Orthopedic & Muscular System: Current Research

Santosh, et al., Orthop Muscul Syst 2015, 4:1
DOI: 10.4172/2161-0533.1000184

Case Report Open Access

Langerhans Cell Histiocytosis on Fine NeedleAspiration Cytology: A Report of 2 cases and Review of Literature

Santosh T*, Manoj Kumar Patro, Atanu Kuamr Bal and Anita Choudhury

Department of Pathology, MKCG Medical College, Berhampur, Odisha, India

Abstract

Langerhans cell histiocytosis (LCH) also known as Histiocytosis X is a clinico-pathologic entity characterized by proliferation of Langerhans cells (LCs) throughout the body including the reticulo-endothelial system, bone& skin. LCs is currently considered as a distinct type of histiocytic cell, not primarily phagocytic in nature. Fine needle aspiration (FNA) is 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 where access to histopathology is limited. We report two cases of LCH in a 3 ½ year old & 6year old patients based on FNA from the site of swelling.

Keywords: Langerhans cell histiocytosis; FNA; Swelling

Introduction

Langerhans cell histiocytosis (LCH), a disorder of antigen-presenting cells is the commonest disorder of the mononuclear phagocytic system. Owing to the relative rarity of the condition, it remains a disease in which thediagnosis is often delayed and in which many questions remain unanswered, ranging from etiology andpathogenesis to therapy [1]. LCH is characterized by clonal proliferation and excess accumulation of pathologic Langerhans cells. The diseasevaries widely in clinical presentation from localized involvement of a single bone to a widely disseminated life-threatening disease [2]. We report two cases of LCH where Fine Needle Aspiration (FNA) was helpful in establishing a rapid andcorrect diagnosis in correlation with radiology. The purpose is to highlight diagnostic cytological features which will help the pathologist in rendering a rapid and accurate cytologic diagnosis avoid unnecessary biopsy and guide for an early and appropriate management.

Case Report

Case 1

A 3 ½ year male child presented to the out patient department with complains of swellings in right frontal & parietal region of the skull for the last three months. After informed consent on examination these swelling were fluctuant, ill-defined soft tissue masses each measuring 2 x 2cm.The patient had no fever or loss of body weight. There was no evidence of lymph node enlargement or hepatosplenomegaly. Peripheral blood picture showed normocytic normochromic anemia. Haemoglobin was 11gm/dL. Differential count showed 40% neutrophils, 08% eosinophils, 1% basophils, 41% lymphocytes, and 10% monocytes. Platelet count was normal. Skull x-ray showed multiple punched out lytic lesion in calvarium (Figure 1).

Case 2

A 6 year old male child presented with swelling in scalp of size 3 x 3cmsince four months. The patient had no fever or loss of body weight. There was no evidence of lymph node enlargement or hepatosplenomegaly. Peripheral blood film showed microcytic hypochromic anemia. Haemoglobin was 9.5gm/dl. Differential count was normal with mild esoniphilia. Platelet count was normal. Skullradiograph revealed a single punched out osteolytic lesion in the parietal region of skull (Figure 2).

FNA was performed in both the cases in an out patient basis without any anaesthesia using a 24 G needle without image guidance as the lesions are superficial and palpable. Cytosmears in both the cases showed essentially identical features. The smears were cellular and are comprised of a mixture of abundant, predominantly dissociated histiocytes and eosinophils. They were accompanied variable number of neutrophils, lymphocytes, macrophages and multinucleated giant cells. The histiocytes were large cells with abundant, pale blue cytoplasm and round to oval, vesicular nuclei. Prominent nuclear indentations and grooves (with a coffee bean appearance) were observed in the mononucleate histiocytes and multinucleate giant cells. The eosinophils were of the natural type and showed bilobular nuclei and numerous large, eosinophilic granules. The cytologic findings were suggestive of LCH (Figure 3). FNA diagnosis of Langerhans cell histiocytosis

