ISSN: 2577-2996

PROGRESS IN MEDICAL SCIENCES, 2023
VOL 7, NO. 1, PAGE 1 – 6
DOI: doi.org/10.47363/PMS/2023(7)186

CASE REPORT

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LCH Case Report in Adult- A Rare and Unusual Event

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a histiocytic neoplasm characterized by clonal proliferation of Langerhans type cells, a normal antigen presenting cells. It most commonly affects children, with an annual incidence of 2–5 cases per million inhabitants, it is extremely rare in adults. Herein we are reporting a case of LCH in 37 year old male which was initially diagnosed by FNAC and confirmed by histopathology and IHC performed later on.

ARTICLE HISTORY

Received December 26, 2022
Accepted January 03, 2023
Published January 31, 2023

Introduction

Langerhans cell histiocytosis (LCH) previously referred to as eosinophilic granuloma and/or histiocytosis X is a rare histiocytic neoplasm, mostly affecting children. Its etiology is largely unknown, however an abnormal immune response is thought to play a potentially important etiological role. Histopathology followed by IHC evaluation is the mainstay for its diagnosis. Herein we are reporting a case of LCH in adult male which is rare event, that too initially diagnosed by FNAC, which was later on confirmed by histopathology and IHC evaluation [1].

Case Report

37 years old male presented to us for FNAC with chief complaint of post auricular swelling since one month. On physical examination, post auricular swelling measuring 3x3 cm was noted, which was firm in consistency, freely mobile and having normal overlying skin. All his blood investigations were normal. Skull Xray showed multiple lytic lesion. His PET-CT scan stated "Hypermetabolic cervical, right supraclavicular, mediastinal and abdominal portocaval lymph nodes with hypermetabolic soft tissue opacification with erosion in the left mastoid air cells.

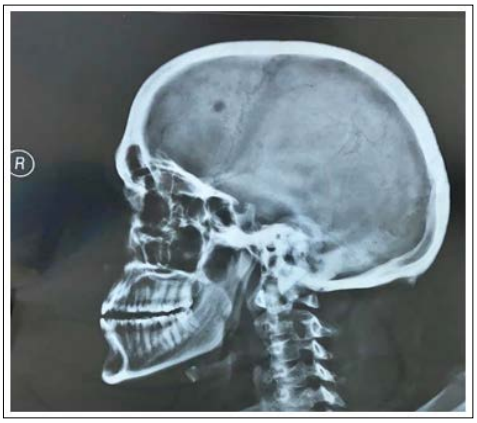

Figure 1: Skull Xray of Patient Showing Lytic Lesions

FNAC was done from post auricular swelling. On aspiration blood mixed material was aspirated, four air dried smears were prepared, all giemsa stained. Smears were highly cellular showing fair number of lymphoid cells admixed with numerous histiocytes, plasma cells, eosinophils and immunoblast. It was reported as lymphohistiocytic disorder, suggestive of LCH, and histopathological and IHC evaluation was advised for confirmation.

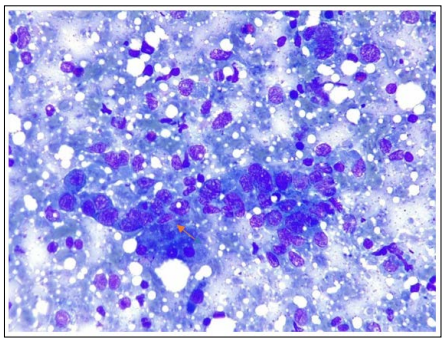

Figure 2: Cytology Smear Showing Cluster of Histiocytic Cell Admixed with Inflammatory Cells. Few Histiocytic Cells are Showing Nuclear Grooving (Marked by Orange Arrow) (Characteristic Feature of Langerhans Cell)

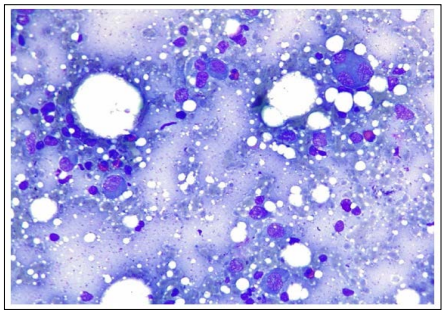

Figure 3: Cytology Smear Showing Fair Number of Eosinophils Admixed with Histiocytic Cells. One Multinucleate Giant Histiocytic Cell is Also Seen in this Smear

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Later on biopsy and IHC was performed. Histology sections showed partially effaced lymph node tissue infiltrated by large histiocytic cells having nuclear indentations, admixed with eosinophils, mast cells and lymphocytes. On IHC hstiocytic cells were positive for S100, CD1a and Langerin, while negative for CD20 and CD3. Based on histomorphology and IHC, diagnosis of LCH was made. No bone marrow involvement was found on bone marrow examination, performed later on.

