

Case Report

Touton like Giant Cell in Lymph Node in a Case of Langerhans Cell Histiocytosis

Mukhopadhyay S*, Mitra PK⁺, Ghosh S[#]

Abstract

Aspiration cytology of lymph node of a one-year old girl suffering from acute disseminated Langerhans cell histiocytosis revealed pleomorphic histiocytes and multinucleated giant cells, including Touton like giant cells. Histology of the same lymph node showed atypical histiocytes but no giant cell. We report this case because presence of Touton like giant cells in aspiration cytology smear of lymph node is a rare finding.

Journal of Cytology 2007; 24 (4) : 191-192

Key Words : Touton giant cell, langerhans cell histiocytosis.

Introduction

Langerhans cell histiocytosis (LCH) is a disease characterized by proliferation of Langerhans histiocytes present in lymph node, skin, thymus etc. The disease presents variable clinical course and prognosis depending upon whether there is uni-system or multisystem involvement. Multisystem LCH presenting at the first year of life is potentially fatal and is classified as acute disseminated LCH (Letterer-Siwe disease). Even with specific treatment, mortality rate is as high as 66% in patients who fail to respond to treatment.¹ The other types are not seen at this age. Identification of Langerhans cells in tissue is necessary for diagnosis of the disease.

Case Report

A one-year old girl presented with fever for one month. She had pallor, otitis media, cervical and axillary lymphadenopathy, hepatosplenomegaly and proptosis of right eye. The child was irritable. There was no skin lesion.

Routine haematological examination revealed an erythrocyte sedimentation rate of 30 mm in first hour and haemoglobin of 7 gm%. Platelet count was 1,25,000/cmm. Total white cell count was 5,000/cmm. No immature cell was found in the peripheral blood smear. Bone marrow smear revealed normo-cellular marrow, reduction in megakaryocytes, mild erythroid hyperplasia, normal myeloid population and no abnormal cell. Chest radiograph was normal.

Fine needle aspiration cytology (FNAC) of an enlarged cervical lymph node in the posterior triangle revealed lymphocytes at different stages of maturation, eosinophils, large number of Langerhans cells, that is, histiocytes showing atypia, abundant cytoplasm and vesicular nuclei with prominent nuclear grooves and indentations. The smear also showed multinucleated giant cells. Some of the giant cells had appearance of Touton like giant cells with wreath-like arrangement of nuclei along the periphery of the cell (Fig 1). A possibility of LCH was suggested and excision biopsy of the lymph node was advised for confirmation.

Biopsy of the same lymph node was done within a week of FNAC. Biopsy revealed effacement of architecture of the lymph node with small lymphocytes and histiocytes in diffuse arrangement. Nuclei of some of these histiocytes were kidney-shaped. Nuclear atypia was present in these cells. No giant cell was found in the

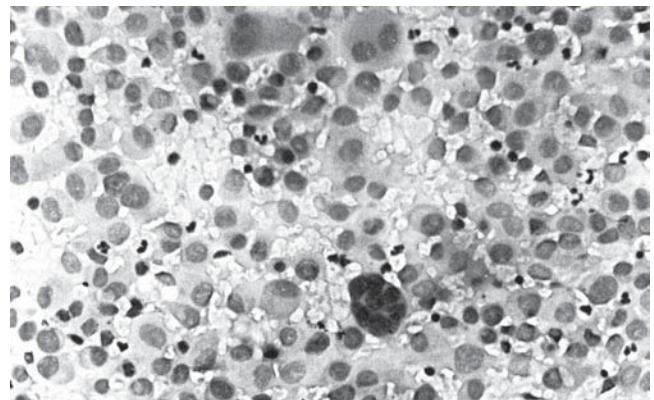


Fig. 1: Photomicrograph of FNAC smear of lymph node showing Touton like giant cells

*Assistant Professor, ⁺Professor of Pathology and Principal, Midnapore Medical College. West Bengal; [#]RMO cum Clinical Tutor, Regional Institute of Ophthalmology, Kolkata.