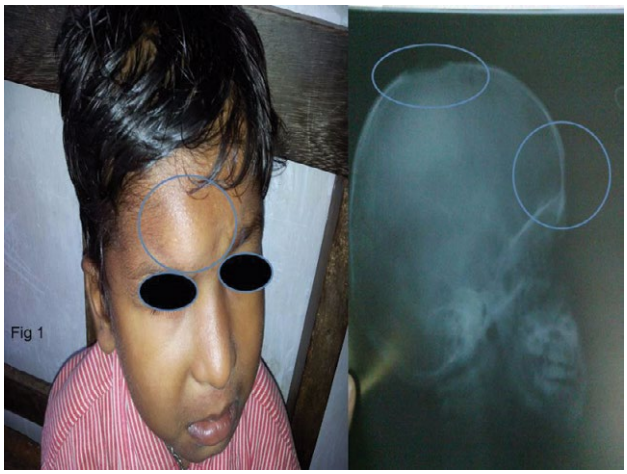

Figure 1: [a] Photograph of Case 1 showing a swelling in the forehead and [b] Photograph of X-ray skull showing two osteolytic lesions over forehead and parietal area.

***Corresponding author:** Dr. T Santosh, Ex PG Student, Department of Pathology, MKCG Medical College, Berhampur, Odisha, India, Tel: 08895495670/09584873660; E-mail: born_vss@yahoo.co.in

Received January 24, 2015; **Accepted** February 16, 2015; **Published** February 23, 2015

Citation: Santosh T, Patro MK, Bal AK, Choudhury A (2015) Langerhans Cell Histiocytosis on Fine NeedleAspiration Cytology: A Report of 2 cases and Review of Literature. Orthop Muscul Syst 4: 184. doi: 10.4172/2161-0533.1000184

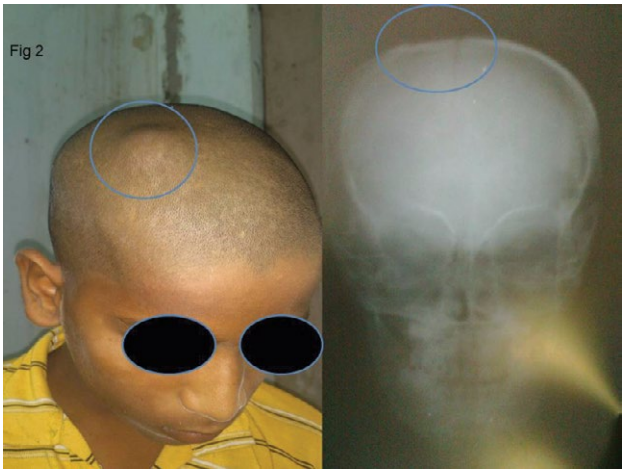

Figure 2: [a] Photograph of Case 1 showing a swelling in the right parietal region and [b] Photograph of X-ray skull showing an osteolytic lesion in the region.

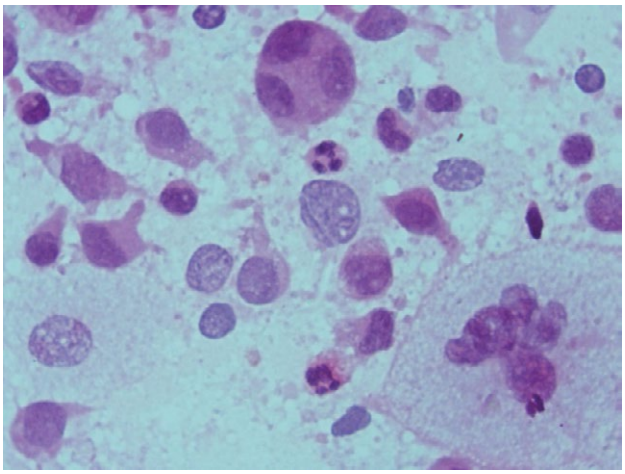

Figure 3: Cytosmears show diagnostic Langerhans cells admixed with eosinophils, histiocytes, polymorphs and giant cells. Langerhans cells are characterized by oval to reniform nuclei and abundant pale cytoplasm. The nuclei show prominent grooving, finely granular chromatin pattern and inconspicuous nucleoli. (H&E x40).

