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Case Report

Multifocal Langerhansian Cell Histocytosis: About O Three Cases

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Abstract: Langerhans histiocytosis is a rare multi-systemic disease defined by the accumulation of Langerhans cells in the various organs of the human body, it predominates in children. Different localizations are described, Including the hematological localization characterized by the presence of histiocytes cell in the bone marrow. The aim of this work is to show the interest of the medullogram in the diagnosis of histiocytosis. We report 3 cases of Multifocal langerhanstic histiocytosis diagnosed in the hematology department in the Hassan II University Hospital in Fez. The diagnosis of Langerhans' histiocytosis was retained on clinical, cytological and anatomopathological criteria**.**
Keywords: Langerhans histiocytosis, multifocal, histiocytes cell.

INTRODUCTION

Langerhans histiocytosis is a rare multi-systemic disease associated with the accumulation of Langerhans cells in different organs. It is not a malignant disease.

All the body's organs can be affected but the most frequent are bone (80%), skin (33%), pituitary (25%) and haematopoietic system (15%) [1].

OBSERVATIONS

Observation 1

A 22-month-old child admitted for inflammatory swelling at the left forearm, extending to the wrist, the remainder of the clinical examination revealed hepatomegaly with no splenomegaly or skin's injuries. Radio of the forearm: Sign of osteolysis with periosteal reaction and rupture of the cortex. Anatomopathological findings: Hemophagocytic Histiocytosis caused by immune deficiency. Medullogram: Marrow hypercellular with the presence of some histiocytes in intramedullary (Figure 1).

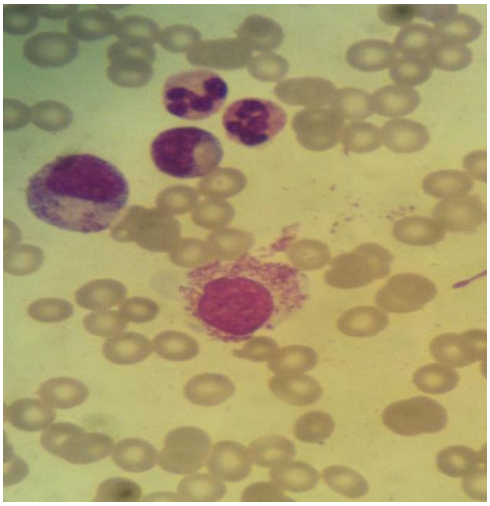

Fig-1: Image of histiocyte

Observation 2

A 2-year-old child admitted for pruritic erythematous Scams injuries in the scalp and for who the clinical examination has demonstrated 2 right cervical adenopathies, hepatomegaly with no splenomegaly. Abdominal echography: Liver discreetly increased in size, regular, homogeneous with absence of other abnormality. Cutaneous biopsy: Dermal localization of a systemic histiocytosis. Medullogram: Hypercellular marrow with intracellular histiocytes (Figure:2).

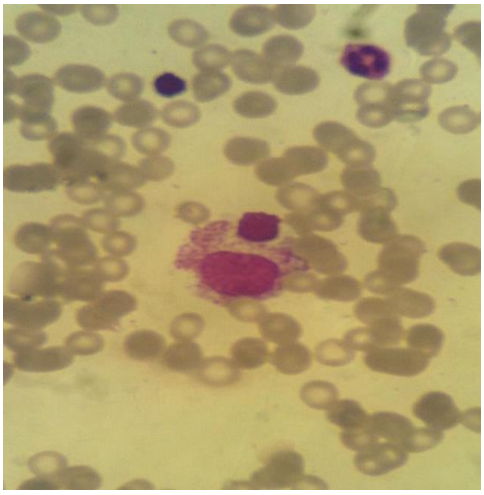

Fig-2: Image of histiocyte

Observation 3

A 4-year-old child admitted for scalp injuries descrobed as automata squamous plaque, the clinical examination has revealed a fever at 38 ° C, hepatosplenomegaly, cervical and inguinal adenopathies. Cutaneous biopsy for histiocytosis osteomedullary biopsy: Medullary infiltration by histiocytes. Medullogram: Hypocellular marrow infiltrated by histiocytes (Figure:3).

