Medico Research Chronicles

ISSN No. 2394-3971

Case Report

POSTMORTEM DIAGNOSIS OF PULMONARY RELAPSE OF LANGERHANS CELL HISTIOCYTOSIS ON FINE NEEDLE ASPIRATION CYTOLOGY

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Submitted on: December 2014
Accepted on: December 2014
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Abstract
Langerhans cell histiocytosis (LCH) is an unusual disease where pathologic Langerhans cells accumulate in various organs. Multiorgan involvement and young age of presentation are associated with frequent relapses and high mortality. We report a three year old girl, presenting with fatal pulmonary relapse following initial treatment, diagnosed on postmortem fine needle aspiration cytology. This case emphasizes the importance of cytology in the diagnosis of LCH.
Keywords: Langerhans cell histiocytosis, Postmortem, Fine needle aspiration cytology, relapse.

Introduction

Langerhans cell histiocytosis (LCH) is a rare disease characterized by monoclonal proliferation and accumulation of histiocytes in various organs including the bone marrow, skin, lymph nodes, spleen and lungs. The annual incidence of this condition ranges from 0.5-0.54 per million persons, with a higher incidence in early years of life [1]. We report a treated case of LCH in a three year old girl, presenting with fatal pulmonary relapse; diagnosed on postmortem fine needle aspiration cytology. This case is being presented for its rarity and to highlight the usefulness of cytology in the diagnosis of LCH.

Case Report A one year old female child presented with moderate grade, intermittent fever of one month duration. X-ray of the skull revealed multiple punched out lytic lesions in the calvarium. Clinical examination of all the organs was unremarkable. The bone marrow biopsy showed nodular infiltrates of large cells with abundant pink cytoplasm and convoluted grooved nuclei rimmed by numerous eosinophils (Figure 1). A diagnosis of Langerhan cell histiocytosis was rendered. She was treated with limited radiotherapy and chemotherapy. Two years later she presented to the emergency department with shortness of breath, collapsed and died. A postmortem chest radiograph showed non homogenous opacities with air bronchogram in bilateral lung fields. A postmortem fine needle aspiration showed large histiocytes

and giant cells, few with nuclear grooves and interspersed reactive endobronchial cells, eosinophils, neutrophils and lymphocytes confirming the diagnosis of LCH (Figure 2)

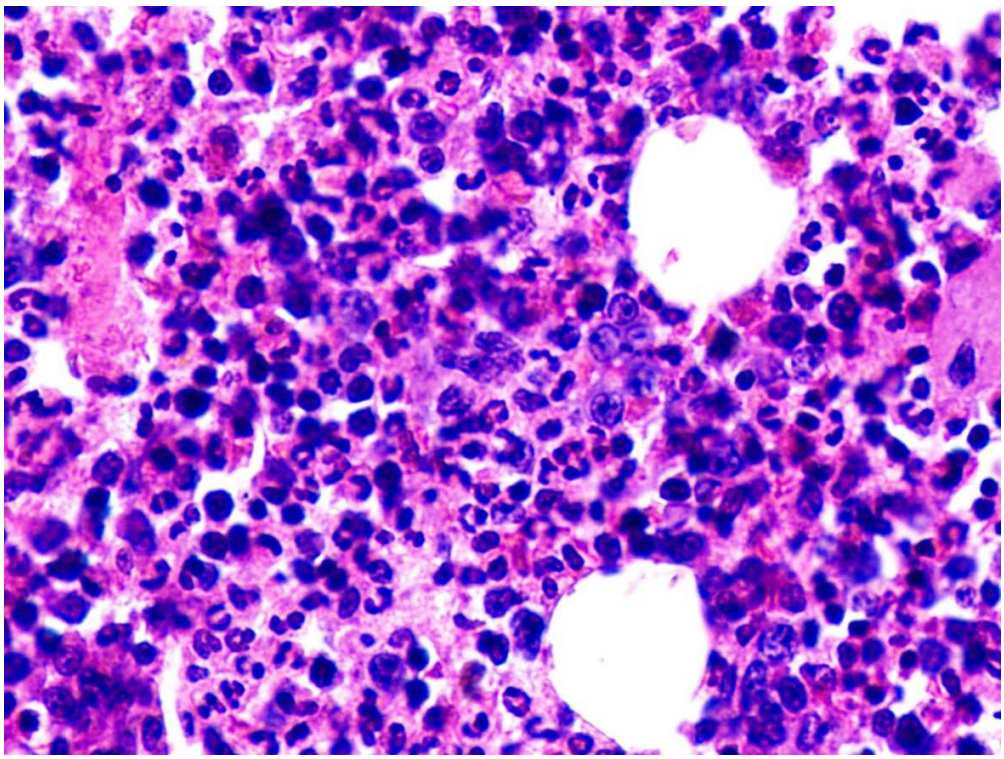

Figure 1: Bone marrow biopsy showing langerhans cells and dense eosinophilic infiltrate (H&E, X40)

