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Fine Needle Aspiration Cytology (Fnac) Diagnosis Of Langerhan Cell Histiocytosis: Presenting With Multifocal Involvement – A Case Report

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ABSTRACT: Langerhans cell Histiocytosis (LCH) is a rare disease affecting predominantly children.A case of LCH may involve single organ, single system or it may involve single site (unifocal),multiple site, multifocal or it may involve multiple organ(multisystem). Here a case of multifocal LCH in a two and half year old child is presented where FNAC was helpful in achieving a rapid and accurate diagnosis in an appropriate clinical and radiological setting.This can avoid unnecessary biopsy and guide the management especially when patient is very sick and doesnot give concent for biopsy . The highly characteristic common and rare cytological features are highlighted with focus on differential diagnosis.

Key words – Langerhans cell histiocytosis(LCH), Fine needle aspiration cytology(FNAC).

I. INTRODUCTION

Langerhan cell histiocytosis is a rare disease ranging from a benign to a rapidly fatal condition affecting predominantly young children(1) may be seen in young adult and also in latter part of life with male predilection(2,3,4). Langerhan cell histiocytosis is a disease involving clonal proliferation of the Langerhan cells(5).The pathogenesis is ill understood; whether it is a reactive process or neoplastic is still not defined. Spontaneous resolution occur in 10% - 20% cases and hence initial period of observation is often advisable. Langerhan cell histiocytosis (LCH) is the preferred terminology over histiocytosis X,

Eosinophilic granuloma,Abt-leterrer Siwe disease,Hand –Schuller-Christian disease and reticuloendotheliosis(4).All of them share common histogenesis and have similar morphological findings(5).We report here a case of Langerhan cell histiocytosis (LCH) of cervical lymph node along with bone and cutaneous involvement.

II. CASE REPORT

A 2 year old male child presented with multiple bilateral enlarged lymphnodes in the cervical region along with skin lesions resembling seborrhoic dermatitis involving scalp, face and trunk for last one and half

years. It was gradual on onset and no associated itching. The patient also had fever on and off accompanied by cough for last few days. The mother of the child complained of decreased appetite and delayed milestones and also gave history of excessive urination by the baby for last 1year.On physical examination the patient was irritable, bilateral multiple enlarged cervical lymphnodes measuring 2x2cm. On systemic examination showed mild bilateral proptosis with features of delayed milestones. Peripheral blood smear showed microcytic hypochromic picture, haemoglobin 6gm%,DLC N72%,L14%,M6%, E8% . Platelet count was within normal limit .TLC was 11920 cell / cu mm of blood. Bone marrow aspiration study showed normal haematopoesis with moderately increased eosinophils. FNAC of cervical lymphnode yielded blood mixed materials. Ethanol fixed and air dried smears were prepared and stained with Pap and Giemsa stains respectively.Smears were hypercellular and showed many atypical histiocytes as predominant cell types scattered singly and in loosely cohesive clusters.The cells have moderate amount of cytoplasm with enlarged vesicular nuclei.There were prominent nuclear grooves with coffee bean appearance. Nuclear lobulations were also observed. These cells displayed marked pleomorphism with variation in size and shape of cells and nuclei. Some of these cell showed cytoplasmic processes. Most were mononuclear and some were binucleated and multinucleated. Multinucleated giant cells have folded nuclei similar to mononuclear atypical histiocytes and easily differentiated from reactive multinucleated histiocytic giant cells. Occasional mitotic figures were also noted. These were admixed with polymorphous population of eosinophils, nutrophils, plasma cells and foamy histiocytes.

Radiological survey of the skeleton showed multiple lytic lesions in the skull over the frontal and parietal bones corresponding to the area of skull lesions.Multiple lytic lesions were also present in the radius and the femur (Fig). On the basis of clinical, radiological and cytological findings a diagnosis of Langerhan cell Histiocytosis was made and biopsy was adviced for confirmation.Initial management was planned and biopsy was advised for confirmation .However,after mild improvement during the stay in the hospital the patient took discharge against medical advice and could not be followed up.

