ISSN: 0975-833X

*Available online at http://www.journalcra.com*

*International Journal of Current Research*
*Vol. 12, Issue, 05, pp.11771-11773, May, 2020*

**DOI: https://doi.org/10.24941/ijcr.38579.05.2020**

**INTERNATIONAL JOURNAL OF CURRENT RESEARCH**

## RESEARCH ARTICLE

# MONOFOCAL LANGERHANS CELL HISTIOCYTOSIS IN CHILD: CASE REPORT

**[1*]Benradi, L., [1]Nour, M., [1]Salhi, H., [1]Elaissaoui, F., [1]Aadem, A. and [1]Belahcen, M., [2]Haloui, A., [2]Mouhoub, M. and [2]Bennani, A.**

[1]Pediatric Trauma-Orthopedics Department, Mohammed VI Universitary Hospital Center, Oujda, Morocco
[2]Anatomopathology Department, Mohammed VI Universitary Hospital Center, Oujda, Morocco

## ARTICLE INFO

***Article History:***
Received 09th February, 2020
Received in revised form
14th March, 2020
Accepted 08th April, 2020
Published online 31st May, 2020

***Key Words:***
Childhood,
Benign tumor,
Surgery.

## ABSTRACT

Langerhans cell histiocytosis is a rare disease progressing by flares which can affect both sexes at any age. The lesions can be localiseted or multifocal; the organs most frequently affected are the bone, the lung, the skin and the endocrine system. We report a case of a young 13-year-old child with single focal clavicular langerhansian histiocytosis kept under surveillance with good clinical and radiological progress.



**Citation:** ***Benradi, L., Nour, M., Salhi, H., Elaissaoui, F., Aadem, A. and Belahcen, M., Haloui, A., Mouhoub, M. and Bennani, A.*.. 2020.** "Monofocal Langerhans cell histiocytosis in child: case report", *International Journal of Current Research*, 12, (05), 11771-11773.

## INTRODUCTION

Langerhans cell histiocytosis is a polymorphic disease which is characterized histologically by tissue infiltration by cells with markers and ultra-structural properties identical to Langerhans cells in the skin. The diagnosis is based on the anatomopathological examination of biopsies where an infiltrate of cells with a pleated reniform nucleus and an abundant clear cytoplasm is demonstrated. Treatment should be adapted to the severity of the disease. It can range from therapeutic abstention to chemotherapy. We report an observation of Langerhansian histiocytosis in a 13-year-old child diagnosticed and treated in Pediatric Surgery Department B, UHC Mohamed VI OUJDA with a good evolution.

### Observation

A 13-year-old child with no notable pathological history with a painful right clavicular mass that gradually increases in size.
The clinical examination found a conscious child stable on the hemodynamic and respiratory planes, with a mass at the internal edge of the right clavicle, firm and painful on palpation without limitation of the articular amplitudes. The rest of the somatic examination is without particularities.

***Corresponding author:*** **Benradi, L,**
Pediatric Trauma-Orthopedics Department, Mohammed VI Universitary Hospital Center, Oujda, Morocco.

The radiological assessment objectified a lytic lesion at the right clavicle internal edge (figure 1), the thoracic computed tomography eliminated an associated pulmonary attack and an abdominal echography returned without particularities (figure 2). We adopted a conservative treatment with the realization of a biopsy with immunohistochemical study which returned in favor of langerhansian histiocytosis (figure 3), given the absence of other associated slur, the decision taken was the therapeutic abstention with a clinical monitoring every 3 months. The evolution was marked by the regression of the disease with complete recovery after two years (Figure 4).

## DISCUSSION

Langerhans cell histiocytosis is a polymorphic disease which is characterized histologically by tissue infiltration by cells with markers and ultra-structural properties identical to Langerhans cells in the skin (Mathilde de Menthon, 2017). The first descriptions date back to the 1920s to 1940s, when Hand-Schüller-Christian disease affecting middle-aged children and adolescents and Letterer-Siwe disease which affects the small child and whose development is often spontaneously favorable were reported (Saliba, 2008). Epidemiological knowledge on this disease is poor, as an indication, the annual incidence rate is 2.6 to 8.9 per 1,000,000 in children (Jean Donadieu, 2017). Bone involvement affects approximately 50% of patients.

