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Case Report

Femoral Pain in a Child Revealing Monofocal Langerhans Cell Histiocytosis: A Radiologist Challenge

Fatima Chait[1]*, Nourrelhouda Bahlouli[1], Nidal Mrani Alaoui[2], Fadoua Boughaleb[2], El alami Zouheir[2], El Medhi Tarik[2], Naoual el Ansari[3], Najat Lamalmi[4], Nazik Allali[1], Siham El Haddad[1] and Latifa Chat[1]

[1]*Pediatric Radiology Department, Pediatric Teaching Hospital, Mohammed V University, Rabat, Morocco*

[2]*Pediatric Surgery Department, Pediatric Teaching Hospital, Mohammed V University, Rabat, Morocco*

[3]*Pediatric Hematology and Oncology Department, Children's Hospital in Rabat, Rabat, Morocco*

[4]*Anatomo-Pathology Department, Pediatric Teaching Hospital, Mohammed V University, Rabat, Morocco*

***Corresponding Author:** Fatima Chait, Pediatric Radiology Department, Pediatric Teaching Hospital, Mohammed V University, Rabat, Morocco.

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Abstract

Langerhans cell histiocytosis is a rare disease characterized by the proliferation of CD1a-positive immature dendritic cells, which can affect both sexes at any age. The lesions can be multifocal or localized. The organs most frequently affected are bone, lung, skin and endocrine glands.

Imaging plays a crucial role in the diagnosis and management of patients with Langerhans cell histiocytosis.

We report an observation of monofocal langerhansian histiocytosis in a 2-year-old girl involving the left femur. Histological examination revealed Langerhansian histiocytosis. The evolution was favourable after systemic chemotherapy.

Keywords: *Femoral Pain; Monofocal Histiocytosis; Imaging*

Introduction

Langerhansian histiocytosis (LCH), previously known as histiocytosis X which is a term that includes three entities: eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease, is a diffuse or localized disorder of the phagocytic mononuclear System, of unknown etiology, characterized by a high degree of clinical polymorphism and a prolonged uncertain evolution [1]. It is a rare disease, affecting mostly children: 2.6 to 8.9 per 1,000,000 child, with a male predominance [2]. Bone locations are the most common (60 - 90%) [3]. Unifocal involvement is more common than multifocal involvement [4].

This review describes the clinical and radiological manifestations of this disease, emphasizing the role of the radiologist in diagnostic orientation, differential radiologic findings, including common disease locations and appearances at various imaging modalities.

We report an observation of monofocal femoral Langerhansian histiocytosis in a 2-year-old girl with a good evolution.

Case Report

Two-year-old girl with no notable personal or family medical history, who presents left femoral pain, which motivated her parents to consult a pediatrician, the clinical examination found a mass at the upper left femoral extremity, firm and painful on palpation without limitation of the articular movement. The rest of the somatic examination is without particularities.

An MRI (Figure 2) and a CT scan (Figure 1) were performed and showed a lesional process of the left femoral diaphysis, lysing the bone cortex in places and infiltrating the bone marrow and the soft parts. Bone scintigraphy did not show any other secondary bone locations (Figure 3).

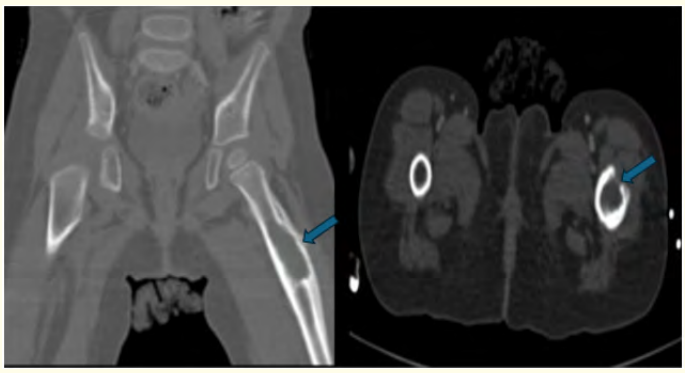

Figure 1: *Axial section and coronal reconstruction of a CT scan of the lower limbs with a bone window showing an osteolytic process (Blue arrow) in the superior part of the left femoral diaphysis with localized cortical rupture and multi-lamellar periosteal reaction.*

