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Antônia Larangeira de Almeida: Approval of the final version of the manuscript; design and planning of the study; drafting and editing of the manuscript; collection, analysis and interpretation of data; critical review of the literature; critical review of the manuscript.

Conflicts of interest

None declared.

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Hiram Larangeira de Almeida Jr. [a,b,*], Fernanda Pinto Garcia [c], Laura de Moraes Gomes [d], Antônia Larangeira de Almeida [b]

[a] *Department of Dermatology, Universidade Católica de Pelotas, Pelotas, RS, Brazil*
[b] *Department of Dermatology, Universidade Federal de Pelotas, Pelotas, RS, Brazil*
[c] *Hospital Erasto Gaertner, Curitiba, RS, Brazil*
[d] *Department of Radiology, Universidade Federal de Pelotas, Pelotas, RS, Brazil*

* Corresponding author.
E-mail: hiramalmeidajr@hotmail.com (H.L. Almeida Jr.).

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Case for diagnosis. Cicatricial alopecia on the vertex - Folliculitis decalvans and lichen planopilaris phenotypic spectrum☆

Dear Editor,

A 34-year-old white female complained of vertex alopecia for six years, associated with pruritus and local burning sensation. She reported that, since the onset of the condition, she observed the development of pustules at the periphery of the lesion. She denied comorbidities or ongoing medication use. She reported having had cycles of trimethoprim-sulfamethoxazole for 60 to 90 days, with partial improvement. The examination disclosed cicatricial alopecia area on the vertex region of the scalp, with follicular pustules at the periphery, erythema and areas with polytrichia (Fig. 1). Dermoscopy revealed follicular pustules, polytrichia, perifollicular desquamation and erythema (Fig. 2). A biopsy of the peripheral region (area with disease activity) was performed, and histopathology showed a mixed perifollicular inflammatory infiltrate, rich in plasmocytes and histiocytes, affecting the region between the isthmus and the infundibulum, with follicular destruction (Figs. 3 and 4).

☆ Study conducted at the Dermatology Outpatient Clinic, Faculty of Medicine, Universidade Estadual Paulista, Botucatu, SP, Brazil.

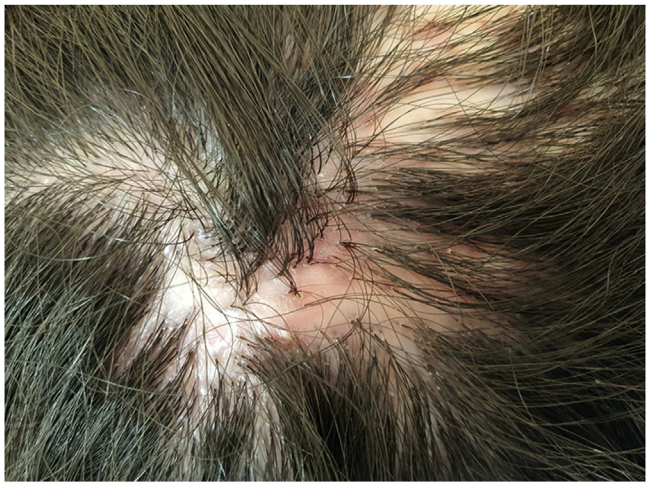

Figure 1 Cicatricial alopecia, with follicular pustules at the periphery, erythema and perifollicular desquamation, in addition to areas with polytrichia.

What's your diagnosis?

a) Folliculitis decalvans
b) Folliculitis decalvans and lichen planopilaris phenotypic spectrum
c) Centrifugal cicatricial alopecia
d) Lichen planopilaris

Discussion

Folliculitis decalvans and lichen planopilaris phenotypic spectrum (FDLPPPS) is a recently described cicatricial alopecia, more prevalent on the vertex of the scalp in adults, which combines clinical and histopathological features of two other alopecias: lichen planopilaris (LPP) and folliculitis decalvans (FD), presenting with lymphocytic and neutrophilic inflammatory infiltrates, respectively.[1]

The pathogenesis of LPP and FD is still poorly understood; however, whereas in LPP there is activation of TCD8+ lymphocytes and loss of follicular immune privilege, in DF the

