## **CASE REPORT**

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

### Zainal Abidin Ibrahim<sup>1</sup>, Wong Siong Lung<sup>2</sup>, Pan Kok Long<sup>3</sup>

- <sup>2</sup> Department of Radiology, Faculty of Medicine and Health Sciences, Universiti Malaysia Sarawak UNIMAS 93400 Kuching, Sarawak, Malaysia
- <sup>3</sup> Department of Orthopaedics, Faculty of Medicine and Health Sciences, Universiti Malaysia Sarawak UNIMAS 93400 Kuching, Sarawak, Malaysia

Submitted: 20 February 2007 Accepted: 3 December 2008

#### 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 ifferential 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.

<sup>&</sup>lt;sup>1</sup> Department of Paraclinical Sciences, Faculty of Medicine and Health Sciences, Universiti Malaysia Sarawak UNIMAS 93400 Kuching, Sarawak, Malaysia