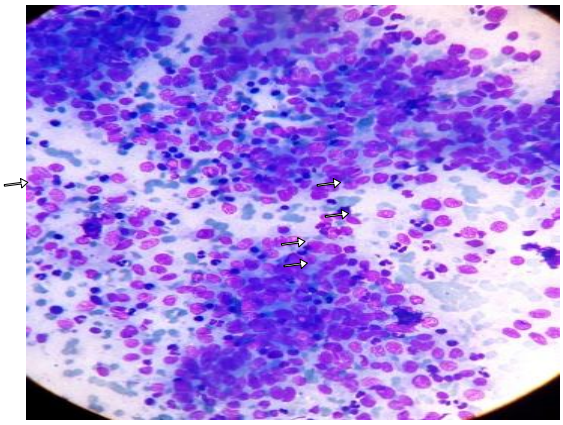


Fig.: Cytopathology of L.C.H. showing histiocytes with grooved nuclei (High Magnified)

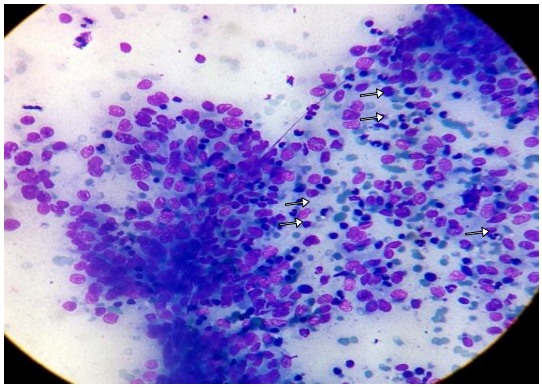


Fig.: Cytopathology of L.C.H. showing Eosinophils (High Magnified)

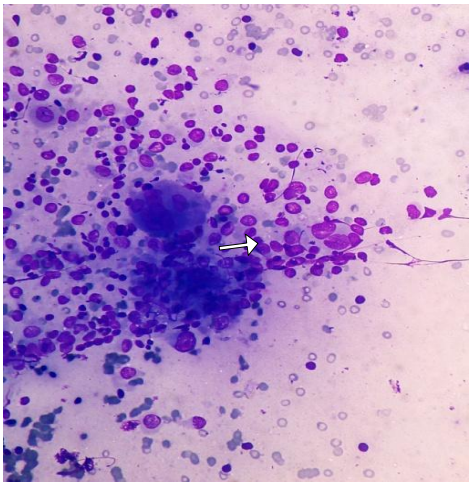
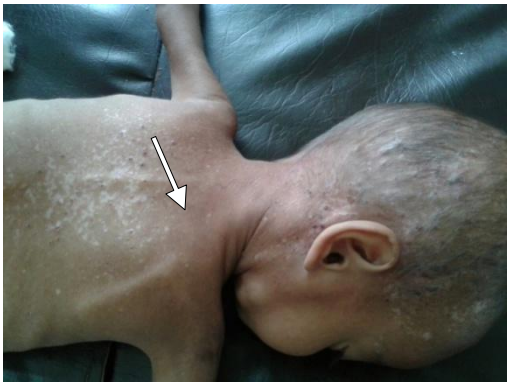
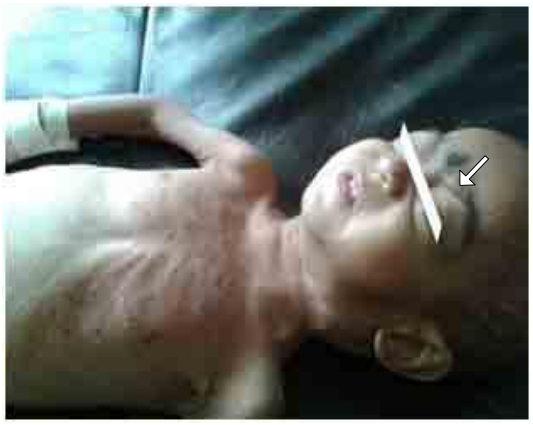


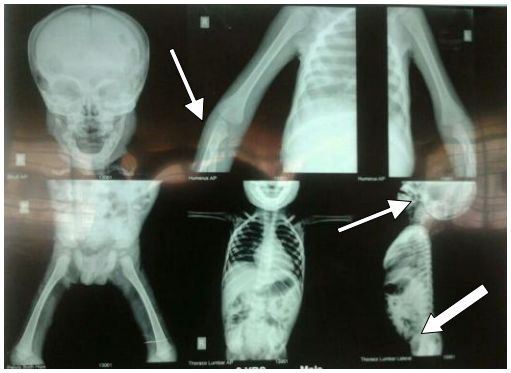
Fig.: Cytopathology of L.C.H. showing Histiocytic Giant Cells (High Magnified)



