

Systemic Congenital Langerhans Cell Histiocytosis Masquerading as Diffuse Neonatal Hemangiomas

A 3-month-old girl was brought to us with complaints of fever and breathlessness for two days. On examination, child was afebrile, had severe tachypnea, hypoxia and firm hepatosplenomegaly. There were diffuse cutaneous lesions – hemorrhagic papules and vesicles on the scalp, face, trunk and extremities, including palms and soles (*Fig. 1*). The mother reported the lesions to be present at birth but were progressively increasing in number. The child had been evaluated for the lesions in the newborn period and a presumptive diagnosis of Diffuse neonatal hemangiomas was made due to the vascularized appearance of the cutaneous lesions; a biopsy was not attempted. As the child was otherwise asymptomatic, she was advised to follow-up.

On laboratory evaluation, she had anemia and hypoproteinemia. Computed tomography of chest revealed diffuse ground glass appearance suggestive of Interstitial lung disease. Skin biopsy demonstrated focal collection of cells with grooved nuclei. Immunohistochemistry revealed S100+ and CD1a+ cells. The final diagnosis was congenital Langerhans Cell Histiocytosis (LCH), possibly with progression from single system (skin) to multi-system (liver, spleen, lung) involvement. The child was treated as per 2009 Histiocyte Society guidelines for LCH protocol. On follow-up, the cutaneous lesions completely regressed and child could be weaned-off oxygen.

Neonatal/congenital LCH is defined when it presents within the first 4 weeks of life (irrespective of age at diagnosis). The largest cohort study of neonatal LCH reported that 61 out of 1,069 LCH patients (6%) met the criteria for neonatal LCH [1]. Skin lesions are the most common initial manifestation, irrespective of disease extent at diagnosis.

Differential diagnosis of hemorrhagic skin lesions presenting in the neonatal period include cytomegalovirus, candidiasis, varicella, herpes simplex, neonatal toxic erythema, infantile acropustulosis, pigmentary incontinence, eosinophilic pustular folliculitis, neonatal erythroptosis, disseminated neonatal



FIG. 1 Diffuse hemorrhagic papules over trunk, palms and soles.

hemangiomas and congenital leukemia cutis. The presence of firm hepatosplenomegaly and ILD (due to deposition of histiocytes) should raise the index of suspicion towards LCH. Disseminated neonatal hemangiomas can be a close mimic as they can present with visceral involvement, and can be best differentiated by skin biopsy [2].

Isolated cutaneous LCH has a high tendency for spontaneous regression, and has been described as Congenital self-healing reticulohistiocytosis. However, approximately 60% can progress to multisystem involvement, necessitating close follow-up.

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