

CASE REPORT

A Gluteal Mass Of Langerhans Cell Histiocytosis Mimicking Malignancy In A Two-Year-Old Boy: A Case Report

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Abstract

Langerhans cell histiocytosis is a disease primarily affects the bone. More than 50 percent of the disease occurs between the age of 1 and 15. We reported a case of a 2 year old boy who presented with a gluteal mass. Radiographic imaging showed an osteolytic lesion suspicious of malignancy. However, the histological diagnosis was Langerhans cell histiocytosis.

Keywords: Langerhans cell histiocytosis, gluteal mass, medical sciences

Introduction

Langerhans cell histiocytosis (LCH) affected 5.4 million children per year. The disease peaks at age 1 to 4 (1). Bone involvement with or without other associated sites is the most common manifestation of LCH, It has been observed in 80–100% of cases (1). Langerhans cells are a member of the dendritic cells family. The cells are believed to arise from multipotent bone marrow stem cells, which are efficient antigen-presenting cells for Tcell mediated immunity (1). Bone involvement of LCH is characterized by expanding erosive accumulation of Langerhans cells usually within the medullary cavity of bone. The aetiology and pathogenesis of LCH remain largely obscured. Fortunately, most cases demonstrate a favourable natural history without treatment (1). Radiologically, the destructive radiographic appearance of lesions may mimic the radiographic appearance of primary bone infection or sarcoma, such as Ewing sarcoma and osteosarcoma (1). For this reason, LCH is sometimes referred to as the “great imitator.” Therefore, it must be definitively differentiated from malignancy. The aim of this paper is to highlight the importance of LCH in the differential diagnosis of an osteolytic lesion in children.

Case Report

A two year-old boy presented with two weeks history of a left gluteal swelling associated with pain, a visible limp and mild fever. He had a fall while playing, two months prior to the swelling. On examination, he had a mild fever of 37.4°C but otherwise well and active. There was a diffuse and mildly tender gluteal swelling measuring 7 cm by 4cm. The range of movement of the left hip was reduced. Examinations of other systems were unremarkable.

Radiological investigations showed an osteolytic main bone lesion at the left ilium and a small lesion in the skull with features suspicious of malignancy. Biopsy led to the diagnosis of Langerhans cell histiocytosis. Immunohistochemical study showed positivity towards S100 and CD1a which confirmed the diagnosis. The patient was later commenced on chemotherapy with prednisolone and vinblastine. He responded well to the treatment. About 1 year after the diagnosis, he is ambulating with no residual limp.

Discussion

Skeletal involvement is one of the most common features presented in LCH which can occur in any bone. In this case, the patient presented with multifocal unisystem LCH. The age group and clinical presentation were consistent with the diagnosis.

Plain radiographs and MRI are the most useful mode of radiological investigation in predicting the nature of osseous disease. Aggressive pathological features with ill-defined margins, bone cortex destruction and soft tissue mass were present (Fig. 1). Hence, malignant diagnosis was seriously considered. However, radiological diagnoses can only suggest whether a lesion is of an aggressive nature or otherwise. Aggressive lesions do not necessarily indicate malignancy as benign bone diseases like LCH and osteomyelitis can also present with similar appearance (2).

On histology, features of LCH were distinctive (Fig. 2). Identification of only a few cells with the above histological appearance in any of these sites not necessarily means that the patient has LCH (3). Significant sizable number of cells needs to be present before the diagnosis can be entertained as exemplified in this case. LCH can also be diagnosed by fine needle aspiration cytology (4).

The diagnosis of LCH is based on the clinical features, histopathology, and special immunohistochemical techniques. For a definitive diagnosis, identification of Birbeck granules and CD1a antigens are required. Electron microscopy

for Birbeck granules could not be performed in this case due to practical constraint.

Laboratory studies are rarely helpful in LCH. We did not encounter eosinophilia. Immunoglobulin levels and tests of cellular immunity are usually within normal limits.

Treatment is directed by the clinical situation. The more aggressive approaches are used in patients with more extensive multisystem involvement (1). Surgical curettage, radiotherapy and chemotherapy can be used alone or in combination. Curettage is commonly performed for unifocal involvement. For a multifocal unisystem involvement, many patients experience spontaneous regression and other can be successfully treated by chemotherapy (5). Recurrence rate depend on the treatment method and location of the lesion. It was reported to range from 1.6% to 25% and patients should be closely followed up for a long period of time (6).

Clinical prognosis of patients with LCH will become worse with the growing number of organs involvement, number of organ dysfunctions, rapid disease progression and limited treatment response. Probably the most significant prognostic factor is the number of involved organs (6).

In conclusion, LCH presentation may closely resembles features of malignancy. Therefore, it should always be considered in a case of paediatric osteolytic lesion.

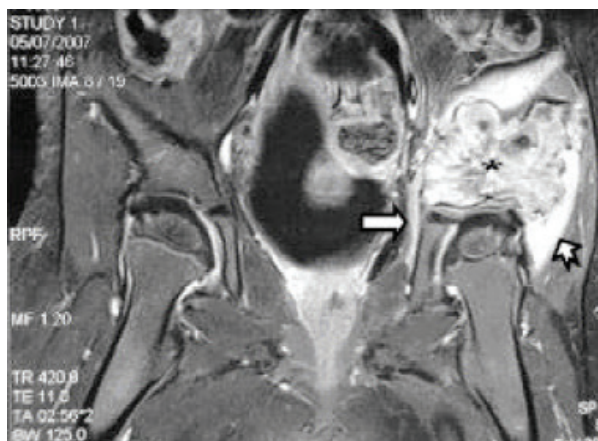


Figure 1: Gadolinium enhanced T1WI MRI in coronal plane showed avidly enhancing left iliac tumor mass (*) with involvement of the left gluteus medius muscle laterally (solid arrowhead) and the left obturator internus muscle medially (arrow). These features are highly suggestive of malignancy.

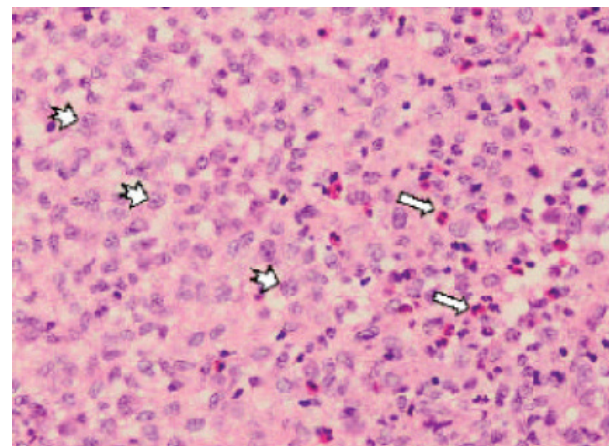


Figure 2: Biopsy revealed Langerhans cells histiocytosis with irregular nuclear margin, nuclear grooving and nuclear indentation (arrow heads) with abundant cytoplasm. There was eosinophils infiltration (arrow).

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