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Langerhans Cell Histiocytosis - A Rare Case Diagnosed Cytologically.

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ABSTRACT

LCH is a rare idiopathic disorder characterized by pathogenic clonal proliferation of bone marrow derived langerhans cells which are immature dendritic cells characterized by presence of Birbeck granules. The annual incidence of LCH is nearly 5.4cases per one million people with male predominance. LCH is primarily a pediatric disease with peak age ranging between 1-4 yrs commonly involving bone. It manifests as single system unifocal, multifocal or multisystem disease. The purpose of this case report is to describe a case of 10 month old male baby with unifocal bone involvement and discuss the clinical, radiological and characteristic cytological features that lead to the diagnosis of LCH.

Keywords: Langerhans cell histiocytosis (LCH), cytological features, unifocal bony lesion.

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a group of idiopathic disorders characterized by the presence of excess immune system cells similar to the bone marrow–derived Langerhans cells juxtaposed against a backdrop of inflammatory cells that is lymphocytes, macrophages, and eosinophils.

LCH was earlier referred to as Histiocytosis X, where histiocytosis denotes proliferation of histiocytes and other inflammatory cells, where as X denotes unknown etiology of the disease Later in 1973,[1] LCH term was adopted seeing Histiocytes involved in disease were phenotypically similar to Langerhans cells found in skin and mucosa Langerhans cells,[2] which help regulate the immune system, are normally found throughout the body, especially in the skin, lymph nodes, spleen, lungs, liver, and bone marrow In people with LCH,[3] these cells multiply excessively and build up in certain areas of the body, causing tumours called granulomas to form.[4]

The working group of the Histiocyte Society has divided histocytic disorders into three groups: Dendritic cell histiocytosis, Macrophage-related disorders, and Malignant histiocytosis.[5] LCH belongs to the group of dendritic cell histiocytosis.

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LHC includes four variants, with different degrees of severity:[6-8]

- Hashimoto-Pritzker disease, a congenital self-healing form
- Letterer-Siewe disease, a severe, acute and disseminate form
- Hand-Schüller-Christian disease, an intermediate chronic form with multiple lesions characterized by diabetes insipidus, bulging of the eye and localized lesions in the bone
- Eosinophilic granuloma, a less severe form, characterized by solitary or few, and chronic lesions of bone or other organs.

Because all the variants have many common symptoms it is thought that they may be manifestations of LCH and not separate syndromes.[6-8]

CASE REPORT

We report a case of 10-month old male baby who presented with a history of gradually progressive diffuse swelling over right cheek. [Figure 1] There was no relevant history of trauma. Medical history and family history were insignificant.

On examination a single diffuse soft to firm swelling noted over right maxilla. The child was investigated and Complete hemogram revealed Anemia (Hb-7.8) and leukocytosis(TLC-15300) and ALC- 8000. HRUS of cheek was performed which revealed a large inhomogeneous hypoechoic soft tissue mass lesion in right axilla causing expansion of maxillary

cortical outlines extending from inferior wall of orbit superiorly to alveolar margin inferiorly, giving an impression as soft tissue right maxillary tumor for further evaluation. CT PNS features suggested the possibility of LCH or Malignant Rhabdomyosarcoma with metastatic deposits [Figure 2]

USG guided FNAC was performed and blood mixed aspirate obtained. H&E stained Cytosmear studied showed sheets and clusters of round to polygonal cells admixed with lymphocytes, eosinophils and other polymorphs. [Figure 3] Individual cells had moderate to abundant eosinophilic cytoplasm with eccentrically placed hyperchromatic pleomorphic nucleus with prominent nucleoli and occasional nuclear grooving. [Figure 4] Also seen many bi/multinucleate giant cell in blood element background. [Figure 5] The above features thus suggested the diagnosis of LCH. Bone scan was done which was negative for any skeletal metastasis.

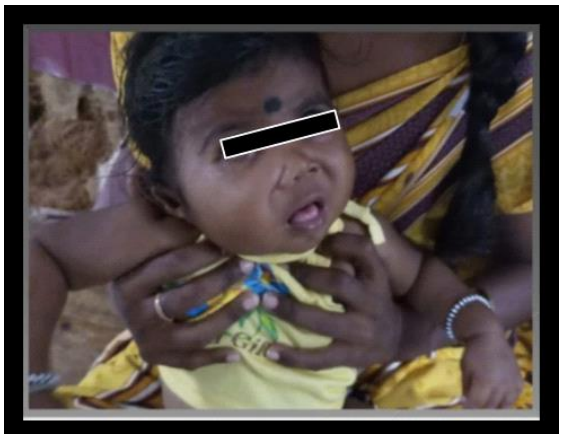

Figure 1: LCH clinical picture showing diffuse non erythematous,non tender swelling over the right cheek

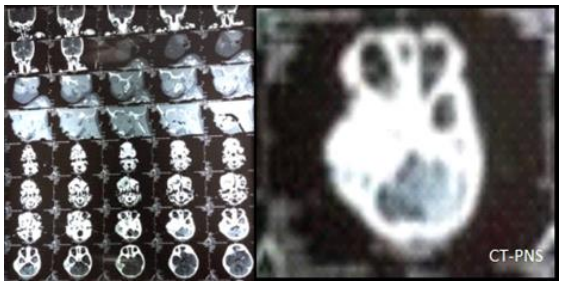

Figure 2: CT-PNS showing hyperdense lesion involving right maxilla with soft tissue component

