Case Report

IAHS Medical Journal
Volume 07 Issue 01 June 2024; 102-104

An 18-Month Case of Langerhans Cell Histiocytosis : Diagnostic Challenges and Treatment Outcome

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ABSTRACT

Background: *Langerhans Cell Histiocytosis (LCH) is a rare disorder characterized by the abnormal proliferation of Langerhans cells, which can affect various tissues and organs, often mimicking malignancies. It primarily affects children and can present with a wide range of symptoms, making diagnosis challenging. The etiology of LCH remains uncertain, though recent studies suggest it involves the aberrant growth of Langerhans cells triggering an immune response.*

Case Presentation: *This was a case of an 18-month-old boy who came with fever of unknown origin in the Department of Paediatrics at BBMH who later on diagnosed as Histiocytosis.*

Conclusion: *This case highlights the importance of considering LCH in the differential diagnosis of paediatric patients presenting with systemic symptoms such as fever, organomegaly and anaemia. Early recognition and appropriate treatment, including chemotherapy and corticosteroids are crucial for favourable outcomes. Continuous follow-up is necessary to monitor therapeutic response and manage potential complications. This report contributes to the understanding of LCH and underscores the need for further research into its etiology and treatment protocols.*

Key words: Bone marrow biopsy; Haematological malignancies; Langerhans Cell Histiocytosis (LCH); Rare disease.

Introduction

Histiocytosis, also known as Histiocytosis X or Langerhans Cell Histiocytosis (LCH) is a set of uncommon illnesses affecting certain cells that typically play significant roles in the immune system. A collection of diseases collectively referred to as histiocytosis are defined by an aberrant rise in the quantity of certain immune cells known as histiocytes. These consist of dendritic cells, macrophages and monocytes. When a disease only affects one area of the body, it is commonly categorized as a single system, when it affects multiple areas of the body, it is categorized as a multisystem disorder. Although persons of all ages can develop LCH, children between the ages of one and fifteen years old account for the majority of cases of histiocytosis. Histiocytes can invade almost any organ or tissue, they are most likely to target the skin, bones, lungs, lymph nodes, central nervous system, and heart.[1] It is estimated that 1-2 out of every 200,000 persons get histiocytosis annually.[2] Recent research, however, suggests that the cause is the growth and development of an aberrant Langerhans cell, which then triggers the other cells of immune system to accumulate and form collections or tumours throughout the body. Certain forms are inherited.

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Date of Submission : 3rd March 2024
Date of Acceptance : 28th May 2024

Case Presentation

An 18-month-old boy immunized, son of non-consanguineous parents, belonging to low socio-economic status was admitted to Bangabandhu Memorial Hospital (BBMH) on October 2021 with a complaint of fever for 1 month with gradual pallor. After admission in the Paediatric department, it was found that the patient had average body built with ill-looking and hepatosplenomegaly, cervical lymph node enlargement, tender multiple nodules in the scalp. Initially patient was suspected of Tuberculosis/ Leukaemia. CBC shows neutrophilic leukocytosis and Hb was 7 mg/dl. 2 bag Packed Red Blood Cell (PRBC) was given. Hb electrophoresis and CSF study were normal. After doing PBF study, it shows Leucoerythroblastic blood picture, serum ferritin = 649ng/mL. An X-ray of skull was done which showed lytic lesions in Figure 1. FNAC was done from the swelling portion of skull and left side of cervical lymph node and found lymphohistiocytic proliferation which is

IAHS Medical Journal
Volume 07 Issue 01 June 2024; 102-104

shown in Figure 2. Later lymphoblastic proliferation was found in bone marrow study. After that, patient was treated with Injection Vinblastine for 1 year and now patient is fully cured.