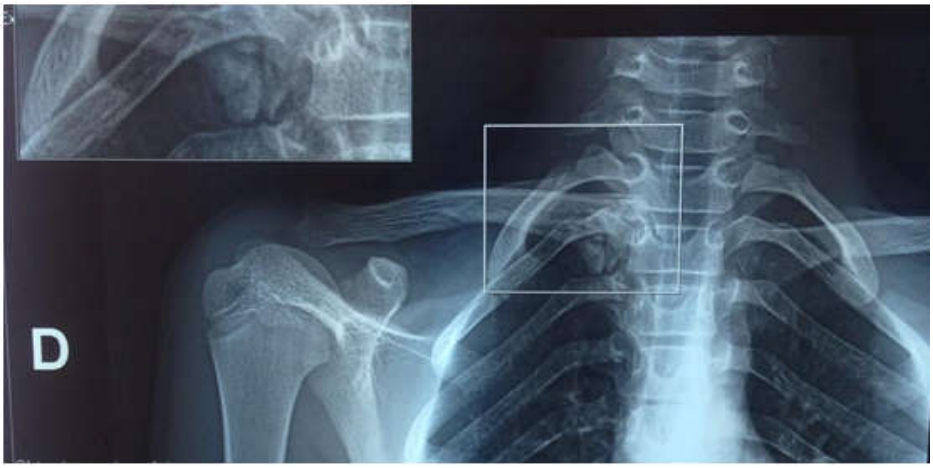


**Figure 1. Lytic lesion at the right clavicle internal edge**

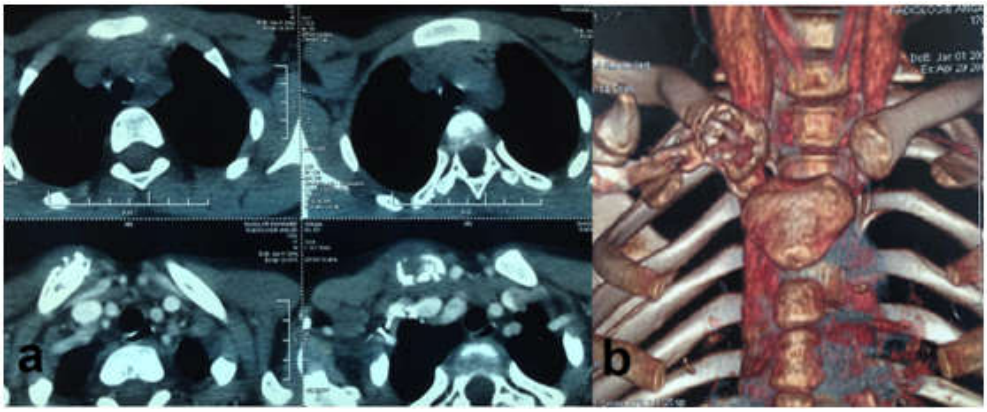


**Figure 2. a. CT objectifying the absence of associated pulmonary involvement, b. 3D reconstruction objectifying the isolated clavicular lytic lesion**

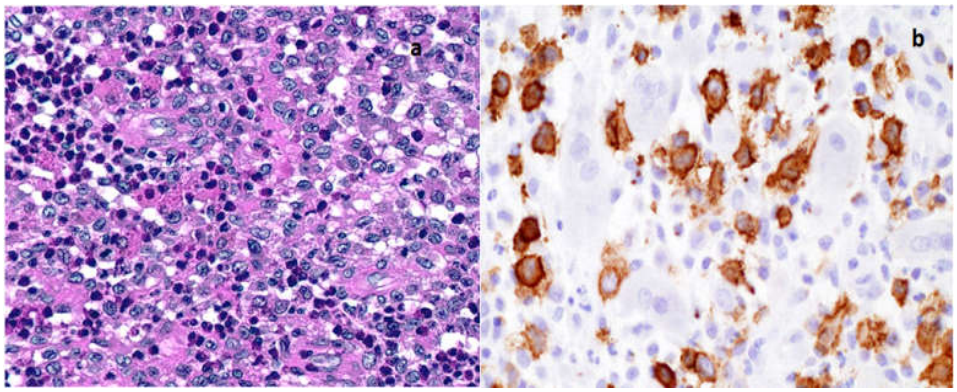


**Figure 3. a. Langerhansian histiocytosis histological aspect on biopsy, b. immunohistochemical study confirming the positivity of the anti-CD1a antibody.**

