

## LANGERHANS CELL HISTIOCYTOSIS OF SHAFT HUMERUS: CASE REPORT

Sanjay Kumar\*, Abha Jyoti and Ajay Raj Pal

Department of Orthopaedics, GSVM Medical College, Kanpur-208002

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### ABSTRACT

A case (4yrs old male child) of Langerhans cell histiocytosis (LCH) of right distal humerus presented with pain and swelling over lower third of right arm. X-ray showed osteolytic expansile lesion of right distal humerus involving metaphysis extending till metaphysio-diaphysial junction. Computed tomography (CT) revealed a large osteolytic expansile bony mass lesion involving lower end of humerus with eccentric cortical erosion and expansions of medullary cavity. Patient managed by Curettage of lytic lesion followed by G-Bone and autologous fibular bone grafting. Histopathological showed sheets of histiocytic cells which are ovoid cells with longitudinal groove and eosinophilic cytoplasm which are suggestive of Langerhans cell histiocytosis.

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### INTRODUCTION

A 4 year old boy was presented to our OPD with complain of pain and swelling over right arm near elbow since 8 month. Pain was localised to lower end of arm, insidious in onset, mild to moderate in intensity, increase on movement and relieved by taking medication and rest. Pain was associated with swelling which develop spontaneously, localised and gradually increase in size.

On physical examination the patient was afebrile with stable vital signs. There is 4x5 cm swelling present on anterior aspect of right arm at distal end. Margin of swelling is indistinct and skin over swelling was normal in colour and freely mobile, no redness, no pulsation or dilated vein seen over swelling.

Local temperature was not raised and there is tenderness present on deep palpation. Swelling was firm to hard in consistency. His elbow range of motion was limited by approximately 15 degree in terminal flexion and extension.

There was osteolytic expansile lesion of right distal humerus involving metaphysis extending till metaphysio-diaphysial junction on X-ray. Computed tomography (CT) revealed a large osteolytic expansile bony mass lesion involving lower end of humerus with eccentric cortical erosion and expansions of medullary cavity. No periosteal reaction seen. The differentiation from normal and abnormal tissue is sharp. No elbow effusion.



Fig 1 Clinical photograph of right elbow of patient.



Fig 2 X-ray right arm with elbow with forearm (AP/Lat)

\*Corresponding author: **Sanjay Kumar**

Department of Orthopaedics, GSVM Medical College,  
Kanpur-208002

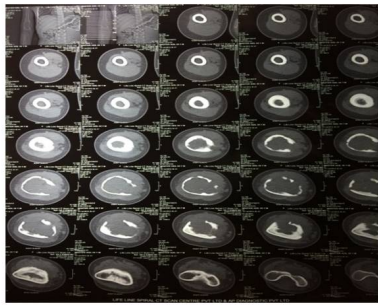


Fig 3 CT scan of right elbow with 3D reconstruction

FNAC was performed and microscopic features were aggregates of large histiocytes with nucleus displaying grooves and folding (coffee bean nucleus) and eosinophilic cytoplasm along with admixture of inflammatory cells, mainly eosinophils.

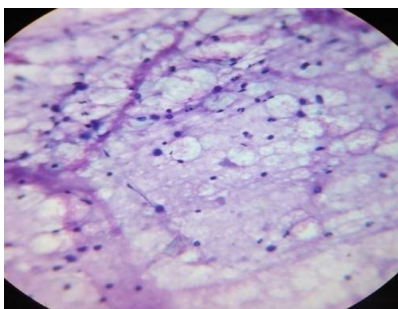
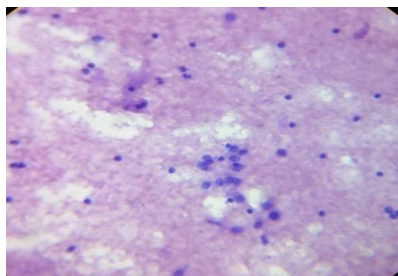
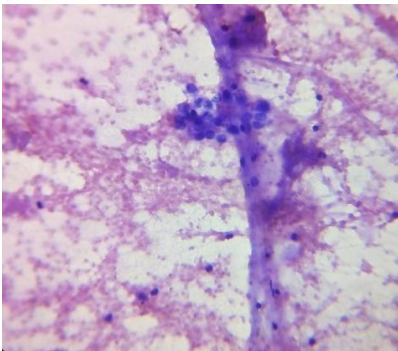


Fig. 4 a,b,c FNAC Cytological findings.

