Adult Langerhans' cell histiocytosis with multisystem bone and lung involvement: About a case

L'Histiocytose Langerhansienne de l'adulte avec atteinte multi systémique osseuse et pulmonaire : A propos d'un cas

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Abstract

Introduction: Langerhans cell histiocytosis (LCH) is a rare systemic disease characterized by the abnormal overproduction of histiocytes that tend to infiltrate single or multiple organ systems leading to significant tissue damage. Although LCH can involve various organs including bone, skin, and lymph nodes, multisystem involvement of LCH is rare in adults

Case presentation: We report the case of a 31-year-old man with LCH involving his lungs and bone. The initial radio-clinical presentation was rather in favor of pulmonary tuberculosis. Through this observation we draw attention to this rare pathology and we discuss the diagnostic elements and the therapeutic management of this pathology.

Conclusion: Although it is occasionally difficult to discriminate LCH from other disorders, systemic evaluation might be helpful for differential diagnosis. As LCH isn't infiltrating malignant cells, strong chemotherapy regimen is not recommended in order to avoid severe toxic and side effects.

Key words : Chest Langerhans cell Histiocytosis, costal involvement, Adult, Chest CT scan, multisystemic form

Résumé

Introduction : L'Histiocytose Langerhansienne est une maladie systémique rare caractérisée par une production importante et anormale d'histiocytes qui infiltrent un ou plusieurs organes entrainant des lésions tissulaires importantes. Bien que cette maladie peut impliquer divers organes, y compris l'os, la peau et les ganglions lymphatiques. L'atteinte multi systémique de L'Histiocytose Langerhansienne est rare chez l'adulte.

Présentation du cas: Nous rapportons le cas d'un homme de 31 ans atteint d'Histiocytose Langerhansienne touchant l'os et les poumons. La présentation radio clinique initiale était plutôt en faveur d'une tuberculose pulmonaire. A travers cette observation nous attirons l'attention sur cette pathologie rare et nous discuterons les critères diagnostiques et les principes de prise en charge thérapeutique.

Conclusion: Bien qu'il est parfois difficile de distinguer l'Histiocytose Langerhansienne d'autres maladies, un bilan de dissémination pourrait être utile pour le diagnostic différentiel. Puisque cette cette maladie n'est pas un néoplasie, une chimiothérapie n'est pas recommandée afin d'éviter des effets indésirables graves.

Mots clés : Histiocytose Langerhansienne pulmonaire, localisation costale, Adulte, Tomodensitométrie thoracique, atteinte multisystémique.

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INTRODUCTION

Langerhans Cell Histiocytosis (LCH) is a rare disease characterized by the infiltration of one or more organs by Langerhans cell type dendritic cells, most often organized into granulomas. The clinical picture of LCH is very variable. Bones, skin, pituitary gland, lungs, central nervous system and lymphoid organs are the main organs involved. The evolution of LCH ranges from fulminant multisystemic disease to spontaneous resolution. Adult LCH has certain specific characteristics and poses distinct therapeutic challenges; knowing that data on these patients are limited (1). The bone is the most common site of LCH involvement, and lung, skin, lymphnode, central nervous system (CNS), and liver can also be involved (2).

Through the present case, authors draw attention for an atypical presentation of LCH occurring in a patient that showed the involvement of multiple organs including bone and lung in a with complete resolution of symptoms under analgesic treatment.

Reported Case

A 31-year-old man, with a family history of pulmonary tuberculosis, smoker 15 pack-years, presented to our consultation of pneumology with right latero-thoracic pain for three weeks associated with productive cough, night sweats and weightloss. Findings of physical examination were unremarkable. Electrocardiogram was normal. The hematology laboratory tests were also without abnormalities. The liver and renal functions were normal. The chest-X-Ray was also normal. Chest computed tomography (CT) revealed multiple bilateral excavated nodules with associated alveolar involvement made of ground glass nodules and parenchymal condensation of the right lower lobe. Bone window showed costal involvement of the middle arch of the 3rd left side with heterogeneous osteolytic lesion with invasion of the soft parts (Figures 1 and 2). The initial clinical presentation was in favor of pulmonary tuberculosis or pulmonary metastasis associated with bone metastasis. Pulmonary tuberculosis was suspected but overturned after the sputum mear results for Koch's bacillus was negative. Bronchial fibroscopy was refused by the patient. For anatomopathological confirmation, we accessed to the bone lesion since it is more accessible to biopsy. Needle biopsy of the rib lesion showed an abundant cellular infiltrate. This infiltrate contains sheets of oval mononuclear cells characterized by unique reniform or cleaved nuclei with pale cytoplasm and many eosinophils, plasma cells and lymphocytes are present with positive and diffuse staining with Cd1a (Figure 3).

Diagnosis of Langerhans cell histiocytosis (LCH) was retained.As part of the disease assessment, we completed by radiological explorations. MRI of brain and pituitary gland was normal. Bone scintigraphy showed a focus of hyper fixation in the sternum, the rest of the skeleton was of normal fixation.A level 2 analgesic has been prescribed .At 3, 6 and 12 months of follow-up, there was an improvement of pain with spontaneous resolution of the other symptoms.