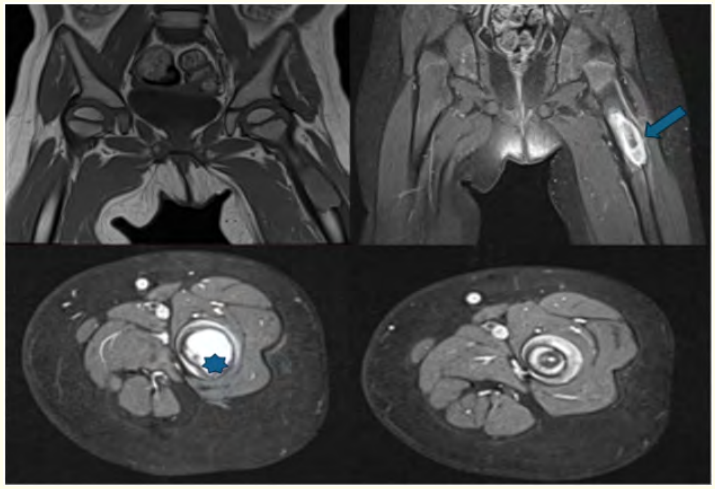

Figure 2: *MRI of the lower limbs showing a lesion process of the upper extremity of the left femur, in T1 isosignal and T2 hypersignal (Blue arrow), enhanced after injection of gadolinium, with rupture of the cortical bone in places and infiltrating the medullary bone (Blue asterix).*

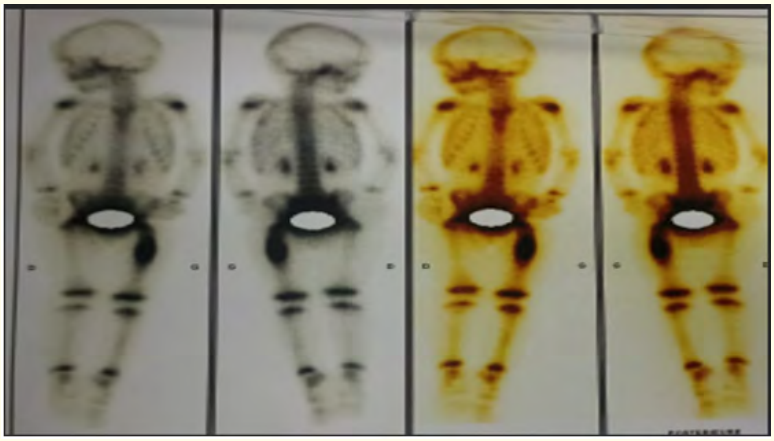

Figure 3: *Bone scintigraphy showing intense hyperfixation of the upper and middle parts of the left femoral shaft, without secondary localisation.*

The child then underwent a bone biopsy with an anatomopathological and immunohistochemical examination, which revealed massive infiltration of bone tissue by histiocytes with abundant eosinophilic cytoplasm and coffee-bean-shaped nuclei compatible with histiocytosis (Figure 4), then underwent chemotherapy treatment with a very positive evaluation, marked by a reduction in the size of the bone lesion.

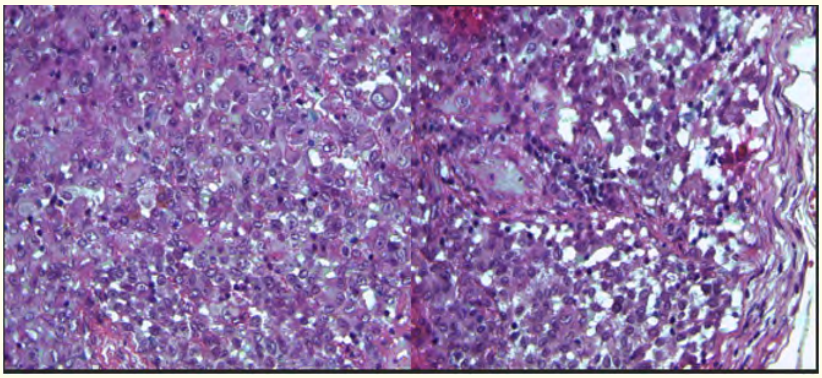

Figure 4: *HE GX 40 massive infiltration of bone tissue by histiocytes with abundant eosinophilic cytoplasm and coffee-bean shaped nuclei.*

Discussion

Langerhansian histiocytosis (LH) is a particularly rare disease with a wide clinical spectrum. The nomenclature of this disease has changed in the last 50 years. Historically LH was called "histiocytosis X", which is a term that includes three entities: eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease [5].

