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Cytological Diagnosis of Langerhans Cell Histiocytosis;A Study At Medical College Hospital

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I. Introduction

Langerhans cell histiocytosis (LCH) is a rare intricate neoplasm affecting predominantly children. It can present as a single isolated lesion requiring no treatment or as a multisystem, life-threatening disorder necessitating aggressive therapy[1]. This disorder is characterized by abnormal, clonal proliferation of antigen-presenting cells, the Langerhans cells (LCs), associated with immunosurveillance and immune regulation. [2] It remains a debatable matter as to whether LCH represents a reactive or neoplastic process.[3] The exact pathogenesis of the disease is, however, unclear.[4]The clinical presentation may be variable, with a clinical spectrum that ranges from an isolated bone lesion with a favorable natural history to a more aggressive disease with multisystem involvement (such as liver, lung, bone, spleen, lymph nodes, hypothalamus, pituitary gland, gastrointestinal tract).[5]The diagnosis of LCH in our patients was made on the basis of FNAC with radiology and clinical correlation. The cytologic diagnosis may be missed due to varied clinic-radiological differentials or due to the lack of characteristic cytological findings as a result of sampling error.Therefore it is obligatory for the pathologist to consider this diagnosis only in an appropriate clinical and radiological setting. It is also important to be familiar with cytological features of other differential diagnoses.

II. Materials And Methods

This study was conducted in the Department of Pathology, Government Medical College, Srinagar. It was a retrospective study done over a period of 7 years November 2009 to December 2016. Six cases of LCH diagnosed on fine-needle aspiration (FNA) were retrieved from the archives. The clinical data from the cytology forms and case files of the patients were collected. The clinical profile including the age, sex, and signs and symptoms with emphasis on the type of lesions were evaluated. FNA was performed by the cytopathologist The smears were air-dried for May-Grünwald-Giemsa (MGG) and wet-fixed in 95% alcohol for hematoxylin and eosin (H and E) and Papanicolaou staining. CD 1a immuno histochemistry was done on cell block preparation.

III. Results

All patients in our study were males with a mean age of 9 years.All the patients presented with swelling in skull ,frontoparietal region being the commonest site of involvement.(Table 1)

Table 1: Distribution of cases according to age, sex and site of lesion

Number	Age/sex	Location
1.	10/male	Parietal region
2.	12/male	Parietal region
3.	9/male	Occipital area
4.	6/male	Frontal region
5.	8/male	Frontal region
6.	10/male	Orbital area

IV. Discussion

LCH is rare, with an estimated incidence of approximately 2-5 per million per year.[6]In the past, the disorder was named as histiocytosis X which included eosinophilic granuloma, Hand-Schuller Christian disease and Letterer – Siwe syndrome. These three conditions actually represent different expressions of the same disorder, now known as LCH.[7] The clinical presentation in LCH is varied. It may present with a solitary lesion, multifocal unisystem involvement to multisystem lesions.[8]Any organ or system can be involved, but the skeleton, the skin and the central nervous system are more commonly affected.[9]In our study all patients

presented with swelling confined to the head and neck region, which included five cases of scalp swelling and one case of orbital swelling. The differential diagnoses clinically were tuberculosis, Ewing's sarcoma, non-Hodgkin lymphoma, and osteomyelitis. Althogh the diagnosis of LCH is based on hematologic and histological criteria,accurate cytological diagnosis of LCH is possible on the basis of characteristic cytological features in the presence of appropriate clinical and radiological setting.[10] The classical cytological features include high cellularity comprising of Langerhans cell admixed with polymorphous population of numerous eosinophils neutrophils and lymphocytes, multinucleated giant cells and macrophages. The key to the diagnosis is identification of large Langerhans cells with large, eccentric, kidney-shaped coffee bean nucleus.[11,12] Cytological smears in our study showed Langerhans cells (LCs) in a polymorphic infiltrate with a preponderance of eosinophils. LCs with grooved folded nuclei were identified(Figure1) A confirmed diagnosis was made after cell blocks showed positivity for anti-CD1a antibody(Fig 2). Ancillary studies may be always necessary for diagnosis in appropriate clinical and radiological setting. However, our patients had no other systemic involvement which was evident from the investigations done.

