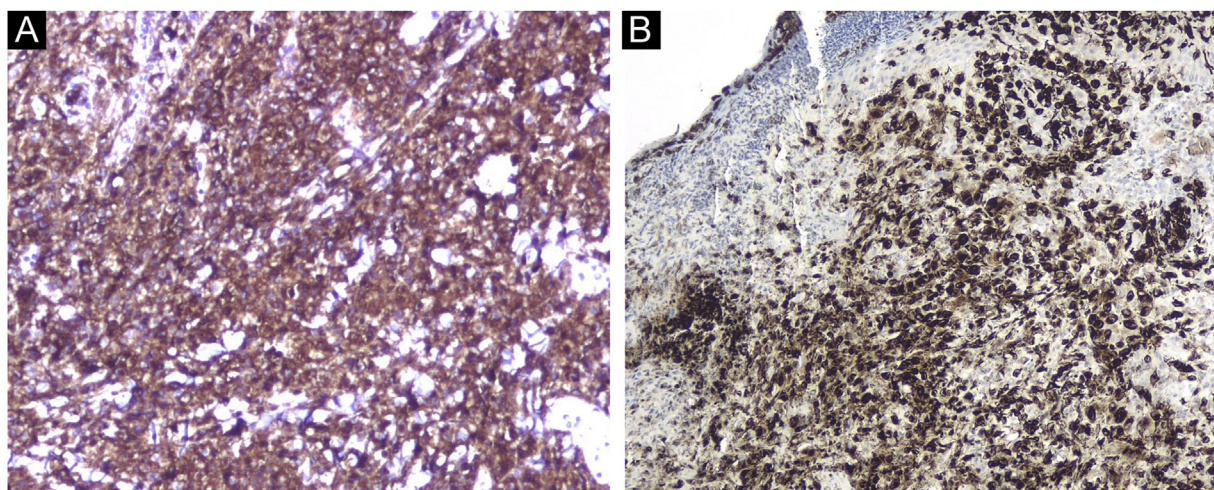


Figure 5 Positive immunostaining for CD1a (A, $\times 200$) and langerin (B, $\times 100$).

Financial support

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Authors' contributions

Tiago Fernandes Gomes: Data collection, analysis and interpretation; critical literature review; preparation and writing of the manuscript.

José Carlos Cardoso: Approval of the final version of the manuscript; data collection, analysis and interpretation; manuscript critical review.

Victoria Guiote: Approval of the final version of the manuscript; data collection, analysis, and interpretation; manuscript critical review.

Felicidade Santiago: Approval of the final version of the manuscript; data collection, analysis, and interpretation;

intellectual participation in propaedeutic and/or therapeutic management of studied cases; manuscript critical review.

Conflicts of interest

None declared.

References

1. Satter EK, High WA. Langerhans cell histiocytosis: a review of the current recommendations of the histiocyte society. *Pediatr Dermatol.* 2008;25:291–5.
2. Dhar S, Srinivas SM, Dhar S, Basu S, Banerjee R, Malakar R, et al. Langerhans cell histiocytosis in children: a retrospective case series of 126 cases. *Pediatr Dermatol.* 2020;37:1085–9.
3. Hansel K, Tramontana M, Troiani S, Benedictis D, Bianchi L, Cucchia R, et al. Congenital self-healing langerhans cell histiocytosis: a rare presentation of Blueberry Muffin Baby "Spectrum". *Dermatopathology.* 2019;6:37–40.

4. Ungari M, Caresana G, Tanzi G, Drera B, Frittoli MC, Gusolino MD, et al. Congenital unilesional cutaneous langerhans cell histiocytosis: a case report. *Am J Dermatopathol.* 2020; 00:1–5.
5. Dupeux M, Boccarda O, Frassati-Biaggi A, Hélias-Rodzewicz Z, Leclerc-Mercier S, Bodemer C, et al. Langerhans cell histiocytoma: a benign histiocytic neoplasm of diverse lines of terminal differentiation. *Am J Dermatopathol.* 2019; 41:29–36.
6. Schwartz Z, Bender A, Magro CM. Solitary congenital Langerhans cell histiocytoma: a pattern of benign, spontaneous regression in patients with single lesion disease. *Pediatr Dermatol.* 2020;37:1009–13.
7. Micali G, Verzi AE, Quattrocchi E, Ng CY, Lacarrubba F. Dermoscopy of common lesions in pediatric dermatology. *Dermatol Clin.* 2018;36:463–72.
8. Pigem R, Dyer A, Podlipnik S, Carrera C, Puig S, Ferrando J. Dermoscopy of congenital langerhans cell histiocytosis. *Dermatol Pract Concept.* 2020;10:e2020063.
9. Zunino-Goutorbe C, Eschard C, Durlach A, Bernard P. Congenital solitary histiocytoma: a variant of Hashimoto-Pritzker histiocytosis. *Dermatology.* 2008;216:118–24.
10. Simko SJ, Garmezy B, Abhyankar H, Lupo PJ, Chakraborty R, Lim KPH, et al. Differentiating skin-limited and multisystem langerhans cell histiocytosis. *J Pediatr.* 2014;165:990–6.

