ABSTRACT

Introduction: Langerhans cell histiocytosis is a systemic proliferative disease. It is a rare disease that can affect all tissues. Evolution can be spontaneously favorable. Multi-organ involvement may be life-threatening. We report the case of an toddler with bone lesion and issues.

Case Report: It was a 33-month-old infant with right shoulder pain. Clinically, she had a swelling in the front side of the right shoulder with a limited abduction. X-ray of the right shoulder showed osteolysis with an irregular boundary of the right humeral head. The blood work was normal. In view of the painful swelling of the right shoulder, a biopsy was performed. Histological examination confirmed the diagnosis of Langerhans histiocytosis. A conservative treatment was decided after a multidisciplinary consultation meeting. After six months, spontaneous improvement was noted.

Conclusion: Langerhans cell histiocytosis is a proliferative pathology that can affect one or more organs. The clinical presentation is polymorphic according to the affected organ. An extension assessment is fundamental in the management in order to determine the treatment.
Keywords: Bone lesion; bone pain; child; Langerhans cell histiocytosis.

1. INTRODUCTION

Langerhans cell histiocytosis, is a systemic pathology resulting from clonal proliferation of dendritic cells with the immunological characteristics of Langerhans cells [1]. It is a rare disease most often affecting children, with an estimated incidence of 5 cases per million children, or 55 new cases per year [2]. It can affect any organ but bone damage is the most common. The clinical presentation is polymorphic and may involve an isolated organ damage or a multi-visceral lesion. We report the case of a 33-month-old girl with Langerhans histiocytosis with bone involvement.

2. PRESENTATION OF CASE

A 33-month-old girl from Fianarantsoa was seen in the pediatric oncology department of the Joseph Ravoahangy Andrianavalona University Hospital in May 2018 for a swelling of her right shoulder. Her disease was evolving 6 months before by an asymmetry of the shoulders whose radiography of the shoulders showed a subluxation of the right shoulder. The child benefited from 5 sessions of motor physiotherapy and his articulation returned to normal. The shoulder then became painful waking up at night in an apyretic context treated by a paracetamol type 1 analgesic every six hours.

On physical examination, she was in good general condition with good staturo-ponderal development. It showed a swelling of 2.5 cm in diameter, painlessly limited to the anterior aspect of the right shoulder with exposure of the underlying muscle. The abduction of the corresponding shoulder was limited to 45°. There was no tumor syndrome, no rash. Stomatological, ophthalmological and ENT examinations were normal. There were no signs of diabetes insipidus or pituitary involvement.

On paraclinical examinations, the blood count and sedimentation rate were normal and there was no evidence of macrophageactivation syndrom. CRP, creatinine and transaminases were normal. No abnormalities were detectable on abdominal ultrasound. Radiography of the right shoulder showed irregularly bounded osteolysis of the right humeral head (Fig. 1). A biopsy of the lesion was performed and confirmed the histological diagnosis of Langerhans histiocytosis. A budding appeared at the level of the surgical scar.

Fig. 1. Osteolysis of the right humeral head

In the light of these clinical, radiological and histological arguments, we made the diagnosis of Langerhans histiocytosis with isolated right scapular bone localization classified as group A according to Oberman’s classification [3]. We offered the parents vinblastine chemotherapy but they refused the treatment. A conservative treatment was decided after a multidisciplinary consultation meeting. Regular monitoring of clinical status was recommended. Six months later, spontaneous improvement was noted and the lesion disappeared. The little girl manages to make a complete abduction of her right shoulder with a reappearance of the humeral ossification nucleus after an X-ray control.

3. DISCUSSION

Langerhans histiocytosis is a rare disease that can affect patients of any age, but preferably affects children. Its incidence is estimated at 4.6 cases per million children. The incidence is higher in infants with a peak in frequency between 1 and 4 years [1]. On the other hand, there is male dominance with a sex ratio of 1.2 [1].

The pathophysiology of this disease is still poorly understood. It is a reactive disease of dendritic cells with no known etiology. In addition, smoking is a risk factor for lung localization [4]. Our case...
had no particular history. Langerrhans histiocytosis is a systemic disease that can affect one or more organs. The clinical presentation is thus polymorphic. Bone damage is found in 80% of cases with localization at the level of flat bones, long bones or vertebrae [5].

Circumstances in which bone damage is discovered are dominated by increased pain at night, swelling, or pathologic fracture [5]. According to some authors, swelling in relation to the lesion is the most common mode of disclosure in children [6]. The telltale signs in our observation were bone pain associated with functional impotence. When locating in the long bone, the diaphysis is the most frequently affected area. Metaphyseal damage is rare and may manifest as joint damage [6].

In a typical clinical-radiological picture, diagnostic confirmation is based on histological examination of a biopsy specimen [7]. Histology is characterized by an often polymorphic infiltrate including, in variable proportions, histiocytes, eosinophilic polymuclear cells, lymphocytes and plasma cells and multinucleated cells. According to the diagnostic criterion of histiocytic society [8], the diagnosis is made after histological examination by light microscopy associated with at least one of the following two criteria: The presence of either Birbeck granules under electron microscopy or the presence of pathological cell labelling with CD 1a antigen on immunohistochemistry. Our diagnosis was only a presumptive diagnosis because immunohistochemistry was not made by the difficulty of accessibility. The main differential diagnoses for the presence of osteolytic lesions in children are Ewing’s sarcoma, bone infection, dermoid cyst or bone lymphoma. In the case of bone damage, histological confirmation is definitive. In addition, bone involvement may be uni- or multifocal. Thus, the search for other locations by means of an X-ray of the entire skeleton is part of the extension assessment at the time of diagnosis in order to decide on therapeutic indications. In our case, it’s focal bone damage. According to Oberman’s classification, group A includes patients in whom the lesion was confined to a single bone [3].

For treatment, simple monitoring or surgical curettage is recommended for localized bone forms [2]. Surgical treatment by curettage or immobilization measures can reduce the risk of fracture. Multifocal forms or forms associated with one or more visceral injuries require chemotherapy with vinblastine and 6 mercaptopurine.

The natural evolution of Langerrhansian histiocytosis is heterogeneous. This evolution can go from spontaneous self-regression of the lesions to a multi-visceral lesion that can be fatal. In addition, certain lesions can evolve into permanent sequelae such as diabetes insipidus, hormonal deficits, deafness or respiratory insufficiency, neurodegeneratives changes, secondary malignancy [9]. According to some authors, an age of less than 2 years [10] and damage to an organ at risk, such as the liver, marrow or lung, are poor prognostic factors [11,12]. In our case, the evolution was favourable without chemotherapy despite the destructive nature of the lesion. The severity of the lesion does not predict the prognosis.

4. CONCLUSION

Langerhans histiocytosis is a systemic proliferative non-malignant disease. The clinical presentation is polymorphic. Bone damage is the most common. The evolution is heterogeneous. A spontaneous improvement can be noticed. The management passes through an extension assessment and multidisciplinary meeting to determine the treatment. Prolonged surveillance is fundamental in this non-malignant tumour pathology.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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