It can develop at any age. However, it peaks in children between the ages of 1 and 4 years with an estimated incidence of 2-9/1000 000/year and a slight predominance in boys (M/F = 1.2-1.4) [5]. Resulting from the monoclonal expansion of immunophenotypically and functionally immature Langerhans cells. The exact cause of LCH is unknown and it is not yet well understood whether it is a neoplastic or a reactive disease. LCH cells often have mutations in the BRAF gene, which explains the therapeutic effect of BRAF-specific agents [6].

According to recent studies, LCH is classified into three distinct forms: a single-site system (SS-s), a single multiple-site system (SS-m) and a multisystem type (MS) [7]. Approximately 65% of patients have the single-site system (SS-s) form [8].

The clinical presentation of LCH includes a wide variety of symptoms, varying from symptoms of severe life-threatening systemic disease, to the presence of non-specific lumps under the skin or a single asymptomatic bone focus [9].

Bone is involved in approximately 80% of histiocytosis cases. Monofocal involvement is more common than multifocal involvement [10]. with a predominance of the axial skeleton, with more than 50% of bone lesions occurring in the flat bones (skull, ribs, pelvis). Among the long bones, the femur is most commonly involved, followed by the humerus, tibia and vertebrae [9]. Symptoms can include pain, swelling of the affected area and pathological fractures. In our case, the patient had pain in the femur with swelling on clinical examination.

On imaging, the bone lesions of histiocytosis are described as osteolytic lesions that may infiltrate the medullary and soft tissue, which poses a diagnostic problem with other bone tumors and makes the radiological diagnosis of this disease difficult.

Diagnostic orientation is radiological, but confirmation is anatomopathological. Depending on whether the lesions are single or multiple, several situations are possible:

- Multiple localizations may suggest Langerhans cell histiocytosis (LCH) in the first instance.
- A monofocal bone lesion raises the question of age, location, local aggressiveness and radiological appearance.
- A slowly or moderately progressive lesion, with few signs of aggressiveness, suggests benign processes, benign tumours or circumscribed osteomyelitis.
- An aggressive lesion, which is not synonymous with malignancy, suggests malignant tumours, particularly Ewing's sarcoma, and osteomyelitis.
- Whatever the presentation, the presence of one or more bone lesions with diabetes insipidus (polyuria, polydipsia) should always prompt consideration of the diagnosis, although a biopsy is required to confirm it.

The definitive diagnosis of histiocytosis is based on anatomopathological and histological study which will show diffuse or focal growth of LCH cells with eosinophilic cytoplasm and coffee bean shaped nuclei, accompanied by eosinophilic infiltration [11].

The radiological assessment of our patient showed a lytic process of the two upper parts of the left femoral diaphysis infiltrating the medullary and soft tissues (Figure 1 and 2), with no metaphyseal or articular involvement and no other bone or visceral locations.

The treatment of bone damage is different, and depends on the mono or multifocal character and the location of the lesions. A single non-threatening bone lesion can be treated by curettage alone or by curettage with local injection of methylprednisolone [12]. The curettage biopsy, which is done for diagnostic purposes, can initiate the process of cicatrization.

Multifocal forms require chemotherapy alone or in combination with corticosteroids and/or radiotherapy [13]. First-line chemotherapy is vinblastine or etoposide. Radiotherapy is questionable due to the risk of secondary cancer in the field of radiation [14].

Nevertheless, it may be useful in the case of very extensive lesions [15]. Broadbent [16] reserves radiotherapy for lesions that threaten vital structures such as the spinal cord or optic nerve. Radical treatment of lesions is generally not recommended, since it may be damaging and increase the cicatrization process.

The prognosis of LCH is variable. It depends on the form of the disease, its location and the response to chemotherapy. In the case of unifocal LCH affecting a single bone, the prognosis is good. Spontaneous remission or disappearance of symptoms after local treatment has been observed. In forms with multifocal bone involvement, relapse of the disease is more frequent, therefore prolonged surveillance is necessary.

Conclusion

Langerhansian histiocytosis in children is a rare and polymorphic disease manifests across a broad spectrum of clinical presentations and radiological findings. While many imaging signs lack specificity, understanding and familiarity with these conditions help in their recognition within the differential diagnosis.

Bone involvement in this disease is one of the most frequent manifestations. The prognosis is often favorable. Management of the disease requires a bone and visceral evaluation, which determines the choice of treatment.

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Conflict of Interest

Declare if any financial interest or any conflict of interest exists.

Informed Consent

Written informed consent was obtained from the parents of patients for the publication of this case report.

Ethical Approval

Our institution does not require ethical approval for reporting individual cases or case series.

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