(LCH) was given in both the cases. Biopsy from site of lesion showed sheets of atypical histiocytes with nuclear grooves, polymorphous inflammatory cells comprising of eosinophils, neutrophils, lymphocytes in background. There was no necrosis, and no mitosis (Figure 4). On immunohistochemical staining, S-100 and CD1a were diffusely positive in all the atypical histiocytes.The patients subsequently received clinical staging for LCH. They were alive and well one month postoperatively and had regular follow-up in the out-patient clinic without any further treatment.

Discussion

Langerhans cell histiocytosis (LCH) is a rare group of disorders with a wide spectrum of clinical presentations. The term LCH was introduced as an alternative to histiocytosis X by Nezelof in 1973. Eosinophilic Granuloma, Hand-Schuller-Christian Disease and Letterer-Siwe Syndrome are the three conditions that are believed to represent different expressions of the same disorder, now known as

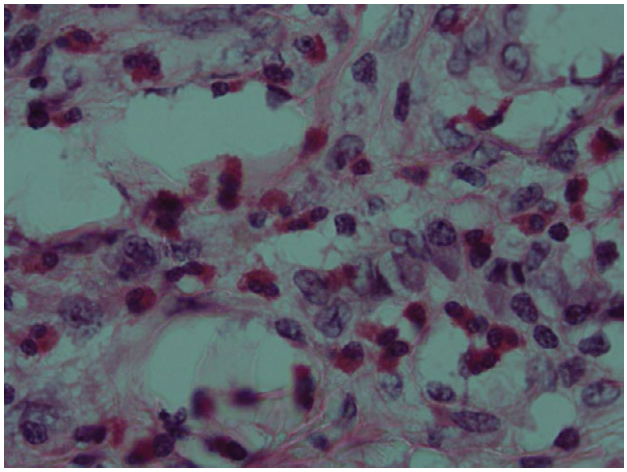

Figure 4: Histopathology sections of curettage material showed sheets of large pale Langerhans cells interspersed with large numbers of bilobed eosinophils. Prominent nuclear indentations or grooves of Langerhans cells are clearly seen. (H&E x100).

LCH [3]. LCH has an estimated annual incidence ranges from 0.5-5.4 cases per million persons [2].

The hallmark of LCH is the abnormal proliferation of the Langerhans Cells (LC). Paul Langerhans first described them in 1868 after making it visible by means of a gold chloride technique [4]. The LC is considered to be a dendritic cell of the epidermis, making up 1–2% of epidermal cells and is believed that it is derived from a multi-potent bone marrow stem cell. The LC is thought to be part of a spectrum of cells including macrophages, dendritic cells. It is a potent antigen-presenting cell that is essential for the integrity of the skin and of the immune system. After antigen encounter, they migrate to regional lymph nodes where they present antigen to paracortical T cells [5].

LCH may occur at any age, although the majority of the cases are diagnosed in children from newborn to 15 years. There is no significant gender difference. The clinical spectrum varies from a solitary lesion to multifocal unisystem to multisystem lesions with related symptoms. The unifocal form usually involves the bone, often seen in children between 5 and 15 years old. Systemic LCH is morecommon in children under 2 years of age. The multifocal unisystem form almost always occurs in the bone. Any bone can be involved, but more than 50% of lesions occur in the skull, spine, pelvis, ribs, and mandible. The multifocal multisystem form involves many organs including the bone, skin, liver, spleen, hematopoietic system, and lymph node [6].