V. Conclusion

The cytologic features of LCH are quite characteristic to suggest the diagnosis with an appropriate clinical radiological back up . A high index of suspicion, knowledge about common and rare cytological features of LCH, its differential diagnoses is necessary. Immunochemistry if available can be performed on cell block. This can obviate the need of biopsy and electron microscopy.

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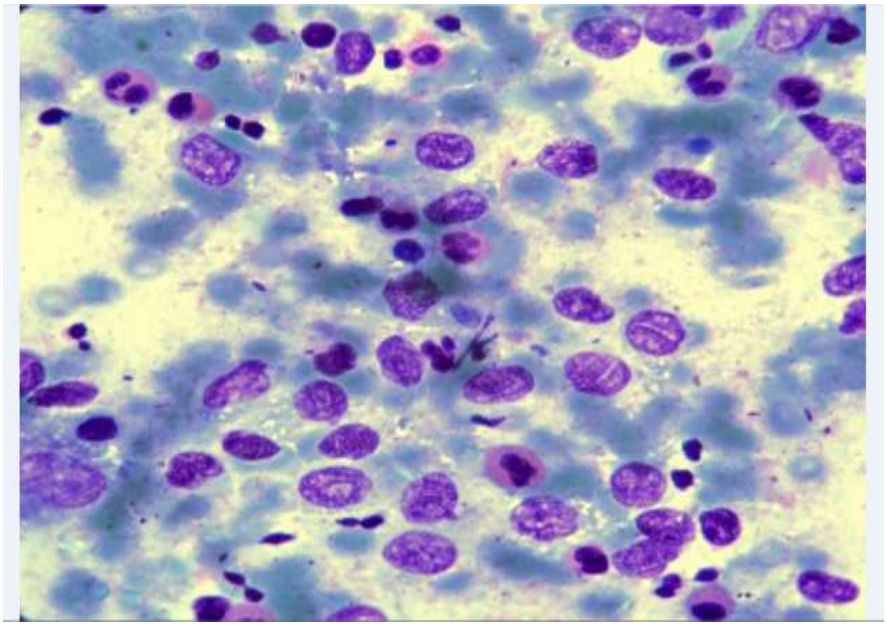

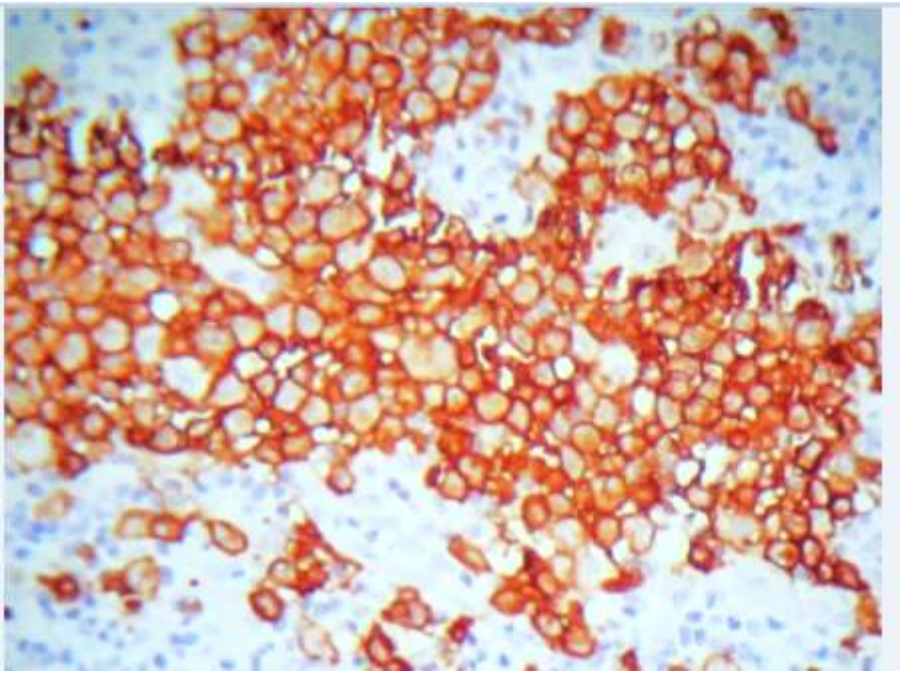

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