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Case Report

Rare yet Critical Manifestations: A Case of Langerhans Cell Histiocytosis in a Child Presenting with Belt-Like Dorsalgia: Diagnostic and Therapeutic Approach

L. Atroune[1*], A. Hachid[1], Y. Djouadi[1], E. Kerboua[2], Z. Benlahrech[3], M. Djaafer[1]
[1]Department of Neurosurgery, Mustapha Bacha University Hospital, Algiers
[2]Department of Medical Oncology, Pierre and Marie Curie Center, Algiers
[3]Department of Medicine, University of Laghouat, Algeria

***Corresponding Author:** L. Atroune
Department of Neurosurgery, Mustapha Bacha University Hospital, Algiers

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Abstract: Langerhans cell histiocytosis (LCH) is a rare disease characterized by abnormal proliferation of Langerhans cells. We present the case of a 9-year-old girl complaining of recent belt-like dorsalgia, whose imaging revealed dorso-lumbar scoliosis and a tumor mass at the posterior arch of T11 with posterior epidural infiltration. Surgical decompression and tumor resection were performed, histopathologically confirming the diagnosis of LCH. This abstract highlight the diagnostic and therapeutic challenges of LCH, emphasizing the importance of a multidisciplinary approach for effective management of this rare disease. Additionally, it underscores the relevance of early intervention and meticulous follow-up to ensure optimal outcomes for patients with LCH.

Keywords: Langerhans cell histiocytosis, LCH, child, dorsalgia, dorso-lumbar scoliosis, tumor mass, surgical decompression, tumor resection, diagnosis, treatment.

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare and complex disorder involving abnormal proliferation of Langerhans cells. We report here the case of a 9-year-old girl presenting recent belt-like dorsalgia without other apparent neurological manifestations. Initial investigations revealed dorso-lumbar scoliosis on radiographs of the dorsal and lumbar spine. Subsequent magnetic resonance imaging (MRI) revealed an expansive lytic tumor process at the posterior arch of T11, with posterior epidural infiltration compressing the spinal cord and infiltration of the para-vertebral soft tissues. Evolving lesions at the TH10-TH11-TH12 vertebrae were confirmed by bone scintigraphy. Treatment involved surgical decompression and tumor resection, despite extensive infiltration of the para-vertebral muscular soft tissues and posterior arches of T11 and TH12 by the tumor. Histopathological examination and immunohistochemical profiling confirmed the diagnosis of Langerhans cell histiocytosis. This clinical presentation underscores the importance of thorough evaluation and appropriate management of LCH, a disease that often remains a diagnostic and therapeutic challenge.

CASE REPORT

This is the case of a 9-year-old girl presenting recent belt-like dorsalgia without other neurological manifestations. Radiographs of the dorsal and lumbar spine revealed dorso-lumbar scoliosis. Dorsal MRI (fig 01) showed an expansive lytic tumor process at the posterior arch of T11 with posterior epidural infiltration compressing the spinal cord, as well as infiltration of the para-vertebral soft tissues. Bone scintigraphy confirmed evolving lesions at the TH10-TH11-TH12 vertebrae. Treatment involved surgical decompression and tumor resection, despite extensive infiltration of the para-vertebral muscular soft tissues and posterior arches of

T11 and TH12 by the tumor. Histopathological examination and immunohistochemical profiling confirmed the diagnosis of Langerhans cell histiocytosis, with subsequent regression of dorsalgia after treatment.