The classical cytological features include high cellularity composed of sheets and many isolated LCs seen admixed with polymorphous population of numerous eosinophils, neutrophils, lymphocytes, plasma cells, multinucleated giant cells andmacrophages. The key to the diagnosis is to identify the LC through its characteristic features, namely nuclear grooves and nuclear pseudo-inclusions. They show variable degree of pleomorphism and mitotic activity.Degree of eosinophil infiltrationvaries in different areas of LCH lesion [7].

The most common differential diagnoses of skull lesions clinically included Ewing's sarcoma, non-Hodgkin lymphoma, and osteomyelitis. Ewing's sarcoma and non-Hodgkin lymphoma are characterized by monotonous population of small round blue cells. In acute osteomyelitis, the neutrophils forma prominent component. The reactive histiocytes are seen and can be easily distinguisheddue to

the absence of distinctive features of LCs. Chronic osteomyeltis shows predominantly plasma cells and lymphocytes [8]. Rarely, LCH can be associated with another malignancy such as malignant lymphoma, leukemia ormetastatic neoplasm [9].

LCs show positivity for S-100, PNA (peanut agglutinin), MHC class II, CD1a, and langerin (CD207). The Birbeck granule is their distinctive ultrastructural hallmark [10]. FNA can be used to establish the extent of disease or recurrence of LCH. For localized lesions in the skeletally immature patients, a simple minimally invasive form of treatment with a low rate of complication is desirable. In view of this and the possibility of spontaneous resolution in localized disease, FNA alone could be used to confirm the diagnosis [7].

Conclusion

Our cases highlight the role of FNA in the diagnosis of LCH in a child with usual clinical presentation. The cytologic features of LCH are highly characteristic to suggest a diagnosis in an appropriate clinical setting with classical radiological findings. A high index of suspicion, awareness of common and rare cytological features of LCH, its differential diagnoses and causes of diagnostic pitfalls is necessary. This can obviate the need of biopsy and electron microscopy. Immunochemistry can be performed on cell blocks prepared from the residual aspirates and is a useful adjunct.

References

1. Sheila W, Egeler RM (2008) Langerhans cell histiocytosis: Update for the pediatrician. Current Opinion in Pediatrics 20: 23–29.

2. Beverley PC, Egeler RM, Arceci RJ, Pritchard J (2005) The Nikolas Symposia and histiocytosis. Nat Rev Cancer 5: 488–494.

3. Nezelof C, Basset F, Rousseau MF (1973) Histiocytosis X: Histiogenic arguments for a Langerhan's cell origin. Biomedicine 18: 365-371.

4. Langerhans P (1868) UberdieNerven der menschilchen Haut. Virchow Arch Pathol Anat. 44: 325.

5. Michael PG, David FC, John PD (2002) Langerhans Cell Histiocytosis: Clinical Presentation, Pathogenesis, and Treatment from the LCH Etiology Research Group at The Children's Hospital of Philadelphia. The University of Pennsylvania Orthopaedic Journal 15: 67–73.

6. Windebank Kand Nanduri V (2009) "Langerhans cell histiocytosis," Archives of Disease in Childhood. 94: 904-908

7. Chandekar SA, Shah VB, Kavishwar V (2013) Cytological diagnosis of Langerhans cell histiocytosis with cutaneous involvement. J Cytol 30: 81-83.

8. Lee LY, Kang CJ, Hsieh YY, Hsueh S (2005) Diagnosis of nodal Langerhans cell histiocytosis by fine needle aspiration cytology, Chang Gung Medical Journal. 28: 735–739.

9. Egeler RM, Neglia JP, Arico M (1998) The relation of Langerhans cell histiocytosis to acute leukemia, lymphomas, and other solid tumors. The LCH-Malignancy Study Group of the Histiocyte Society, Hematology/Oncology Clinics of North America 12: 369–378.

10. Kumar N, Sayed S, Vinayak S (2011) Diagnosis of Langerhans cell histiocytosis on fine needle aspiration cytology: A case report and review of the cytology literature. Patholog Res Int 2011: 439518.