Tiago Fernandes Gomes ^{a,*}, José Carlos Cardoso ^b,
Victoria Guiote ^a, Felicidade Santiago ^a

^a Department of Dermatology, Centro Hospitalar de Leiria, Portugal

^b Department of Dermatology, Centro Hospitalar e Universitário de Coimbra, Portugal

* Corresponding author.

E-mail: tiagofgomesd@gmail.com (T.F. Gomes).

Received 4 October 2021; accepted 22 December 2021;

Available online 26 April 2023

<https://doi.org/10.1016/j.abd.2021.12.012>

0365-0596/ © 2023 Published by Elsevier España, S.L.U. on behalf of Sociedade Brasileira de Dermatologia. This is an open access article under the CC BY license (<http://creativecommons.org/licenses/by/4.0/>).

Dermoscopy of circumscribed palmar hyperkeratosis[☆]



Dear Editor,

A 71-year-old female patient of Asian descent was referred to the Dermatology Outpatient Clinic for treatment of seborrheic keratoses. Two years later, in one of the consultations, an asymptomatic lesion on the left thenar region was observed and described, characterized at the time as possible excoriation. However, due to the persistence and increase in the size investigation of the lesion was initiated. The patient denied local trauma. The lesion remained asymptomatic, measuring 1.6 × 1.2 cm, characterized by a single, rounded area, with a depressed center and slightly erythematous, non-scaling, with well-defined and raised borders (Fig. 1).

Dermoscopy was performed using the Dermlite DL4[®] device (3 Gen, San Juan Capistrano, USA) at level 0 (10×), without immersion gel. The central region of the lesion showed white dots, red dots and whitish striae, with regular and symmetrical distribution on a light pink background. In addition, a border with peripheral desquamation similar to a “step ladder” was observed (Figs. 2–4). The proposed diagnostic hypotheses were Bowen’s disease, porokeratosis, granuloma annulare, and self-inflicted trauma.

A skin biopsy was performed, and histopathology revealed a well-circumscribed area in which there was a decrease in the stratum corneum (Fig. 5). After evaluating the literature and considering the result of the anatomopathological examination, the authors considered

circumscribed palmar hyperkeratosis. However, the patient was lost to follow-up as of the submission of this article.

Discussion

In 2002, Perez et al. published a series of ten cases, in which they described a new entity, characterized by solitary, asymptomatic, circular, circumscribed, erythematous and depressed palmar or plantar lesions – mainly on the thenar and hypothenar eminences of the palms. At the time, the authors described it as an epidermal malformation, calling it circumscribed palmoplantar hyperkeratosis (CPH) and reported that it is more prevalent in elderly patients and women, preferentially affecting the palms.¹

Since then, about 100 cases have been described, but their etiology remains unknown. In 2013, Rocha & Nico described the first cases in Brazil, corroborating the main findings of Perez et al., although one of the cases had more than one lesion.²

The first dermoscopic description of CPH in the literature was performed by Ishiko et al.,³ in 2007, reporting two cases in Asian women. Since then, the reports have grown in frequency, containing descriptions similar to the case reported herein: “step-ladder-like” desquamation on the periphery of the lesion, well-demarcated erythema, with scattered white dots, which correspond to the acrosyringia, and small reddish dots, which could correspond to enlarged capillaries.^{3–7} Vilas Boas da Silva et al. described the presence of whitish striae in two of their three cases reported.⁸

Several etiological hypotheses have been made since the first report by Perez et al. – which suggested the possibility of a benign clonal epidermal malformation,¹ – such as dynamic proliferative disorder, a primary disorder of keratinization located in the granular layer and stratum corneum, trauma, infection of keratinocytes by HPV, and

[☆] Study conducted at the Dermatology Clinic, Santa Casa de Misericórdia de São Paulo, São Paulo, SP, Brazil.