Received: 04.07.2007; **Accepted:** 15.10.2007

Corresponding Author: Dr. Subhalakshmi Mukhopadhyay, BB 41/8, Salt Lake City. Kolkata-700064.

E-mail: subhamukhopadhyay@yahoo.com

multiple tissue sections examined. Presumptive diagnosis of LCH was made, though immunohistochemistry or electron microscopic confirmation could not be done.

Unfortunately the child died of her illness within one week of the biopsy.

Discussion

In this case, presence of pleomorphic histiocytes in FNAC smears in abundance, eosinophils and lymphocytes were expected findings in support of the diagnosis of LCH but presence of multinucleated giant cells with morphology like Touton giant cell was the most striking feature. Histopathological findings were corroborative but giant cells were absent. Whether the lapse of one week between the FNAC and biopsy caused some change in the lymph node morphology was not known.

Usually a suggestive diagnosis of acute disseminated LCH is made by clinical findings like fever, hepatosplenomegaly, lymphadenopathy with matching radiological findings suggestive of pulmonary infiltrates and haematological abnormalities like anaemia and thrombocytopenia.² Bone involvement is uncommon in this group, except in mastoid region of temporal bone giving rise to otitis media. Cutaneous lesions are absent in about 20% of cases.² In the absence of skin lesions diagnosis is difficult. Identification of Langerhans cells in the tissue is important.

Occurring in infants, acute disseminated LCH, a malignant condition, is often diagnosed retrospectively.² According to Histiocyte Society, typical clinical and light microscopic findings can help in presumptive diagnosis.³ A definite diagnosis requires either CD1a positivity or demonstration of Birbeck granules by electron microscopy.³ CD1a is positive in 100 % cases whereas Birbeck granules are present in 2-69% and S-100 is positive in 2-100%.⁴ Literature review indicates that role of FNAC as a diagnostic technique in case of LCH has not been assessed or widely accepted till now, though a few reports are available.^{4,5}

The giant cells in the smear in this case, other than Touton giant cells, are possibly multinucleated histiocytes since these cells retained some of the nuclear features like irregular contour, prominent grooves and folds traversing in all directions and abundant eosinophilic cytoplasm as helpful identifying features. Foreign body type giant cells were not found in the smear, which are usually seen in the skin lesions of granulomatous LCH.

Though skin lesions of juvenile xanthogranuloma (JXG) often show giant cells of similar morphology, but presence of Touton giant cell in lymph node is not seen in any condition, to the best of our knowledge.

Lymph node involvement is not seen in cases of JXG with systemic complications.⁶ Moreover, this patient was never a case of JXG clinically as she had no skin lesion at all and clinically did not show any feature like JXG. Interestingly, FNAC was more revealing as well as informative than histology in this case. Presence of giant cells in FNAC, particularly those with morphological similarity with Touton giant cells prompted us to report the case.

References

1. Gardner H, Grois N, Arico M, et al. A randomized trial of treatment for multisystem Langerhans cell histiocytosis. *J Pediatr* 2001; 138: 728-34.
2. Walter HC Burgdorf. The histiocytoses. In: Elder D, Elenitsas R, Jaworsky C, Johnson B Jr, editors. *Lever's histopathology of the skin*. 8th ed. Philadelphia: Lippincott-Raven Publishers; 1997. p. 597-9.
3. Writing Group of the Histiocyte Society. Histiocytosis syndrome in children. *Lancet* 1987;1:208.
4. Kilpatrick SE. Fine needle aspiration of Langerhans cell histiocytosis of bone: are ancillary studies necessary for a definitive diagnosis? *Acta Cytol* 1998; 42:820-3.
5. Kumar PV, Mousavi A, Karimi M, Bedayat GR. Fine needle aspiration of langerhans histiocytosis of lymphnodes: a report of six cases. *Acta Cytol* 2002; 46:753-6.
6. Zvulunov A, Barak Y, Metzker A. Juvenile xanthogranuloma, neurofibromatosis and juvenile chronic myelogenous leukaemia. World statistical analysis. *Arch Dermatol* 1995; 131: 904.