

## Case Report

# Langerhans cell histiocytosis in an infant: a case report from North-East India

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## ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare disorder of reticuloendothelial system characterized by uncontrolled proliferation and accumulation of CD1a and CD207 dendritic cells (DCs). 3-4 cases per million occur annually in children under 15 years of age, with a male:female ratio of 2:1 and peak incidence in infants aged 1 to 2 years. Report a case of LCH in a month-old female infant with skin lesions for 2-3 weeks. Skin biopsy was suggestive of LCH and it was confirmed with immunohistochemistry markers that were positive for S100, CD1A and negative for CD68.

**Keywords:** Children, Cutaneous manifestations, LCH

## INTRODUCTION

Langerhans cell histiocytosis (LCH) is the name proposed by Risdall et al in 1983, for all group of disorders previously called histiocytosis X.<sup>1</sup> LCH is a rare clonal disease of the monocyte-macrophage system characterized by uncontrolled proliferation and accumulation of CD1a+/CD207+ dendritic cells (DCs) due to continuous immune stimulation,<sup>2</sup> and accumulation of these pathological Langerhans cells causes infiltration and destruction of the local tissues.<sup>3</sup> Clinical feature is highly variable and may range from isolated, self-healing skin and bone lesions to life-threatening multi-system disease.<sup>2</sup> The cutaneous lesions may be the sole manifestation of LCH where typical manifestation is a seborrheic dermatitis-like lesion on the scalp and the flexural regions.<sup>4</sup>

## CASE STUDY

A month-old female infant presented with lesions on face, arms, abdomen, trunk, palms and soles for 2-3 weeks, which was first noticed on forehead as whitish papule. The baby was delivered at full term by elective

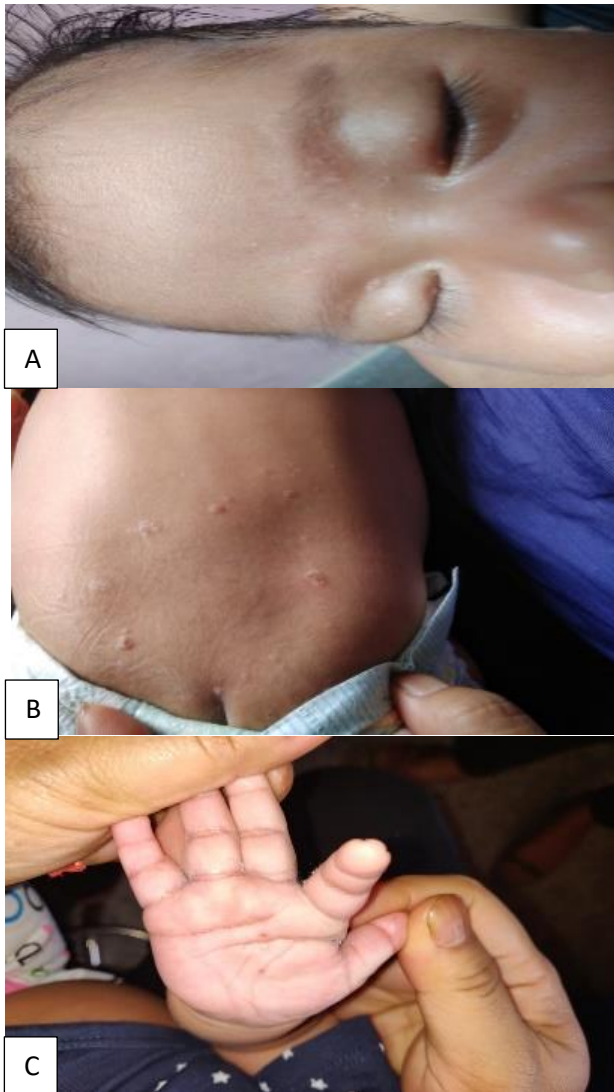
caesarean section and postnatal period was uneventful. There was no history of fever or any other systemic illness preceding the cutaneous lesions. Family history was not significant.

On examination multiple hyperkeratotic and erythematous papules with few pustules was noticed on face, bilateral arms, abdomen, trunk, buttocks, palms and soles (Figure 1 A-C). Oral and genital mucosa were spared, general physical examination was unremarkable.

