Abstract: Congenital self-healing reticulohistiocytosis is the benign spectrum of Langerhans Cell Histiocytosis, characterized by cutaneous lesions at birth or in the neonatal period, absence of systemic manifestations and spontaneous resolution of clinical status. Despite the benign and often self-resolving course in most patients, studies show that in some cases there may be metastasis or recurrence of the disease, emphasizing that the clinical course is variable, requiring long-term follow-up. The monitoring of the patient for a long period is important to detect possible systemic involvement, as there is a report of recurrence involving the skin, mucosa, bone and pituitary gland.

Keywords: Histiocytosis; Histiocytosis, Langerhans-Cell; Histiocytosis, Non-Langerhans-Cell

Resumo: A retículo-histiocitose congênita autolimitada é o espectro benigno das histiocitoses de células de Langerhans, caracterizada pela presença de lesões cutâneas ao nascimento ou no período neonatal, ausência de manifestações sistêmicas e resolução espontânea do quadro clínico. Apesar de curso benigno e frequente autorresolução na maioria dos pacientes, estudos mostram que, em alguns casos, pode haver disseminação ou recaída da doença, enfatizando que o curso clínico é variável, havendo necessidade de seguimento em longo prazo. O acompanhamento do paciente por longo período é importante para detectar possível envolvimento sistêmico, pois existe relato de recorrência, envolvendo pele, mucosa, ossos e glândula pituitária.

Palavras-chave: Histiocitose; Histiocitose de células de Langerhans; Histiocitose de células não Langerhans

Received on 21.06.2010.
Approved by the Advisory Board and accepted for publication on 21.07.2010.

Conflict of interest: None / Conflito de interesse: Nenhum
Financial funding: None / Suporte financeiro: Nenhum
INTRODUCTION

Langerhans cell histiocytosis (LCH) is a generic term that encompasses clinically distinct diseases, but that present as a point in common clonal proliferation of Langerhans cells with positive Birbeck granules and S100/CD1A in immunohistochemistry tests. It is considered a rare disease, with an annual incidence rate of three to four cases per million in pediatric patients. The most common clinical manifestations are osteolytic lesions, lymphadenomegaly and skin lesions. The skin lesions are frequently the first manifestation of LCH and cutaneous involvement is observed as the only disease site in approximately 10% of cases. A congenital self-healing reticulohistiocytosis of Hashimoto-Pritzker is the benign spectrum of Langerhans cell histiocytosis, characterized by the presence of skin lesions at birth or in the neonatal period, absence of systemic manifestations and spontaneous resolution of the clinical picture.

CASE REPORT

A full-term female infant, delivered by cesarean section, presented papulonodular reddish-brown erythematous lesions and crusts of different sizes located predominantly on the face, distributed on the trunk, upper and lower limbs (Figures 1 and 2). The physical examination did not otherwise reveal any abnormalities. Routine laboratory tests did not show alterations. The serological tests done for the TORCH group (toxoplasmosis, rubella, cytomegalovirus, herpes and syphilis) were negative. Skull, chest and long bone X-rays and abdominal ultrasound were normal. The skin biopsy revealed histiocyte aggregates with granulomatous formation, surrounding vases and annexes with a great quantity of infiltrated erythrocytes. The aggregates are disposed in the middle and deep reticular dermis, quite distant from the papillary dermis and without any epidermis involvement (Figure 3). Some of the histiocytes have reniform nuclei, while others have small granules in the cytoplasm. FITE and GROCOTT staining were negative for microorganisms. Immunohistochemical tests revealed positivity for CD1A/S100 (Figure 4). The lesions completely regressed in five weeks (Figure 5). With these findings the diagnosis of congenital self-healing reticulohistiocytosis of Hashimoto Pritzker was completed. The patient has been under follow-up for two years and up to the present moment has not shown signs of recidivation of the disease.

DISCUSSION

Congenital self-healing reticulohistiocytosis was described in 1973 by Hashimoto-Pritzker. The disease has as characteristic the presence of cutaneous lesions such as papules, nodules and vesicles, usually at birth or in the neonatal period, without systemic involvement. Histopathology presents infiltrate of Langerhans cells, with a positive immunohistochemical profile for S100/CD1A and spontaneous resolution in the first year of life. Spontaneous involution and absence of systemic symptoms are marked characteristics for differentiation with the other clinical spectra of Langerhans cell histiocytoses.

Most cases are limited to the skin, but there are reports of cutaneous symptom complex accompanied by pulmonary and ophthalmic involvement. Despite the benign course and frequent sponta-
neous resolution in most of the patients with this form of LCH, studies show that in some cases there may be dissemination or relapse of the illness, demonstrating that the clinical course is variable and that a long-term follow-up is required. 4,10,11

Patients with LCH in a single compromised system present excellent survival rate prognosis and no reports of death have been found in the literature concerning this group of patients. 12

There is no specific treatment for congenital self-healing reticulohistiocytosis. The conduct recommended by the Histiocyte Society consists in following the clinical picture and awaiting spontaneous regression. If the lesions persist, topical corticosteroids or topical nitrogen mustard may be effective. In case of cutaneous recurrence, the adopted conduct is the same: await spontaneous resolution of the symptoms, which presents favorable results. In cases of systemic recurrence, chemotherapy with vinblastine or etoposide, with or without corticosteroids is recommended. 13,14,15

It is important to follow the patient for long periods to detect possible systemic involvement, since there are reports of recurrence involving skin, mucosa, bones and pituitary gland. 4

Differential diagnosis should be made comparing it with cytomegalovirosis, candidiasis, varicella, herpes simplex, neonatal toxic erythema, infantile
acropustulosis, pigmentary incontinence, eosinophilic pustular folliculitis, neonatal erythropoiesis, disseminated neonatal hemangiomatosis and congenital leukemia cutis.\textsuperscript{15}

As this is a rare disease with very variable clinical behavior, it is of the essence to establish uniform clinical and histopathological criteria for diagnosis and patient stratification. According to the Histiocyte Society, the definitive diagnosis requires the demonstration of Birbeck granules in the histiocytes by means of electronic microscopy or surface antigen CD1a positivity by immunohistochemistry.\textsuperscript{13,3}

Actual incidence should be higher than observed in literature, which is justified by the fast spontaneous resolution.\textsuperscript{5}

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