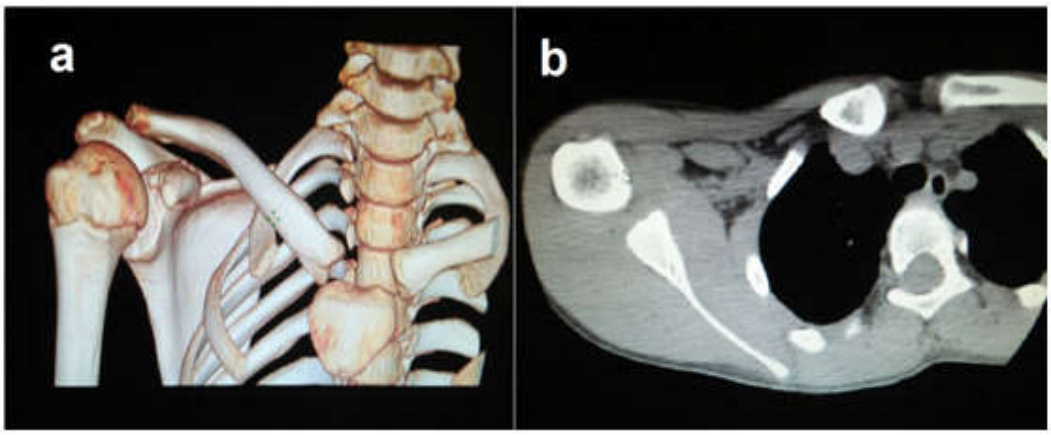


**Figure 4: control by CT objectifying the spontaneous regression of the lesion.**

It can be isolated, or observed in the context of systemic forms. Clinically, the main manifestation is inflammatory pain and responds very well to nonsteroidal anti-inflammatory drugs (Bernstrand, 1996). Sometimes the lesion can spread to the adjacent soft tissue and then manifest as a palpable mass. All the bones can be affected, the most frequently affected being those of the axial skeleton, first of all the skull and the dorsal or lumbar vertebrae, but also the ribs, the pelvis, the diaphyses or metaphyses of the femurs, the mandible (Baptista, 2012). Standard radiographs show one or more lytic puncture lesions, without peri-lesional condensation, ranging from a few millimeters to a few centimeters in diameter and extending from the medulla to the cortical. The scanner centered on the affected bone region makes it possible to measure the size of the lesion, to assess its fracture risk by showing cortical lysis and to specify the extension to the soft tissues, which are then enhanced by the injection of contrast product (Girschikofsky, 2013). However, the only systematic means of assessing bone damage recommended is a complete skeleton x-ray (Haupt, 2013).

The diagnosis is based on the anatomopathological study. The bone damage risk is variable. Unifocal forms have a good prognosis, spontaneously or after local treatment. For multifocal forms, remission is often obtained, but relapses occur with a variable rate from 12% to 27% depending on the studies (Baptista *et al*., 2014). In the event of a non-threatening local focal bone form, local treatments are recommended. The curettage biopsy, which is done for diagnostic purposes, can itself initiate a process leading to healing. Complete excision is generally not recommended, as it can be decaying and increase the healing time. In all cases, bone reconstruction is often slow and takes several months or even a year (The French Langerhans', 1996).

**Conclusion**

Langerhansian Histiocytosis is a rare and polymorphic disease, which sometimes makes its diagnosis difficult. It evolves by pushes. The disease sometimes leaves significant functional consequences, with big physical, psychological and social repercussions.

## REFERENCES

Baptista AM, Ferrari A, Camargo F, Pires de Camargo O, Odone Filho V, Enzo Cassone A. 2012. Does adjunctive chemotherapy reduce remission rates compared to cortisone alone in unifocal or multifocal histiocytosis of bone? Clin Orthop Relat Res 2012;470:663–9.

Bernstrand C, Bjo¨rk O, Ahstro¨m L, Henter JI. 1996. Intralesional steroids in Langerhans cell histiocytosis of bone. Acta Paediatr. 1996;85:502–504.

Girschikofsky M, Arico M, Castillo D, *et al*., 2013. Management of adult patients with Langerhans cell histiocytosis: recommendations from an expert panel on behalf of EuroHistio-Net. *Orphanet J Rare Dis.,* 8:72.

Haupt R, Minkov M, Astigarraga I, *et al*., 2013. Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. *Pediatr Blood Cancer.,* 60:175–84.

Jean Donadieu, Sébastien Héritier, 2017. Histiocytose langerhansienne de l'enfant Presse Med. 46: 85–95

Mathilde de Menthon, 2017. Histiocytose à cellules de Langerhans de l'adulte Presse Med. 46: 55–69.

Saliba I, Sidani K, El Fata F, Arcand P, Quintal MC, Abela A. 2008. Langerhans' cell histiocytosis of the temporal bone in children. *Int J Pediatr Otorhinolaryngol*.72:775–786.

The French Langerhans' Cell Histiocytosis Study Group. 1996. A multicentre retrospective survey of Langerhans' cell histiocytosis: 348 cases observed between 1983 and 1993. Arch Dis Child.

*******