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Case Report

Rare diagnosis in a neonate with isolated skin lesions

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ABSTRACT

Langerhan Cell Histiocytosis (LCH) represents a group of diseases with varied spectrum of presentation. LCH limited to skin is not very common with 100 cases reported in literature. A definitive diagnosis is important for both management and follow up. A 30 days old male infant was brought with vesiculopustular skin lesions over the trunk and back since day 12 of life. Systemic examination was normal. Preliminary investigations revealed a normal haemogram with normal liver and renal function tests. He was initially managed symptomatically. In view of persisting skin leisons, biopsy done was suggestive of Langerhans Cell Histiocytosis and confirmed by CD1a postivity on immunohistochemistry. Skeletal survey did not reveal any lytic leison, bone scan was normal. There was no biochemical evidence of diabetes insipidus. The child was diagnosed to have neonatal isolated cutaneous LCH. He was followed up periodically without any therapy. Presently, he has an Event Free Survival (EFS) of 2 years without any progression. Though isolated cutaneous Langerhan Cell Histiocytosis is rare in neonates, it should still be considered in the differential diagnosis by the treating paediatrician. Though it can be a self-limiting entity, it is very important to recognize this condition, since it is mandatory to look for multisystem involvement by LCH as it determines the treatment intensity. It is essential to closely follow up these children to know if this heralds involvement of risk organs by LCH or a self-limiting disorder. This case also highlights the multidisciplinary dialogue required in the diagnosis and management of such rare cases.

Keywords: Langerhan cell histiocytosis, Neonates

INTRODUCTION

A 30 days old male infant, first born to non-consanguineous parents with an uneventful antenatal and postnatal period was brought with vesicopustular skin lesions predominantly on the trunk and gluteal region noted since day 12 of life.

Child was born at term with birth weight of 2 kg and 3.3kg on day 30 of life. No significant family history was present. On physical examination, his vitals were normal. He was alert and healthy. His skin had vesicopustular lesions involving the hands; multiple hypo pigmented scaly macules with central puckering over the front and back of trunk (Figure 1, and Figure 2). There was no seborrhea or petechial lesions. His respiratory, cardiac, abdominal and neurological examinations were all within normal limits.

Blood investigations done revealed white blood cell count of $8.6x10^9$ per liter, Hb 8.7g/L, platelets 640 x 10^9 /L. Liver enzymes and renal function tests were normal. His chest radiograph was normal.

Hospital course

In view of persisting skin lesions, pediatric dermatologist was consulted and skin biopsy was done. Skin biopsy from one of the lesion revealed hyperplastic epidermis and upper dermis with dense infiltration by many eosinophils, polymorphs, mononuclear inflammatory cells with reniform nuclei as shown in Figure 3.

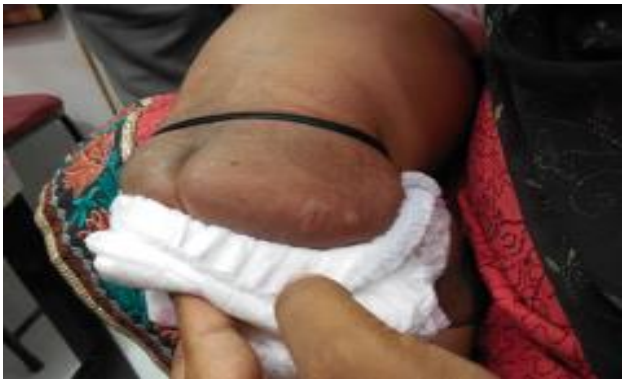

Figure 1: Vesicopustular lesions on the buttocks.

Figure 2: Vesicopustular lesions on the hand.

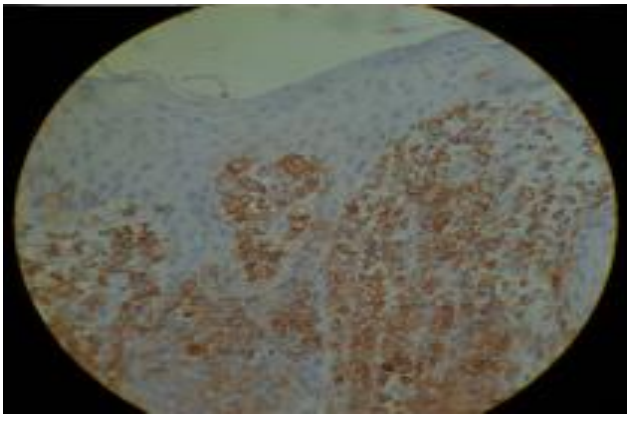

Figure 3: CD1a skin biopsy (immunohistochemistry) showing positive cell membrane staining.

