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Langerhans Cell Histiocytosis with Nail Involvement: A Rare Case

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Abstract: ***Langerhans cell histiocytosisis a proliferative disease characterized by excess accumulation of CD1a + Langerhans cells in various sites, leading to tissue damage. More common in males (m:f = 2:1) with estimatedprevalence of 2–5 children per million per year. Common sites involvedare skin, ears, periodontal, bone, lymph nodes, endocrine glands and lungs. Herein, we report a case of 3-year-old boy presenting with a hypopigmented maculopapular lesion in the seborrheic area and lymphadenopathy. Nail dystrophy was also present. Histopathological and immunohistochemical studies were suggestive of LCH. Langerhans cells histiocytosis is a rare disease with a high rate of misdiagnosis and definitive diagnosis depends on pathological biopsy and X-ray examination. The prognosis is related to the age of onset and the number of affected organs. Here we report a rare case of LCH with Nail dystrophy.***

Keywords: Langerhans cell histiocytosis(LCH), Langerhans cell, Nail, Histopathology, Immunohistochemistry

1. Introduction

Langerhans cell Histiocytosis is a rare tumor in which Langerhans cells derived from bone marrow, infiltrate in various organ and cause tissue damage. It can affect any organ systems, but primarily involve bone, skin lymph node, liver, spleen, and endocrine glands[1].LCH was previously known as histiocytosis X and was classified into three types: eosinophilic granulomatosis, Hand-Schüller Christian disease, and Letterer-Siwe disease. These are all characterized by the proliferation of Langerhans cells and have been collectively referred to as LCH.[2] At present, LCH is classified into three groups: single-system single-site (SS type), single-system multi-site (SM type), and multi-system multi-site (MM type), and treatment differs depending on the location and type of disease.[3]Nail involvement with varying morphology of changes has been reported, though rarely, in Langerhans cell histiocytosis. Nail involvement has also been considered an unfavorable prognostic sign and seems to be more frequent in multisystem Langerhans cell histiocytosis[4,5,6]. The medical literature currently reports only 38 cases of Langerhans cell histiocytosis with associated nail involvement[7]. Here we report a case 3 year old boy with LCH with prominent nail dystrophy.

2. Case Report

History- A 3 years old boy presented with complains of multiple light colored raised lesions on forehead, scalp, interscapular area of back and on chest for 6 months and a painful swelling on right side of neck.

On examination- Multiple hypo- pigmented maculopapular lesion with fine scaling seenon forehead, chest, retro auricular area, interscapular area, scalp. Right cervically mphadenopathy was seen. Nails of ring and little finger of left hand, and those of thumb, middle and little finger of right hand were brittle, brownish in colour and distorted.

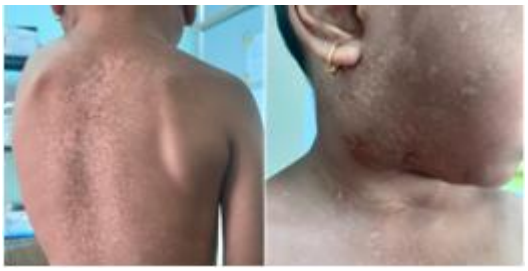

Figure 1: Maculopapular lesion on back and face

Figure 2: Dystrophic nails of hand.

X ray skull showed punched out lytic lesions with slightly sclerotic margins and irregular outline in the parietal area of skull.

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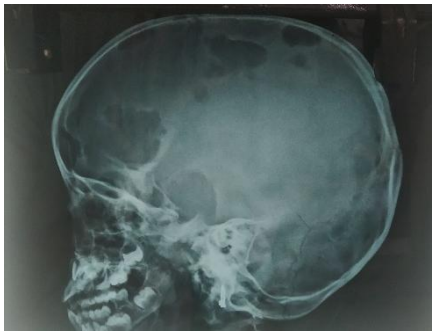

Figure 4: X ray skull showing lytic lesion.

On histopathology right cervical lymph node showed fibro-collagenous tissue along with inflammatory infiltrate consisting of Langerhans cells, dense eosinophils, neutrophils, foam cells, plasma cells and multinucleated giant cells.

On immunochemistry atypical histiocytes were positive for cd 1a and cd 207.

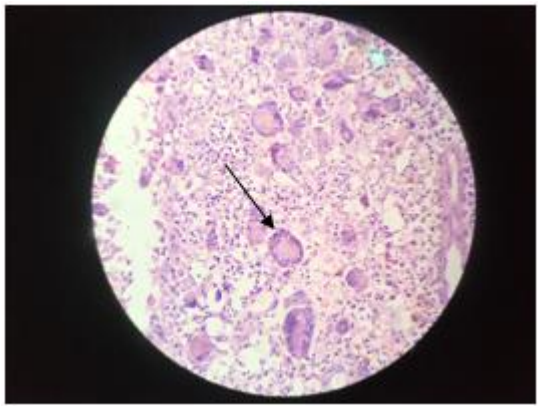

Figure 3: Lymph Node Biopsy showing Langerhans Cell

3. Discussion

Langerhanscell histiocytosis is a rare disorder having varied presentation, with skin lesion mostly mimicking seborrheicdermatitis. Nail involvement has been documented in LCH but is very rare and is reported only in children. In 1985 in a study of 32 children with LCH in the United States, no nail unit involvement was reported [6]. Another study in 1988 evaluated 124 pediatric patients over a period of 14 years and no nail changes were recorded. However, there are few individual reports and small case series with nail involvement in LCH.In a review study by Bonometti et al in 2021 of cases with LCH with nail involvement they reported the following Nail changes in LCH onycholysis (73.7%), subungual hyperkeratosis (60.5%), purpuric striae (55.3%), nail dystrophy (55.3%), paronychia (28.9%), pachyonychia (21.0%) and longitudinal grooving (21.0%). In most cases, more than one nail presented with changes and 55.3% of cases, all fingernails were involved[7]. Bonometti et al highlighted an association between nail involvement in Langerhans cell histiocytosis and multisystem presentation with lung involvement in pediatric patients. Our Patient, a 3-year male child showed skin manifestation with Nail dystrophy.

4. Conclusions

We reported a case of LCH who presented with Nail dystrophy. It is suggested that LCH should be considered in differential diagnoses of children with isolated severe nail lesions. Further studies should be done to determine the association of Nail changes with multisystem involvement.

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