The patient was managed operatively by curettage of lytic lesion followed by G-Bone and autologous fibular bone grafting.



Fig.5 a,b,c Intraoperative picture



Fig.6 Post op Xray

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The tissue was sent for histopathological examination which was reported as follows:-

**Gross-**Specimen consists of multiple greyish white soft to firm tissue pieces altogether measuring 3.5\*3 cm.

**Microscopic Findings-** H&E stained sections show sheets of histiocytic cells which are ovoid cells with longitudinal groove and eosinophilic cytoplasm. These are mixed with inflammatory cell infiltrate comprising of eosinophils, lymphocytes.

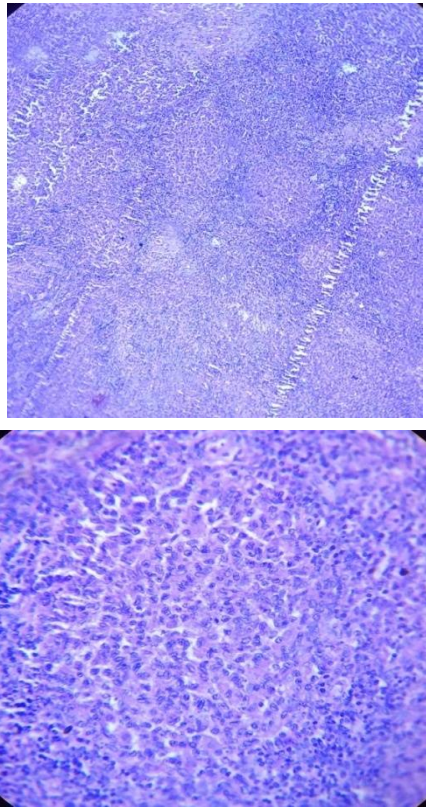


Fig. 7a,b Histopathological findings

**Impression-** Findings are suggestive of Langerhans cell histiocytosis.

### DISCUSSION

Langerhans cell histiocytosis is the terminology currently preferred over histiocytosis X, eosinophilic granuloma, Abt-Letterer-Siwe disease, Hand-Schuller-Christian disease, or Diffuse reticuloendotheliosis. LCH results from clonal proliferation of immune henotypically and functionally immature, morphologically rounded LCH cells along with eosinophils, macrophages, lymphocytes and occasionally, multinucleated giant cells. The term LCH cells is used because there are clear morphologic ,phenotypic ,and gene expression differences between Langerhans cells of the epidermis and those in LCH lesion (LCH cell).

The recent discovery that approximately 60% of LCH biopsy specimen demonstrate the V600E mutation in the BRAF oncogene, regardless of stage or organ involvement, has led to the conclusion that LCH is a clonal neoplastic disorder.

LCH may involve a single organ (single- system LCH), which may be single site (unifocal) or involve multiple sites (multifocal); or LCH may involve multiple organs

(multisystem LCH), which may involve a limited number of organs or be disseminated.

Generally, the choice of therapeutic regimen is based on disease severity. The international LCH study of the Histiocyte Society proposes the stratification of LCH cases by number of system involved. They further categorize those cases with single system involvement by number of sites with in those system (eg, monostotic vs polyostotic bone disease, solitary vs multiple lymph node involvement). Solitary bone lesion are treated locally with curettage or excision. Painful bone lesion may require intralesional steroid injection. Bisphosphonates such as zoledronic acid can also be used to reverse bone destruction and mitigate the pain of bony lesion. Early treatment with vinblastin and prednisolone has been suggested for bony lesion at vital anatomic locations requiring prompt resolution. Rarely, lesions that are unusually large and painful occur in inaccessible sites or involve vital structures require radiation therapy (3-6 Gy [300-600 rad]). Polyostotic bone lesion are best treated with indomethacin or a short course of systemic steroids. Systemic chemotherapy is indicated for multisystem disease and cases of single system not responsive to other treatment. The combination of cytotoxic drugs and systemic steroids is generally effective. Low-to-moderate doses of methotrexate, prednisolon, and vinblastin are used.

Primary musculoskeletal LCH is an uncommon disease and can be challenging to diagnose at initial presentation. Presentation of primary musculoskeletal LCH is similar to many other disease processes, such as tumor or infection. The key to differentiating eosinophilic gralunoma from other condition is histology, using open or needle biopsy. Diagnosing primary musculoskeletal LCH can spare the patient from receiving potentially harmful treatment for a benign and self limiting condition.

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