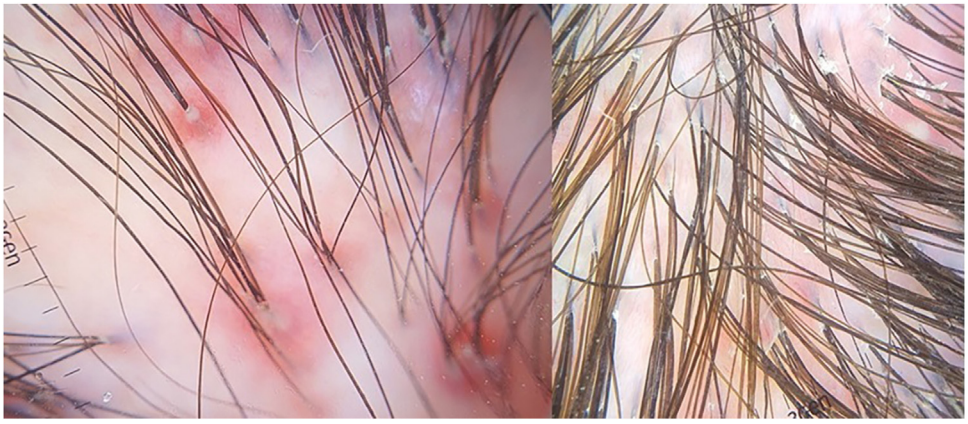

Figure 2 Follicular pustules, desquamation and perifollicular erythema on dermoscopy. Areas with absence of follicular ostia; however, with the presence of polytrichia.

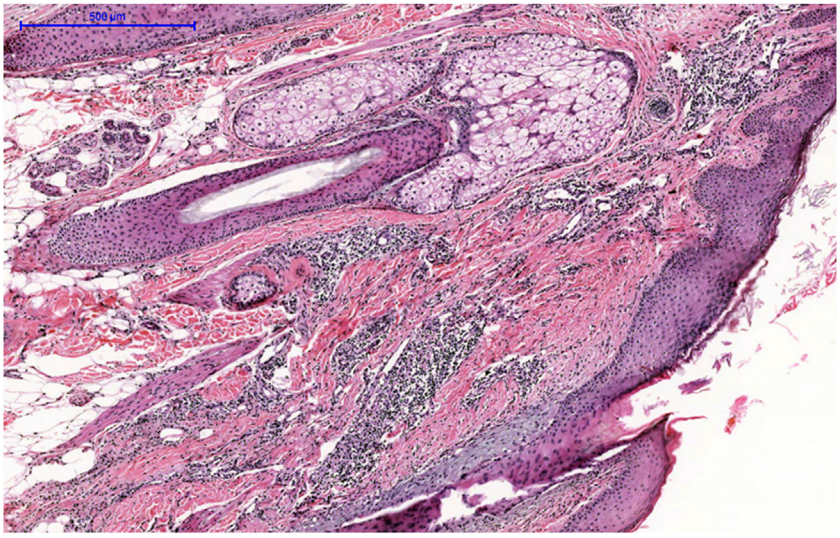

Figure 3 Mixed perifollicular inflammatory infiltrate in the region between the isthmus and the infundibulum, with follicular destruction (Hematoxylin & eosin, ×100).

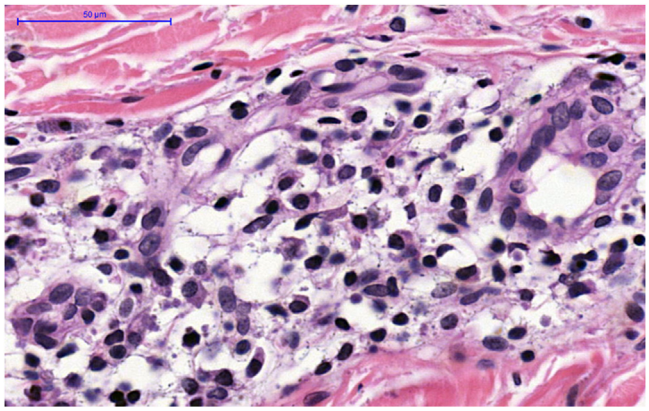

Figure 4 Presence of numerous plasma cells and histiocytes amidst the mixed perifollicular infiltrate (Hematoxylin & eosin, ×400).

