Case for diagnosis*

Caso para diagnóstico

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CASE REPORT

A 1-month-old boy, born at term after an uneventful pregnancy, with a birth weight of 3,480g. First child of healthy and unrelated parents. At birth his mother noticed a 1 x 1.5 cm erythematous, infiltrated, perianal plaque that rapidly progressed to ulceration (Figure 1). The patient was otherwise healthy. A second evaluation at age two and a half months showed a complete and spontaneous resolution of the lesion (Figure 2). Serologic test for syphilis (VDRL) was negative both for the mother and the patient. Histopathological exam revealed a dermal infiltrate with predominance of large, round histiocytic cells with dense eosinophilic cytoplasm, with ground glass appearance, and eccentric reniform nuclei (Figure 3A). Immunohistochemical staining was positive for S100 and CD1a (Figures 3B and 3C). Routine laboratory workup and radiographs of chest, skull, pelvis and long bones were within normal ranges. The patient remains asymptomatic with no signs of recurrence.

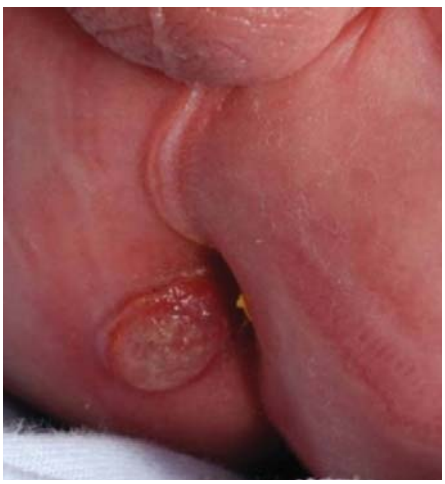

FIGURE 1: Erythematous, infiltrated, perianal plaque that rapidly progressed to ulceration

DISCUSSION

Langerhans cell histiocytosis (LCH) is a rare and clinically heterogeneous condition with monoclonal proliferation of this type of histiocyte.[1-3] Four clinical subtypes, which share significant clinical overlap, are known: Letterer-Siwe disease, Hand-Schüller-Christian disease, eosinophilic granuloma and congenital self-healing reticulohistiocytosis (CSHRH) or Hashimoto-Pritzker disease.[1,4,5] Histopathological and immunohistochemical studies are essential for diagnosis showing a dermal infiltrate with predominance of large, round histiocytic cells with dense eosinophilic cytoplasm with eccentric, reniform nuclei and Langerhans cells stain positive for S100 and CD1a (Figure 3A).[1-8]

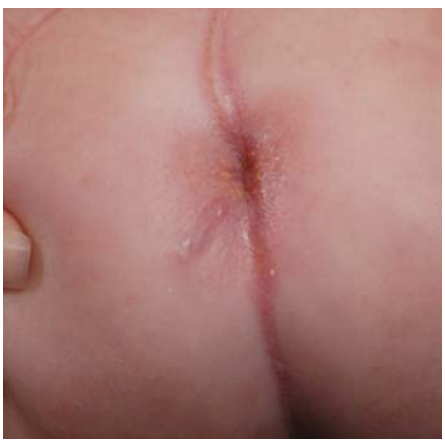

FIGURE 2: A second evaluation at two and a half months of age showed a complete and spontaneous resolution of the lesion

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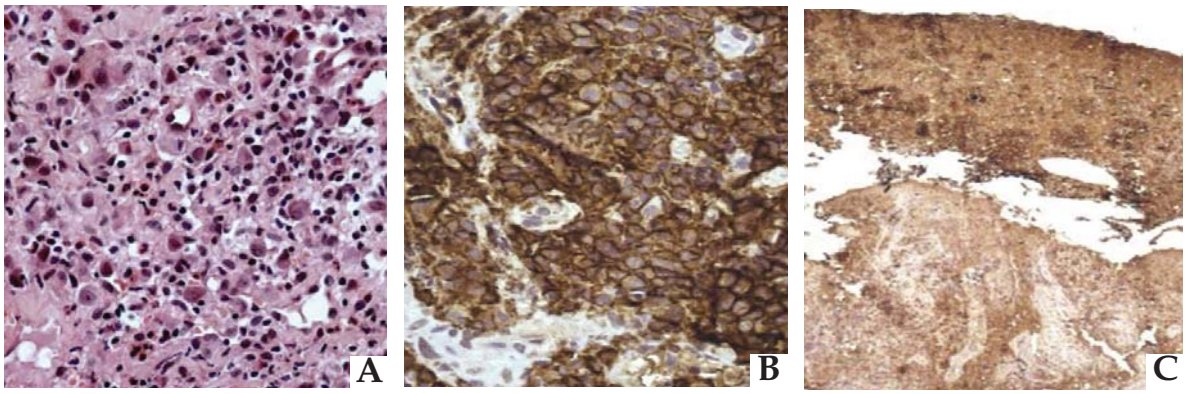

Figure 3: A Dermal infiltrate with prredominance of large, round histiocytic cells with dense eosinophilic cytoplasm, with ground glass appearance, and eccentric reniform nuclei. B and C Immunohistochemical staining was positive for S100 and CD1a

Birbeck granules on electron microscopy are specific for Langerhans cells.[1-9] Once the diagnosis is established, the extent of the disease must be carefully evaluated. Patients with systemic involvement may have a mortality rate as high as 20%.[9]