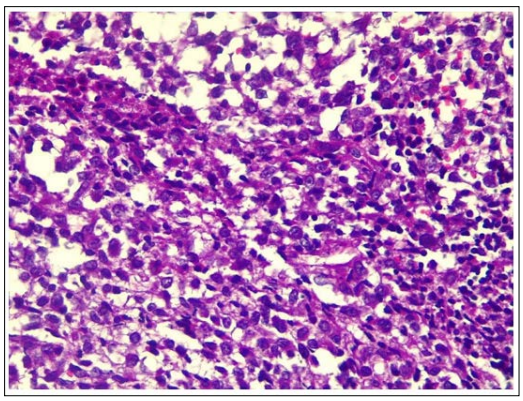

Figure 4: Histology Section Showing Effaced Lymph Node Architecture, With Lymph Node Infiltrated by Multiple Histiocytic Cells (Langerhans Cells)

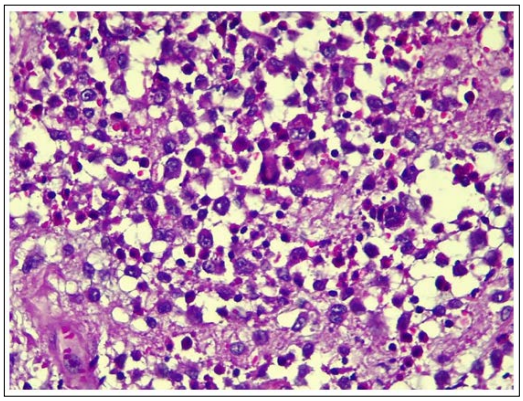

Figure 5: Histology Section Showing Langerhans Cells Which are Polygonal in Shape Having Abundant Eosinophilic Cytoplasm, Oval Nuclei with Prominent Nucleoli and Nuclear Indentation. Langerhans Cells are Admixed with Numerous Eosinophils, Lymphocytes, Plasma Cells and Mast Cells

Discussion

Rare systemic disease characterized by the abnormal overproduction of Langerhans cells that tend to infiltrate tissues and organ systems leading to organ damage. Langerhans cells are mononuclear dendritic cells with a function to present and process antigens to T lymphocytes. They are found normally in epidermis, lymph node, mucosa and bone marrow [2]. LCH predominantly affects pediatric population. It is extremely rare in adults with estimated frequency of approximately one to two cases per million persons [3]. In adults age range varies from 21 to 77 years with male predilection twice that of female [4,5].

Etiology of LCH is largely unknown, however it is considered as reactive disorder of immune regulation and not true neoplasm. Recent Studies have considered it as neoplastic lesion, due to monoclonal proliferation of lesional cells.

Depending on clinical features and age of presentation LCH is classified into three broad categories (1- Acute disseminated (2-Chronic localized and (3- Chronic disseminated [6,7]. Present case comes under chronic localized category. Clinical presentation depends on site of involvement. Most common clinical presentation is solitary or multiple lytic lesion in bones. In the present case multiple lytic lesion was seen in the skull bone.

LCH can involve various organ system of body such as bone, skin, lymph nodes, bone marrow, liver, spleen, lung, endocrine system, ear and brain, with highest frequency in bones followed by skin [8,9]. In the present case site of involvement is bone and lymph node. Estimated frequency of involvement for bone is 80% and for lymph node is 33% [8].

The diagnosis of LCH depends on clinical manifestation, light microscopy (histopathology and cytopathology), immunohistochemistry and electron microscopy. Light microscopy of LCH is characterized by presence of abundant Langerhans cells having grooved coffee bean like nuclei intermixed with eosinophils and plasma cells. Main Immunohistochemistry manifestation is positivity for CD1a and S100. In electron microscopy characteristic feature is presence of birbeck granules in Langerhans cells, langerin (CD107) is a monoclonal antibody which is involved in formation of birbeck granules, it has higher specificity than CD1a [10,11].

In the present case, similar histopathological and cytopathological features were seen along with LCH consistent IHC staining. Prognosis of LCH depends upon age of onset, site of involvement and numbers of organ involved. Single organ involvement carries better prognosis than multiorgan involvement. Skin and bone involvement have favourable prognosis, while prognosis is poor when lung, liver, spleen and bone marrow is involved [12]. In this case only bone and lymph node is involved, rest all organs and bone marrow are spared, so there is a chances of better prognosis for this case.

Conclusion

Purpose of reporting this case is that, it is rare and unusual in terms of age of presentation and was diagnosed by FNAC initially, later on confirmed by histopath and IHC.

Declarations

Ethics approval and consent to participate: Not applicable

Consent for publication: Not applicable

Competing Interests: Not interested

Author contributions: Case reporting

Funding: Not applicable

Availability of data and materials: Available

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