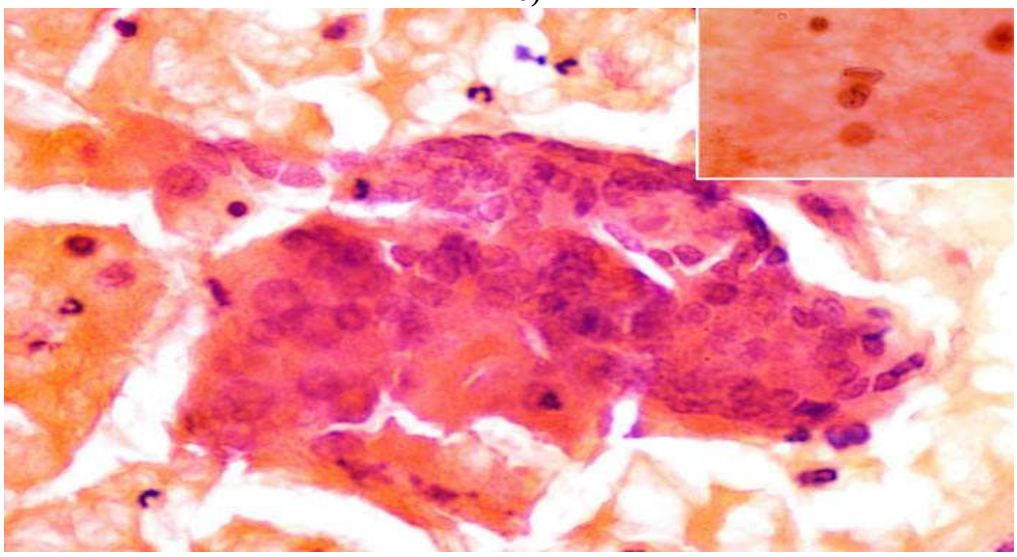

Figure 2: Fine needle aspiration cytology showing langerhans cells, eosinophils and multinucleated giant cells (PAP stain, X40). Inset: Langerhan cell with grooved nuclei (PAP, X100)

Discussion

Langerhan cell histiocytosis may involve a single organ or multiple organs. This condition encompasses various syndromes including Letterer- Siwe disease, Hand Schuller-Christian disease which involves lung and bones, eosinophilic granuloma and isolated pulmonary LCH. Letterer- Siwe disease is the most fulminant form affecting children less than 2 years of age with lesions in the bone, lung and reticuloendothelial system. (2, 3)

The most common age of occurrence of LCH is less than fifteen years with systemic LCH commonly affecting children below 2 years of age. The various organs involved in systemic LCH are bone (skull, spine, ribs, and pelvis), lung, liver, spleen, lymph node and hematopoietic system. (4)

The diagnosis of LCH is based on histopathological features of the lesion in corroboration with clinical history, radiology and haematology. It has been proved that cytology can be effectively used in the diagnosis of LCH thereby avoiding the need of other ancillary tests or techniques (4,5).

In cytology smears, LCH is characterized by highly cellular smears containing langerhans cells (LC) having nuclear grooves and pseudo inclusions admixed with numerous eosinophils, lymphocytes, plasma cells, giant cells and macrophages (4).

Immunoreactivity with S100, CD1a and langerin along with demonstration of Birbeck granules on electron microscopy aids in confirming the diagnosis (6). The present case was a treated case of LCH, with a pulmonary relapse confirmed on a postmortem cytology from lung lesion.

Prognosis of LCH is based on the age of occurrence, number of organs involved and the presence of organ dysfunction. Younger children with multiorgan involvement and dysfunction, especially lung, liver, bone marrow and spleen, have very high relapse and mortality rates. The mortality rates can be as high as 50% (1). Young age and multiorgan involvement by LCH portend poor prognostic factors. Our case was a case of LCH with bone and bone marrow involvement at the age of one year followed by a disease free interval of two years post treatment, and fatal demise due to pulmonary relapse. Postmortem diagnosis of LCH on biopsy of the autopsied organs has been in reported (8). To the best of our knowledge, the cytology diagnosis of pulmonary relapse leading to death has never been reported in literature.

Conclusion

The diagnosis of LCH is confirmed on biopsy however, in the setting of a classical clinical picture, fine needle aspiration cytology can be a very useful minimally invasive diagnostic tool.

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