

CONGENITAL SELF HEALING CUTANEOUS LANGERHANS CELL HISTIOCYTOSIS WITH MULTIPLE HYPOPIGMENTED MACULES AFTER RECOVERY IN A NEONATE

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Abstract- Langerhans cell histiocytosis (LCH) is the most common type of childhood histiocytic disorder with an incidence of 0.2 to 1 per 100,000 children under the age of 15 years (1). The clinical picture of LCH varies from single system (S-S) bone or skin disease to multi-system disease (M-S) (2). We report a neonate with disseminated papulonodular eruption containing mononuclear CD1a and S100 positive histiocytic cells infiltration at epidermis and underlying dermis. The diagnosis of a congenital self healing Langerhans cell histiocytosis (CSHLCH) was made and follow up showed a complete recovery of the eruptions, leaving hypopigmented macules in the sites corresponding to the initial findings.

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INTRODUCTION

The childhood histiocytosis is a rare proliferative disorder of cells of the monocyte-macrophage system of bone marrow origin. Childhood histiocytosis constitute a divers group of disorders. Congenital cutaneous langerhans cell histiocytosis was report for the first time in 1973 and subsequently named Hashimoto Pritzker disease after the authors of the first description (3). Reports of Congenital cutaneous LCH describe high

tendency to spontaneous regression so Hashimoto Pritzker disease called congenital self healing histiocytosis (4). However there is growing evidence that progression to multi-system disease with fatal outcome is possible (5). The histopathologic findings consist of dense nodular infiltrate of mononucleated and polynucleated cells in the dermis. The cell stain positively for CD1a and S100 (2-10).

We report a neonate with findings of Congenital cutaneous langerhans cell histiocytosis.

CASE REPORT

A new born girl with body weight of 2900gm was born as the third child of healthy parents. The pregnancy and delivery were uncomplicated. Physical examination at birth was normal.

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Fig. 1. (A) At age 4 months with multiple red papulonodular lesions with hypopigmented macules in the lower limbs corresponding to the initial lesions . (B) At age 9 months

Mother and her newborn baby were discharged one day after delivery without any complication. On the fifth day of birth , red papulonodular lesions begun to appear from her legs. The lesions disseminated gradually on scalp, face and trunk. The patient had not any symptoms except a little irritability, probably because of pruritus.

The diagnosis was not known until the age of 4 months, when her parents brought her to our hospital (Central Pediatric Clinic of Tehran-Iran).

We examined the patient , she had red papulonodular lesions measuring 0.5 cm in diameter , almost crusted located on the scalp, face, trunk and upper limbs (Fig.1A). In the lower limbs, lesions resolved and leaving hypopigmented macules corresponding to the initial lesions. There was no hepatosplenomegaly or lymphadenopathy and the patient's general condition was good. Complete blood count was within normal limits, electrolytes,

urea, creatinine and liver function test were also normal. Bacterial cultures of blood and urine were negative. TORCH study was negative. The chest, skull x-ray and abdominal ultrasound all were normal. Histopathologic examination of the skin lesion on her back revealed infiltration of mononuclear cells in epidermis and also upper dermis composed of medium size histiocytes with reniform nuclei, eosinophilic cytoplasm and some degree of atypia admixed with eosinophils and rare giant cells. Immunohistochemical study show positive reaction for CD1a and S100 (Fig. 2A,B). We made a diagnosis of CSHLCH.

The lesions gradually resolved within the following 5 months and at 9 months of age hypopigmented macules were seen in the sites corresponding to the initial findings (Fig.1B). In general examination, the infant was normal and laboratory tests were in normal range too.

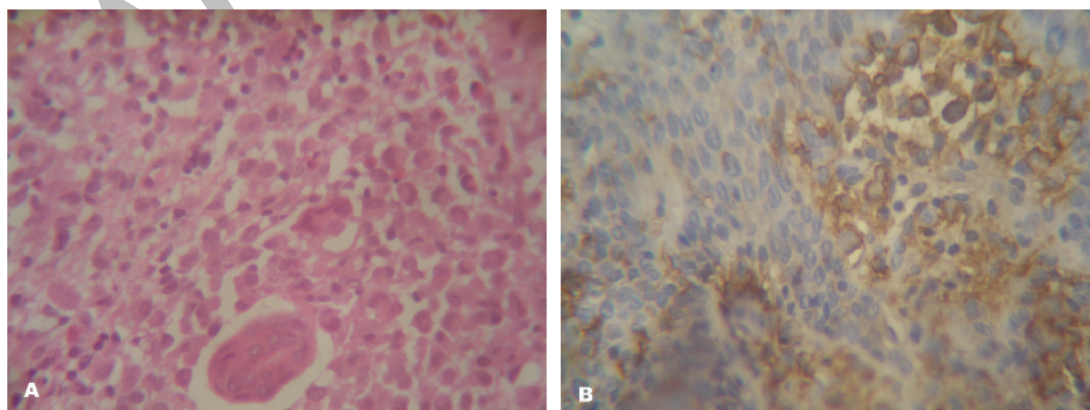


Fig. 2. (A)H&E stained skin biopsy showing histiocytic infiltration at upper dermis and epidermis. (B)CD1a stained skin biopsy (immuno histochemistery) showing positive cell membrane staining.

DISCUSSION

Hashimoto and Pritzker first describe CSHLCH in 1973. The disease was initially described as an independent entity, and later it was classified as part of the LCH (1-3).

Skin rash is the predominant first manifestation of both (S-S)LCH and (M-S)LCH in neonates (9-10). Due to the possibility of clinically unapparent involvement of other organs, through clinical, laboratory and imaging evaluation is mandatory for the categorization of a case as (S-S)LCH or (M-S)LCH.

The disease course is unpredictable upon diagnosis and ranges from spontaneous healing to chronic course of fulminant deterioration. The "Wait and See" approach could be justify in patient with isolated cutaneous LCH.

We report this case because this disease is very rare and this case spontaneously healing without any treatment.

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