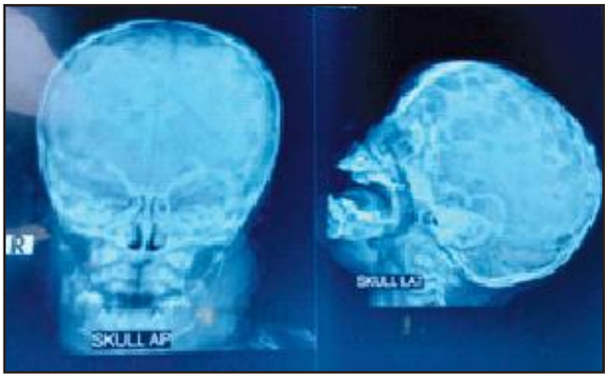

Figure 1 X-ray of skull showing lytic lesions

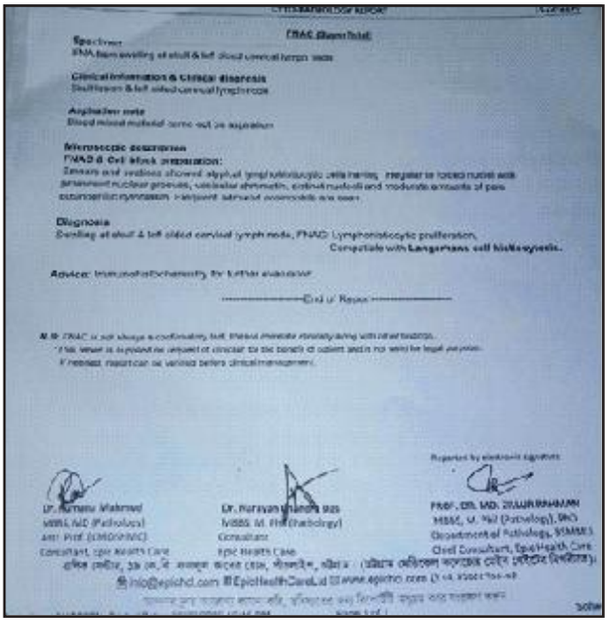

Figure 2 Cytopathology report depicting Lymphohistiocytic proliferation which is compatible with Langerhans Cell Histiocytosis

Figure 3 Situation of the patient during treatment

Figure 4 Improved condition of the patient after treatment

Discussion

For children under the age of fifteen, the annual incidence of LCH is 5-9 cases per 1000000 persons.[3] The features that LCH presents are incredibly varied. The most common ones include bone discomfort or fractures, cutaneous papules, lymphadenopathy, palpable tumours, polyuria and polydipsia associated with diabetes insipidus and exophthalmos. The most common types of skin lesions are polymorphic and multiple: painful ulcerations in skinfolds, vesicles, pustules and reddish-brown crusted papules. Multiple LCH papulovesicular lesions may be a sign of progressive multisystemic Letterer disease or more seriously, self-healing Hashimoto-Pitzker disease.[4] These lesions may even be present at birth. In contrast, one to three skin nodules are indicative of Langerhans cell histiocytoma, an uncommon form of LCH that is typically discovered at birth.[5] Lesions in the bones are common. They may not cause any symptoms at all or show up as swelling, persistent pain or in rarer cases, a fracture. Lytic bone lesions are indicative of childhood LCH and can be single (Eosinophilic granuloma) or numerous. Over 50% of these lesions are on flat bones, such as the skull, ribs or pelvis. [3]Vertebral plana may arise from vertebral body fractures, primarily in the thoracic spine. It is possible for bone diseases to spread into nearby soft tissues, such as the dura.A third of those with lymphadenopathy have LCH. The majority of young children with multisystem LCH have splenomegaly. Exophthalmos is a common orbital involvement of LCH condition that is usually bilateral in nature.

Chronic otorrhea or deafness may indicate mastoid involvement. The most common sign of hepatic involvement in the context of systemic Letterer-Siwe disease in young infants is significant hepatomegaly. Another uncommon but distinctive sign of LCH is bile duct involvement leading to sclerosing cholangitis. Children who have diffuse gastrointestinal tract infiltration may experience diarrhoea and malabsorption.[6] Diabetes insipidus can be brought on by posterior pituitary involvement, which affects 15% of children with LCH, anterior pituitary hormone deficiencies can also occur. The hematological involvement of LCH often results in splenomegaly, macrophage-activation syndrome or hepatomegaly or a combination of these. In this case, the patient suffered from fever, organomegaly, severe anemia and multiple swelling in the scalp. Initially it was thought as a case of leukemia or tuberculosis. Conventional treatment couldn't get the patient cured. After doing bone marrow study when it was diagnosed that patient is affected with Langerhans Cell Histiocytosis, then the patient

IAHS Medical Journal
Volume 07 Issue 01 June 2024; 102-104

was referred to Oncology department. LCH is a rare disease with a varied course of presentation, there are no recognised guidelines for treatment.[7,8,9] Among the available treatment options are radiotherapy, chemotherapy, surgery and steroid injection. Various methods were implemented at a Canadian centre from 1984 to 2007. In the majority of patients, the LCH I regimen (Prednisolone plus vinblastine or etoposide) was employed.[10] This patient was treated with Vinblastine and corticosteroid. It is the 1st line treatment.The first-line therapy regimen produces a response after six weeks in 65% of children with risk organ involvement and 86% of children without risk organ involvement.[11] The patient was advised to follow up regularly.

Conclusion

This case underscores the importance of considering LCH in differential diagnoses for paediatric patients with similar symptoms. Early and accurate diagnosis followed by appropriate treatment can significantly improve outcomes. Regular follow-up is essential to monitor the patient's response to therapy and manage any potential complications. This case adds as an evidence supporting the efficacy of first-line treatment for LCH and highlights the necessity for continued research into the etiology and optimal management strategies for this rare disease.

Disclosure

All the authors declared no competing interests.

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