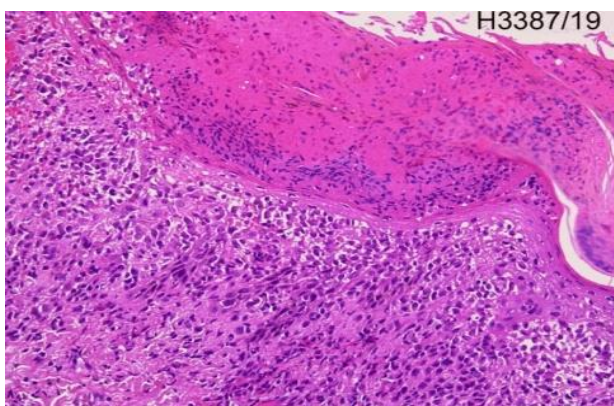
Histopathological examination of a lesion on abdominal wall revealed histiocytes with eosinophilic cytoplasm arranged in sheets, occasional giant cell with scattered eosinophils and sub corneal neutrophilic plasma exudate (Figure 2).

Immunohistochemistry markers were positive for S100 (Figure 3), CD1A and negative for CD68, confirming the diagnosis of LCH. Routine blood tests, urine R/E, chest X-ray and USG abdomen revealed no abnormalities.

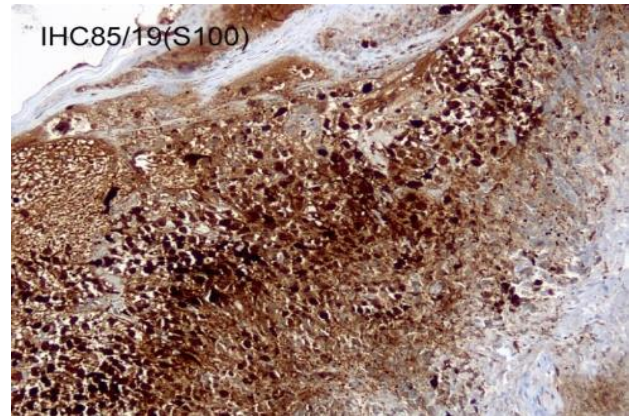
The patient was started on topical tretinoin with plan for oral isotretinoin but was lost to follow up.



**Figure 1: Multiple hyperkeratotic and erythematous papules with few pustules on (A) face, (B) lower back and (C) palm.**



**Figure 2: Histiocytes with eosinophilic cytoplasm arranged in sheets, occasional giant cell with scattered eosinophils and subcorneal neutrophilic plasma exudate (H and E 40x).**



**Figure 3: Positive S100 immunohistochemistry marker staining of LCH cells.**

## DISCUSSION

LCH is a rare disease that generally affects children but can affect all age groups. Annually 3-4 cases per million occur in children under 15 years of age, with peak incidence in infants aged 1 to 2 years.<sup>4</sup>

LCH has a broad clinical spectrum that ranges from isolated cutaneous, pulmonary or bone localization to acute widespread cutaneous and visceral disease.<sup>5</sup>

In this case presented with cutaneous lesions without evidence of any systemic involvement which was evaluated with all routine blood tests, USG abdomen and X-ray chest.

The identification of characteristic cells and the presence of immunohistochemistry markers, such as CD1a and or CD207 (langerin), confirms the diagnosis of LCH.<sup>2</sup>

Treatment of LCH depends on the site of the lesion, its extent, and the number of lesions present.<sup>3</sup> In patients with LCH limited to the skin, lesions may spontaneously resolve, otherwise a variety of methods may be used, including topical steroids, oral methotrexate or thalidomide, topical nitrogen mustard or psoralens with UV light.<sup>2</sup>

Patients may experience long-term permanent consequences that include endocrine disorders (DI and growth hormone deficiency), orthopaedic problems, decreases or loss in hearing and sight, the loss of teeth, neurological defects and impaired lung and liver function. Patients with LCH have a higher risk for developing secondary cancers (leukaemia and non-Hodgkin lymphoma).<sup>2</sup>

Recurrence rates around 1.6 to 25% is reported, therefore a close and regular follow-up for a long period is advised.<sup>3</sup> A wide range of clinical spectrum from an asymptomatic form to those with multi-system

presentation, diverse prognosis and the risk of long-term complications cause LCH to remain a major challenge.<sup>2</sup>

## CONCLUSION

A proper history, histological examination and immunohistochemistry markers are necessary for a definitive diagnosis as LCH can present with wide range of clinical spectrum. Risk of long-term complications and high recurrence rate demands long term regular follow up.

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