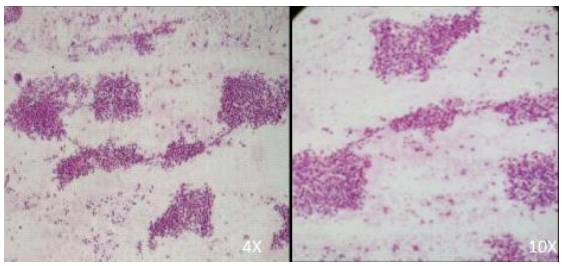

Figure 3a) & 3b): H and E stain showing sheets and clusters of cells.

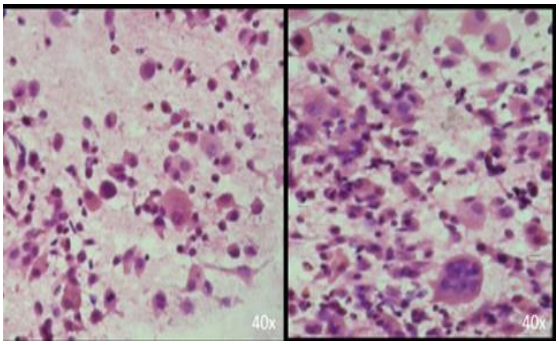

Figure 4: High power (40x) H and E stain showing round to polygonal cells with abundant eosinophilic cytoplasm and eccentrically placed nuclei with prominent nucleoli, bi/multinucleate giant cells, eosinophils and polymorphs.

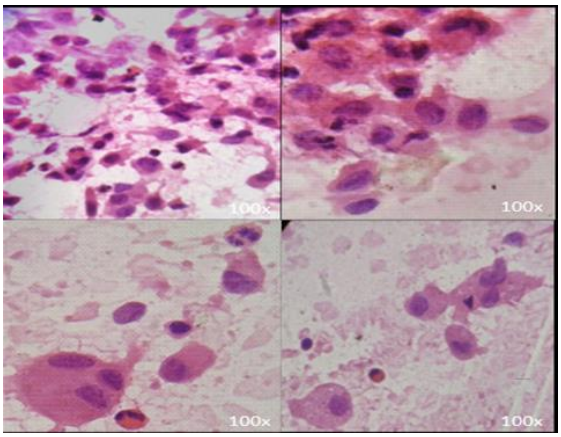

Figure 5: Oil immersion (100x) H and E stain showing histiocytes, eosinophils and polymorphs with few cells showing features of nuclear grooving.

On follow up the diagnosis of LCH was confirmed by Histopathology at his native place.

DISCUSSION

LCH formerly called histiocytosis X, was introduced as a collective term to represent a spectrum of clinicopathological conditions ranging from the most benign form Eosinophilic Granuloma where there is only bone involvement with single or multiple osteolytic lesions to very debilitating form like Hand Schuller christian disease which is characterised by triad of bone lesions, exophthalmous and polyuria or the fulminant disease called letter-siewe disease which impairs the functioning of internal organs and presents with hepatosplenomegaly, lymphadenopathy, bone lesions, skin rash and pancytopenia.[9] The pathogenesis of Langerhans cell histiocytosis (LCH) is unknown. It has been debated whether LCH is a reactive or neoplastic process. Arguments supporting the reactive nature of LCH include the occurrence of spontaneous remissions, the extensive elaboration of multiple cytokines by dendritic cells and T-cells (the so-called cytokine storm) in LCH lesions, and the good

survival rate in patients without organ dysfunction.[10] On the other hand, the infiltration of organs by a monoclonal population of aberrant cells, the possibility of lethal evolution, and the cancer-based modalities of successful treatment are all consistent with a neoplastic process.[11]

LCH is usually encountered in children between 1- and 15 years old with a peak incidence between 2 and 4 years of age, with male predilection twice that of female.[12] Unifocal bone lesions are the predominant clinical form of LCH and choice of approach should take account of symptoms, the size of lesion and organ effected.[13] The relative frequency of organ system involvement is as follows: bone, 80%; skin, 60%; liver, spleen, lymph nodes, 33%; lungs, 25%; orbit, 25% and maxillofacial, 20%.[14]

The present case thus belonged to the category of eosinophilic granuloma with evidence of unifocal lytic lesion without any systemic manifestation. Rhabdomyosarcoma which was considered as differential was ruled out with cytological findings which showed absence of tumour cells with mitotic figures.

Treatment for Langerhans cell histiocytosis (LCH) depends upon the individual patient; it may differ depending on the type and severity of the condition as well as what parts of the body are affected. Due to site, extension of lesion and age of our patient, chemotherapy was chosen as treatment. After such systemic therapy our patient showed gross improvement.

CONCLUSION

A careful clinical examination, good diagnostic skills and awareness of characteristic cytological features of LCH lead to earlier diagnosis and treatment with minimum deformity. Although a unifocal lesion of LCH makes treatment successful, the possibility of recurrence still remains making a long term follow up necessary.

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