presence of *Staphylococcus aureus* induces a biofilm that stimulates the innate immune response, perpetuating the neutrophil-mediated inflammatory process.[2,3]

The etiology of FDLPPPS is still under discussion, given the coexistence (or alternation) of both inflammatory processes. It has been suggested that dysbiosis of the DF follicular microbiome may induce the exposure of follicular autoantigens, stimulating a Th1 response pattern.[1,4]

As for the clinical picture, patients with FDLPPPS usually show a sequential biphasic process with the characteristics of FD preceding LPP, or even concomitant clinical characteristics: areas of cicatricial alopecia with follicular pustules, erythema and perifollicular desquamation, which may evolve with polytrichia, as in the present case.[5,6]

Regarding histopathology, a mixed infiltrate is observed in the infundibulum region, with destruction and atrophy of the follicular epithelium and prevalence of plasma cells and histiocytes, in contrast to the predominance of neutrophils in the FD and lymphocytes in the LPP.[4,7,8]

There is yet no standardized treatment for FDLPPPS, given its recent description. However, treatments used in FD and LPP are usually associated, such as corticosteroids, sulfonamides, doxycycline, retinoid antimalarials, and immunosuppressants, according to the predominance of clinical and trichoscopic characteristics. In this case, oral hydroxychloroquine 400 mg/d and isotretinoin 30 mg/d, and clobetasol 0.05% gel were introduced, with stabilization of the condition after six months. The rationale for choosing anti-inflammatory drugs and oral retinoids was based on the predominance of clinical and trichoscopic signs of the LPP spectrum, in this case, mainly after previous treatment with antibiotics, aiming to reduce the neutrophil activity intensity.[1,9] However, it is noteworthy the lack of controlled studies on the efficacy of therapeutic strategies in cicatricial alopecia.

The cases described in adults and children with FDLPPPS do not seem to reach a large area of the scalp, despite the delay in its diagnosis.[1,4–8]

Dermatologists should be aware of the diagnosis of FDLPPPS in the presence of less characteristic cases of cicatricial alopecia on the vertex of the scalp, with pustules, erythema and follicular desquamation, in which the inflammatory infiltrate is mixed, or with alternating patterns in subsequent biopsies, and containing plasma cells.

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Authors' contributions

Miola AC: Design and planning of the study; collection, analysis and interpretation of data; critical review of the literature; critical review of the manuscript; writing and approval of the final version of the manuscript.

Ramos PM: Critical review of the literature; critical review of the manuscript, writing and approval of the final version of the manuscript.

Miot HA: Design and planning of the study; effective participation in research orientation; critical review of the literature; critical review of the manuscript, writing and approval of the final version of the manuscript.

Conflicts of interest

None declared.

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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).

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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.

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

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

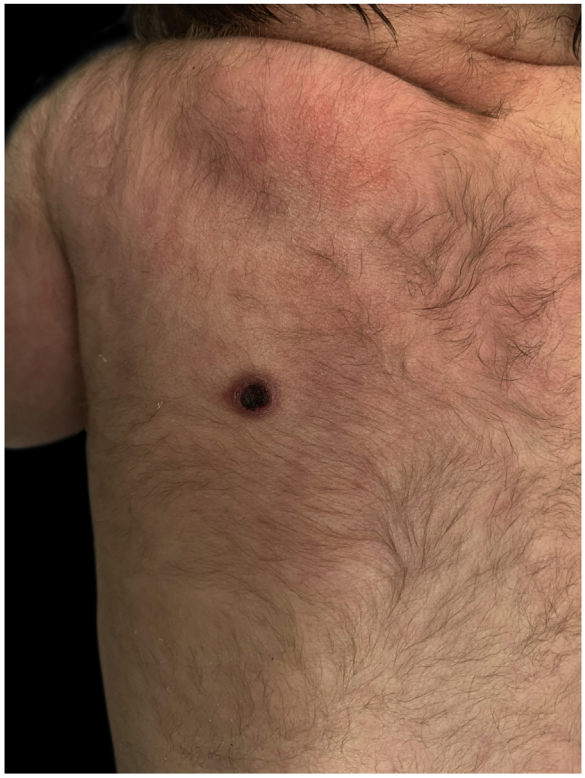

Figure 1 Ulcerated lesion with central crust and elevated pink border located on the left scapular region.