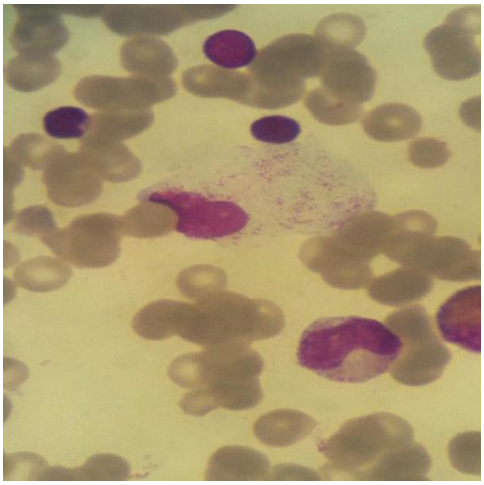

Fig-3: Image of histiocyte

DISCUSSION

Langerhans histiocytosis is characterized by localized or disseminated proliferation of cells with no histological criteria for malignancy and primarily affecting organs rich in reticuloendothelial tissue [2]. Langerhans histiocytosis affects in the majority of cases children more than adults [3].

The clinical spectrum of langerhansian histiocytosis is very wide, different localizations are possible, initially or during a relapse, the most frequent are followed.

-Bone damage: It may be single or multiple; the symptoms that lead to this diagnosis are pain, painless swelling and sometimes a pathological fracture [4].
-Skin involvement: appearing as papulosquamous lesions, sometimes petechial disseminated touching in the first place the scalp, the large folds and the thorax.
-Hematological damage: characterized by the presence of langerhansian histiocytosis cells in the myelogram. Its repercussion on hematopoiesis is more or less marked, It can sometimes be responsible for anemia or deep thrombocytopenia [4].
-Lung injury: responsible of interstitial syndrome, nodular image and localized pneumothorax [5].
-Hepatic damage: it is useful to distinguish acute involvement often multivisceral due to the initial thrust of the disease from chronic sequelae involvement [6].

Many other disorders can be present: hypophyseal, neurological, digestive, spleen, ganglions, subcutaneous tissue. Actually, the diagnosis of Langerhans histiocytosis is always based on histological or cytological examination [7].

In our series 3 cases of children whose age varies between 22 months and 4 years, cutaneous lesions are present in the two first cases and a swelling of the forearm in the 3rd one. the hepatomegaly was present in clinical examination for all the 3 cases, the lymphadenopathy in 2 cases, splenomegaly and fever in 1 case, Radiological signs were predominantly characterized by lesions of osteolysis in the patient with swelling of the forearm.

The diagnosis of histiocytosis was confirmed by pathological examination and reinforced by the myelogram which confirmed the presence of histiocytes in intramedullary.

Additionally, the cells of the langerhansian histiocytosis are large cells with an eccentric or kidney-shaped nucleus. They are often associated with inflammatory cells damage: Polynuclear eosinophils, lymphocytes, and macrophage cells. The accurate diagnosis can not be established if these cells are labeled with an anti-CD1a antibody [8].

Moreover, the detection of the S100 protein is less interesting because it is non-specific to the Langerhans cells as well as for the peanut lectin antigens, The search for Birbeck granules by electron microscope is also possible, very specific, but long and expensive.

Lastly, the histiocytic society has advocated a simplified classification taking into account only one variable: The number of affected organs (1 or 2 organs: localized forms, 3 or more organs: multisystemic forms) [9].

CONCLUSION

Langerhans histiocytosis is an oligoclonal proliferation of langerhans cells. The clinical spectrum is very wide; the diagnosis is always based on a histological or cytological examination. The treatment of langerhans' histiocytosis remains very controversial, as the prognosis is very variable and depends on age, focal or diffuse localizations and the presence of visceral damages as well.

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