CSHRH carries a good prognosis.[1,4,7] Its true incidence may be underestimated since spontaneous resolution often occurs before assessment by a dermatologist.[1,6,8] CSHRH classic features include **1)** painless papules, nodules or plaques present at birth or during the first days of life; **2)** spontaneous regression in months; and **3)** proliferation of histiocytes with features of Langerhans cell.[8,10]

Most patients present with multiple lesions, but solitary lesions are seen in 25% of cases and spontaneous regression takes place in two to three months.[4-5] Development of lesions in adulthood, as well as pulmonary and ocular involvement, are extremely rare.[9] CSHRH may eventually show multisystem recurrence with considerable increase in morbidity and mortality.[2,3,9]

Routine laboratory workup should include full blood count and ESR, electrolytes, urea, liver function tests, C-reactive protein and radiographic study of chest, skull, pelvis and long bones.[3,9] Differential diagnosis comprises vesicular and pustular neonatal eruptions such as congenital candidiasis, herpes simplex, varicela, *Listeria monocytogenes* infection and neonatal hemangiomatosis.[7,8]

Since the differentiation between CSHRH and other forms of LCH cannot be made solely on clinical and histopathological grounds, patients must have a multidisciplinary follow-up since recurrence and multisystemic involvement are reported in 5-10% of all cases.[5,8] ❑

Abstract: Langerhans cell histiocytosis is a rare, clinically heterogeneous desease. Since there is considerable clinical overlap among the four described variants (Hand-Schüller-Christian, eosinophilic granuloma, Letterer-Siwe and Hashimoto-Pritzker), the concept of spectral disease applies to this entity. The Hashimoto-Pritzker variant was first described in 1973. Characteristically, it is present at birth or during the first days of life, impairment is limited to the skin and prognosis is favorable with spontaneous resolution. We report a newborn male patient with Hashimoto-Pritzker disease presenting as a S100 + and CD1a + single congenital perianal lesion with rapid involution in two months.
Keywords: Histiocytosis, Langerhans-cell; Immunohistochemistry; Prognosis

Resumo: A histiocitose de células de Langerhans é doença rara, clinicamente heterogênea. Como há considerável sobreposição clínica entre as quatro variantes descritas (Hand-Schüller-Christian, granuloma eosinofílico, Letterer-Siwe e Hashimoto-Pritzker), o conceito de doença espectral aplica-se a esta entidade. A variante de Hashimoto-Pritzker foi descrita em 1973. Classicamente, está presente ao nascimento ou nos primeiros dias de vida, acomete exclusivamente a pele e o prognóstico é favorável, com regressão espontânea. Relatamos caso de paciente recém-nascido, masculino, com doença de Hashimoto-Pritzker, que se apresenta com positividade para S100 e CD1a, observando-se lesão congênita única perianal com involução rápida em dois meses.
Palavras-chave: Histiocitose de células de Langerhans; Imunoistoquímica; Prognóstico

REFERENCES

1. Jensen ML, Bygum A, Clemmensen O, Fenger-Gron J. Congenital self-healing reticulohistiocytosis - an important diagnostic. Acta Paediatr. 2011;100:784-6.
2. Querings K, Starz H, Balda BR. Clinical spectrum of cutaneous Langerhans' cell histiocytosis mimicking various diseases. Acta Derm Venereol. 2006;86:39-43.
3. Ricart J, Jimenez A, Marquina A, Villanueva A. Congenital self-healing reticulohistiocytosis: report of a case and review of the literature. Acta Paediatr. 2004;93:426-9.
4. Huang CY, Chao SC, Ho SF, Lee JY. Congenital self-healing reticulohistiocytosis mimicking diffuse neonatal hemangiomatosis. Dermatology. 2004;208:138-41.
5. Parentin F, Ventura G, Pastore S, Kiren V, Bibalo C, Pensiero S, *et al*. A singular case of congenital self-healing histiocytosis with skin, liver and atypical eye involvement. Ocul Immunol Inflamm. 2011;19:337-9.
6. Kapur P, Erickson C, Rakheja D, Carder KR, Hoang MP. Congenital self-healing reticulohistiocytosis (Hashimoto-Pritzker disease): Ten-year experience at Dallas Children's Medical Center. J Am Acad Dermatol. 2007;56:290-4.
7. Ersoy-Evans S, Gursoy T, Yigit S, Akcoren Z, Sahin S. Solitary congenital self-healing Reticulohistiocytosis in monozygotic twins. Pediatr Dermatol. 2006;23:273-5.
8. Orle J, Mósca AM, Sousa MA, Lima CM, Adriano AR, Rezende PM. Congenital self healing reticulohistiocytosis in a newborn (Hashimoto Pritzker). An Bras Dermatol. 2011;86:785-8.
9. Lau L, Krafchik B, Trebo MM, Weitzman S. Cutaneous Langerhans cell histiocytosis in children under one year. Pediatr Blood Cancer. 2006;46:66-71.
10. Nakahigashi K, Ohta M, Sakai R, Sugimoto Y, Ikoma Y, Horiguchi Y. Late-onset self-healing reticulohistiocytosis: Pediatric case of Hashimoto-Pritzker type Langerhans cell histiocytosis. J Dermatol. 2007;34:205-9.

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