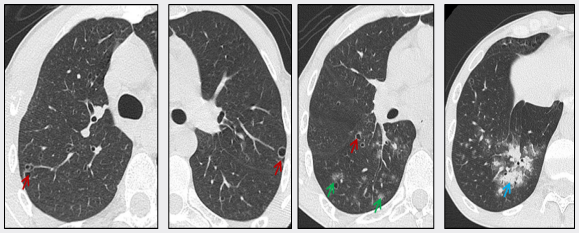

Figure 1. Thoracic CT, axial sections in parenchymal window**,** Multiple bilateral excavated nodules **(à)** with associated alveolar involvement made of ground glass nodules **(à)** and parenchymal condensation of the right lower lobe**(à)**

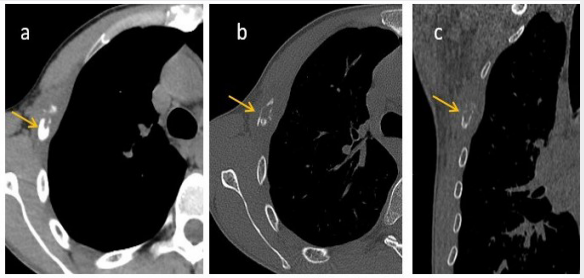

Figure 2. Thoracic CT without injection of contrast product **, a)** Axial mediastinal window, **b)** Axial bone window**, c)** Coronal reconstruction of bone window**,** Costal involvement **(à)** of the middle arch of the 3rd left side: heterogeneous osteolytic lesion with invasion of the soft parts

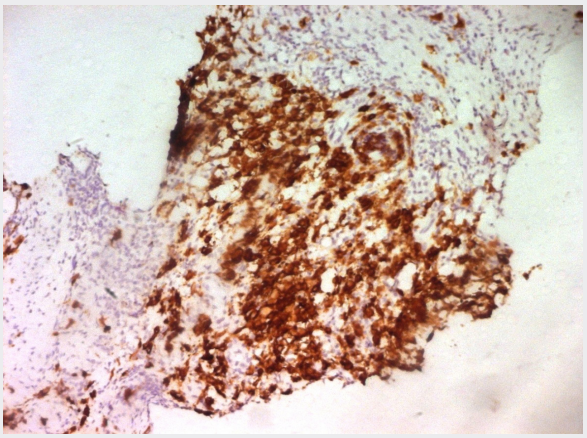

Figure 3. Positive and diffuse staining with Cd1a (IHCx200)

DISCUSSION

Langerhans' Cell Histiocytosis (LCH) is a rare disorder of unknown etiology, caused by clonal proliferation of altered Langerhans Cells (LC). LCH may affect any organ. Adult LCH has some specific features and poses distinct therapeutic challenges, knowing that data on these patients are limited (1). Manifestations of LCH are totally

diverse according to the sites and extents of the lesions. All lead to its high rate of misdiagnosis and missed diagnosis. The present case presented with symptoms suggestive of pulmonary tuberculosis associated to recurrent right latero-thoracic pain.The most affected organs of LCH are bone (80% of patients), skin (33%), and pituitary (25%) (3-5).

The clinical manifestation varies widely due to the differences among age of onset, the proliferation rate of Langerhans cells and the involved tissues and organs (6). LCH is also subdivided into single and multi-system types according to the number of involved organs (7). In our case, although only one enhancing lesion in the 3rd right rib was pathologically confirmed, typical imaging findings of LCH were observed in lung. LCH in our case was considered to be the multisystemic form.

Bone is the most common site of involvement in LCH. Bone involvement presents as a single or multiple lesions within the same bone and sits mainly in the bones of the skull, mandible, pelvis, ribs, clavicles and vertebrae. Clinically, these lesions may be asymptomatic or responsible for localized pain with or without swelling of adjacent soft tissues (8).

In patients with localized, unifocal bone disease, LCH may resolve spontaneously or even surprisingly, by a simple biopsy. In the case of a single bone lesion without symptoms, a wait and see approach or diagnostic curettage is the standard method of care (1). In our case, the patient reported a decrease in pain spontaneously after the biopsy.

Lung involvement is not a common form of LCH and is associated with tobacco smoke which is considered to be a main factor inducing pulmonary form of LCH, but the mechanisms of the process are unclear (9).

On a high-resolution chest tomography (CT), Langerhans cell histiocytosis of the lungs presents with a nodular and cystic pattern (10). In our case, multiple bilateral excavated nodules were predominant, therefore, our patient may have been in the later stage of pulmonary LCH. Histological examination found that nucleus was folded within a groove, making it looking like coffee beans. Eosinophils and other inflammatory cells was sporadic round in shape. The golden standard of diagnose is to find Birbeck particle by electron microscopy. The main immunohistochemical manifestation, as presented in our case, is S-100 protein and CD1a (+) (6).

Treatment strategies are currently based on extent and location of disease. Patients with more limited disease, such as single bone lesions or skin-only disease may not need systemic therapy. Single skin lesions may be surgically resected, and single bone lesions can be treated with curettage and local corticosteroid injections (5). Systemic chemotherapy is recommended in multi systemic LCH. Overall prognosis in LCH seems to be favorable. The prognosis of LCH is closely related to the age of onset, number of involved organs and degree of functional lesion. The prognosis of single-organ involvement is better than multiplied-organ involvement, and the latter has higher case-fatality rate (6).

CONCLUSION

Although it is occasionally difficult to discriminate LCH from other disorders, systemic evaluation might be helpful for differential diagnosis. Our case draws its originality from its clinical and radiological presentation and its histological confirmation.

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