Photograph showing Skin Lesions



Photograph showing Exopthalmos



Radiograph of Skeleton showing multiple lytic lesions

III. DISCUSSION

Langerhan cell histiocytosis(LCH) is a rare disease caused by abnormal proliferation of antigen presenting cells of dendritic lineage, known as Langerhan cell (8).It has reported incidence of 0.2-2.0 per100000 children under age of 15 years(8,9).LCH can be subdivided in three clinicopathological entities: acute disseminated LCH, chronic unifocal ,and multifocal unisystem and multisystem LCH (7).LCH results from clonal proliferation of immunophenotypically and functionally immature morphologically rounded LCH cells(6).Presence of either CD1a cell surface antigen or the findings of Electron microscopic Birbeck granules in the lesional cells are valuable diagnostic adjuncts.(6)

The disease has got varying spectrum ranging from single osteolytic lesion(Eosinophilic granuloma) seen in children between 5 and 15 years of age, multisystem disease (Hand Schuller –Christian –disease) with skeletal and extra skeletal , reticuloendothelial and pituitary gland involvements seen in children 1-5 years of age to rapid fulminant cause (Letterer-Siwe-disease) seen most commonly less than 2 years of age(8,10).Hypothalamic pituitary axis involvement shows varying degree of

involvement. Most common is diabetes insipidus and after that growth hormone involvement(8,11,12).In children growth hormone involvement shows mainly decreased growth velocity (8,11).Other organs which are also commonly involved are lung , bone and skin. Patients who presents with isolated diabetes insipidus (DI) should be observed for onset of other symptoms or signs characteristic of LCH since 80% of them may have involvement of other organ systems(6,13). Cervical nodes are most commonly involved(6).The distinction between single and multisystem disease is very important as management and prognosis depends on this differentiation (7).In case of our patient the diagnosis of LCH was made on the basis of FNA which showed characteristic features of LCH after excluding cytological features of other differential diagnosis (NHL ,Ewing's sarcoma,Osteomyelitis).This was corroborated by characteristic radiological and clinical setting. Apart from cervical lymphadenopathy (clinically),Lytic bone lesions(radiologically)our patient had cuteneous lesions (in scalp and body)with polyurea ,growth retardation and mild exopthalmos thus falling in to the category of multifocal disease(Hand schuller-christian-disease)which is a rare findings.

Fine needle aspiration cytology (FNAC) is a rapid useful technique for immediate diagnosis that allows concurrent institution of therapy(7,14).The typical cytopathological picture consists of a mixture of abundant predominantly dissociated atypical Langerhan cells accompanied by many eosinophils and a varying numbers of neutrophils and lymphocytes, macrophages and multinucleated giant cells with pale ill defined eosinophilic cytoplasm and lobulated nuclei with longitudinal grooves best visualised in pap smears(7,14,15).

Management of LCH is controversial because of unpredictability of outcome and possibility of spontaneous healing(7).Modalities vary from observation,curettage,interventional steroid,low dose radiation,high dose systemic corticosteroid and chemotherapy, bone marrow transplantation and antibody therapy for recalcitrant cases. Rapid onset cases require active intervention(7). Our patient had multifocal involvement and treatment was planned accordingly. However we could not trace the patient after discharge.

IV. CONCLUSION

THE PRESENT CASE HIGHLIGHTS THE ROLE OF FNA IN THE DIAGNOSIS OF A RELATIVELY RARE DISEASE NAMELY LCH IN CHILDREN HAVING UNUSUAL CLINICAL PRESENTATION OF MULTIFOCAL INVOLVEMENT OF LYMPHNODES,SKIN, SKULL AND SKELETAL BONES, PITUITARY AND ORBIT. SINCE FNA IS EASY, NONINVASIVE PROCEDURE AND CAN BE PERFORMED IN OPD ,IN AN APPROPRIATE CLINICO-RADIOLOGICAL SETTINGS,A TYPICAL CYTOPATHOLOGY ALONE CAN BE USED FOR EFFECTIVE DIAGNOSIS AND DEFINITE PROOF OF LCH.

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