Case 18

A newborn Thai boy from Bangkok

Chief complaint: multiple skin lesions presented at birth

Present illness: A 2690-g male infant was born by natural spontaneous delivery at 37 weeks of gestation to a healthy woman.

Apgar scores were 9,10 at 1 and 5 minutes consecutively. At birth, the infant had about 10 skin lesions distributed over scalp, trunk and extremities. The baby was otherwise well.

Physical examination

Skin: multiple discrete red-brownish papules with hemorrhagic crust, about 0.5cm in diameter at posterior aspect of scalp, back, left arm, right arm, right hand, left leg, one pustule at dorsum of right foot, 2 pustules at plantar surface of left foot, and one ulcer at left groin, no mucous membrane involvement

Abdomen: no hepatosplenomegaly

Investigation

Tzank smear from vesicle: no multinucleated giant cell

Gram stain from pustule : no organism

Serum PCR for Herpes simplex type I,II : non reactive

CBC: WBC 17400 (N65% L23% Mo8% Eo2% Ba2%) Hct 55.9%
Hb 20.2 Plt 228000

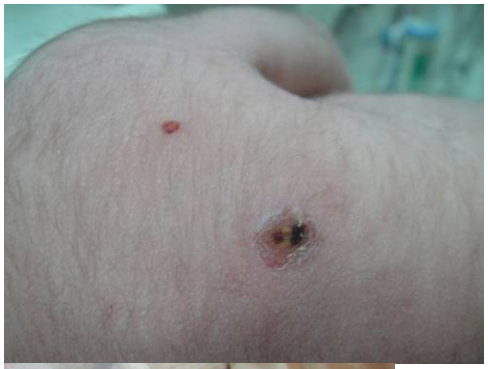
Histopathology (S10-10395) :

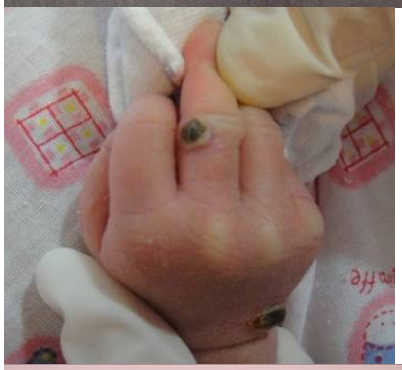
Dense diffuse proliferation of Langerhan's cells characterized by eccentrically located, reniform vesicular nuclei and abundant eosinophlic cytrophasm, in the upper dermis with some infiltrate in the epidermis (epidermotropism) covered with scale-crust, erosion and intermingled with inflammatory- cell infiltrate lymphocytes neutrophhlis and eosinophils

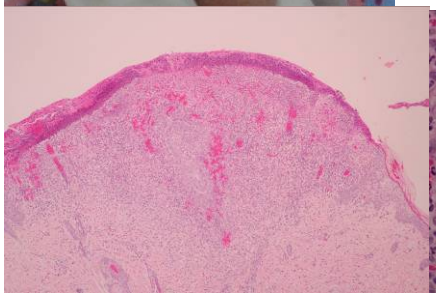
Immunohistochemical study (S10-10664)

S100 : positive

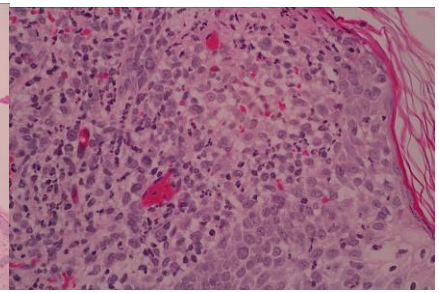
CD1a : positive



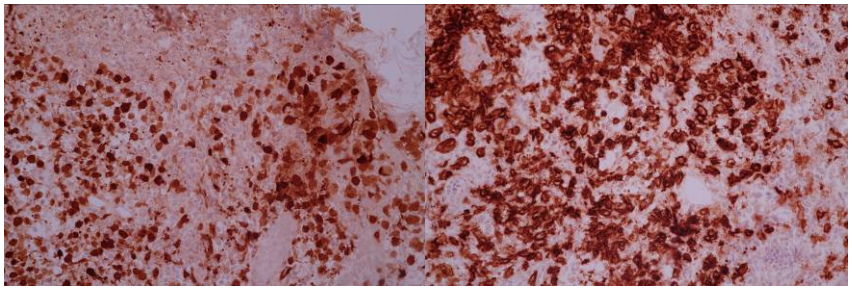




CD1a



S100



Diagnosis: Congenital self healing reticulohistiocytosis
Treatment: no.

At one month follow up, all lesions had disappeared leaving scars. No new lesion.

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Discussion:

Congenital self healing reticulohistiocytosis (CSHR) or Hashimoto-Pritzker is an autoinvoluting, self limited form of LCH. CSHR is usually present at birth or during the neonatal period. It has been described in two forms: a solitary[1] and a multinodular variant. Solitary or generalized lesions can affect any part of the cutaneous surface. Lesions range form 0.2-2.5 cm in diameter. Lesions may grow postnatally. Exceptionally large tumors up to 8 cm in diameter can occur. At presentation the lesions can be papules or nodules with or without erosion[2] or ulceration. Individual lesions are red, brown, pink, or dusky. Lesions may rarely appear as hemorrhagic bullae[3]. Lesions greater than 1 cm characteristically ulcerate as they regress. Lesions are asymptomatic and spontaneously heal over 8 to 24 weeks, leaving atrophic scarring from the ulcerated nodules. Internal involvement is not found.

On histologic examination large mononuclear cells and multinucleated giant cells with ground-glass or foamy cytoplasm are present in the dermis and epidermis. Immunoperoxidase staining is positive for CD1, HLA-DR, and S-100. By electron microscopy 10% to 25% of cells have Birbeck granules. This histology is characteristic but cannot distinguish this entity from other forms of LCH, so a definitive diagnosis cannot be made histologically.

This is a proliferation of Langerhans cells. Because LCH with systemic involvement may present in identical fasion, systemic evaluation is recommended, including a physical examination, complete blood count, liver function test, and bone survey. A liver-spleen scan and bone marrow biopsy should be considered[4-6]. Long- term follow-up must be carried out to detect evidence of relapse or progression of the disease[5-7].

References

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