Immunohistochemistry showed CD 1a: positive, S-100 positive, CD 68: focal positive, features consistent with the diagnosis of Langerhans Cell Histiocytosis (LCH) as shown in Figure 4. Further work up was done to rule out systemic involvement of LCH. His serum osmolality was 282 mosm/kg of water; urine osmolality 180mosm/kg water. His skeletal survey was normal. Bone scan revealed no abnormal tracer uptake. In the absence of systemic involvement, the child was managed conservatively. Child was kept under close follow up and skin lesions showed complete resolution by 3 months of age. The child has been on regular follow up since then and is now currently 3 years of age.

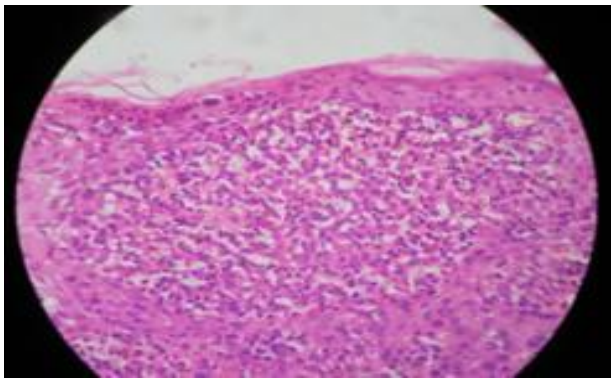

Figure 4: H and E stained skin biopsy showing histiocytic infiltration at upper dermis and epidermis.

Final diagnosis

Neonatal skin only Langerhans cell histiocytosis.

DISCUSSION

The differential diagnosis of vesicopustular lesions range from transient, benign pustular eruptions to serious and life-threatening conditions. The infectious etiology could be mild as candidiasis-neonatal, impetigo neonatorum and scabies to serious infections like Chlamydia, Escherichia coli, Hemophilus influenza, Klebsiella pneumonia, Listeria monocytogenes, Pseudomonas aeruginosa, Staphylococcus aureus, Group A beta hemolytic Streptococcus, Staphylococcal Scalded Skin Syndrome (SSS).[1]

A thorough workup to rule out TORCH infections, listeriosis, and congenital candidiasis are to be done with quick tests like Direct Fluorescent Antibody (DFA) with or without Tzanck smear, KOH and Gram stain. Second line investigations like skin biopsy, RPR, VDRL and cultures are next. Empiric therapy with acyclovir is usually initiated and if ill appearing broad spectrum antibiotics; if syphilis is suspected, penicillin is used.[1] The noninfectious causes can be benign like Acropustulosis of infancy, Eosinophillic pustular folliculitis, Erythema toxicum, Miliaria, Transient neonatal pustular melanosis to potentially serious conditions like Acrodermatitis enteropathica, Epidermolysis bullosa, Epidermolytic hyperkeratosis, Incontinentia pigmenti, Urticaria pigmentosa, Neonatal Herpes gestationis, and Pemphigus vulgaris.[2] Incidence

of LCH in infants is about 25 per 1 million infants, and <5% of these cases are found to be neonates.[3] Vesicopustules are the most common morphology in congenital or neonatal LCH. Solitary lesions, necrosis and involvement of extremities were seen more commonly in self-regressive LCH, whereas intertriginous involvement was significantly more frequent with non-self-regressive LCH.[4] A retrospective cohort study of nineteen neonates with LCH showed that the skin lesion morphologic traits did not correlate with extent of extra-cutaneous disease and patients with disease limited to skin and mucus membranes had excellent outcome.[4]

This finding is contrary to earlier reported literature of nodular, whether congenital or later, to generally have had a better prognosis.[5,6] Mortality rates as high as 50% have been recorded in a study of neonates with disseminated LCH.[7] 'Hashimoto-Pritzker disease' first described in 1973 refers to the skin only LCH congenital or neonatal usually self-resolving in a few months.[8] Because of the potential for recurrence in skin or systemically, it has been suggested that the diagnosis of congenital self-healing LCH be made retrospectively, after a patient has remained free from systemic involvement for several years. There are no absolute criteria that can reliably distinguish self-regressive cutaneous LCH and non-self-regressive LCH in the neonatal and early infancy period.[9]

In a study of 9 patients with skin- only LCH who were younger than 28 days, 5 had spontaneous regression, whereas 4 progressed to have multisystem disease.[10] In another study of 12 patients with skin only LCH who were younger than 1 year, 4 progressed to have multisystem disease within 1 to 6 months.[11] Another separate study revealed that 21 of 31 patients had a self-regressive course.[12] A thorough investigation is required to rule out multisystem involvement. In our case, child presented with only skin lesions and no other systemic involvement and is being followed up periodically without any therapy. It is essential to closely follow up these children to know if this heralds involvement of risk organs by LCH or a self-limiting disorder.

CONCLUSION

Isolated cutaneous Langerhans Cell Histiocytosis is rare in neonates, but should still be considered in the differential diagnosis by the treating Pediatrician. Though it can be a self-limiting entity, it is very important to recognize this condition, since it is mandatory to look for multisystem involvement by LCH as it determines the treatment intensity. This case also highlights the multidisciplinary dialogue required in the diagnosis and management of such rare cases.

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