CASE LETTER

Bullous congenital diffuse cutaneous mastocytosis

Dear Editor,

Mastocytosis is a heterogeneous group of disorders whose signs and symptoms are characterized by mast cells infiltrate in tissues. There are two main variants, cutaneous mastocytosis (CM) that is limited to the skin, and systemic mastocytosis (SM) in which extracutaneous organs are affected, such as the bone marrow, liver, spleen and lymphoid tissue. The skin is the most commonly affected organ, and in children it is often the only one. Studies suggest that in 15–31% of all patients the disease is congenital. Mastocytosis is also classified into two groups according to the age of onset: childhood and adult mastocytosis. The adult mastocytosis tends to progress to SM with worse prognosis, whereas childhood mastocytosis rarely progresses to a SM and tends to improve during adolescence. Diffuse cutaneous mastocytosis (DCM) is a rare and severe variant that typically presents in the neonatal period with a controversial prognosis.

A full-term male newborn, with no significant family or gestational history, presented at birth diffuse erythematous leathery plaques distributed throughout the body (Fig. 1). Physical examination did not present alterations in other organs or systems. On the third day of life, there were tense bullae (filled with yellowish fluid) in the cephalic segment, upper and lower limbs and anterior trunk. In addition, the erythematous plaques became brownish, with peau d’orange appearance (Fig. 2). The spread of new bullous lesions was related to the heat of the incubator, handling with latex gloves and contact with clorexdine during hygiene. Laboratory and imaging investigations did not show any evidence of abnormalities.

At that moment, prednisone 1 mg/kg/day was introduced and a skin biopsy performed. The skin biopsy revealed numerous of diffuse mast cells cutaneous infiltrates stained in metachromically toluidine blue staining, confirming the diagnosis of mastocytosis (Fig. 3). Immunohistochemistry CD117 (c-kit) evidenced massive positivity of mast cells

Figure 1 Erythematous plaques present at birth.

Figure 2 Erythematous brownish plaques on lower limb, topped by blisters at different stages of development (stray blisters, flaccid blisters and exulcerations).

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Study conducted at the Hospital e Maternidade Santa Joana, São Paulo, SP, Brazil.
Reliable prognostic criteria are still lacking to predict the risk of systemic involvement in DCM and maculo-papular mastocytosis. Most cases of DCM resolve spontaneously in childhood or adolescence, however these patients are at a greater risk of complications such as hypotension, anaphylaxis and diarrhea and they should be monitored regularly.

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Authors’ contributions
Julia Marcon Cardoso: Approval of the final version of the manuscript; conception and planning of the study; elaboration and writing of the manuscript; critical review of the literature; critical review of the manuscript.
Camila Angelico S. Cabral: Conception and planning of the study; critical review of the literature.
Rute Facchini Lellis: Approval of the final version of the manuscript; obtaining, analysis, and interpretation of the data.

Conflicts of interest
None declared.

References

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