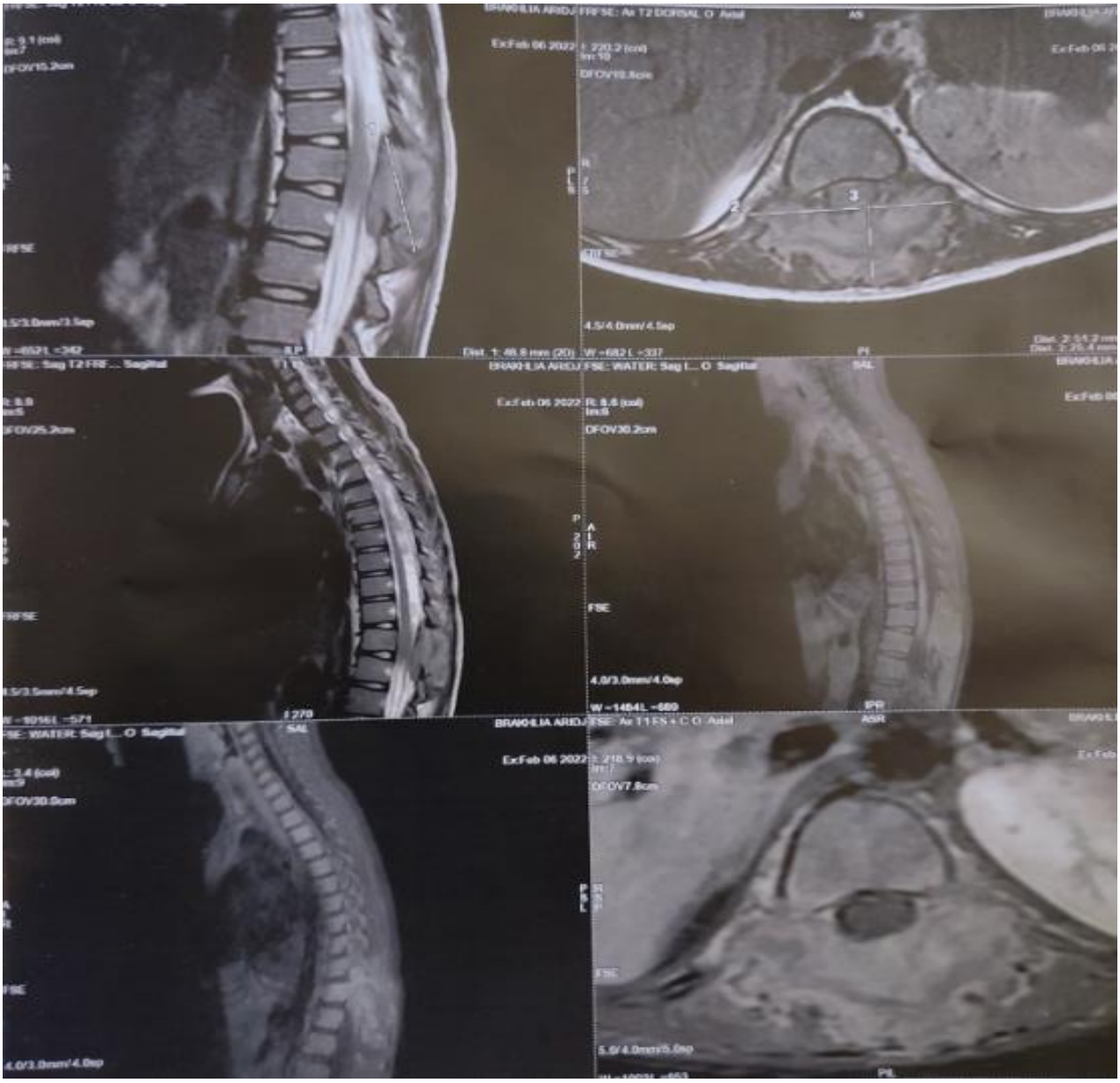

Figure 1: Dorsal MRI in axial and sagittal planes, in T1 and T2 sequences, showing spinal cord compression at the level of TH11

DISCUSSION

Langerhans cell histiocytosis (LCH) represents a rare and complex disease, the understanding of which has been progressively refined since its first description by Langerhans in 1868. Formerly known as histiocytosis X, LCH encompassed three distinct clinical subtypes: eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease. Today, the term Langerhans cell histiocytosis is preferred, reflecting a better understanding of its histiogenetic origin [1].

Primarily observed in children and adolescents, LCH can also affect adults, albeit less frequently. Males seem to be slightly more affected than females, with an incidence ratio of 2.5 to 1 in children. The incidence in adults is considerably lower, with only one to two cases per million people per year. LCH presents with varied manifestations and can affect different systems, with a slightly higher prevalence in certain populations [2].

Incidence rates also vary by age, with a peak between 1 and 5 years in children and a broader distribution in adults, where multisystem involvement is more common than involvement of a single system. In the specific case of adults, vertebral lesions are less frequent than in children, most commonly manifesting in the thoracic spine, although the lumbar and cervical spine may also be affected. LCH represents a diagnostic

and therapeutic challenge, requiring an individualized approach based on the patient's age, the extent of the disease, and its specific clinical manifestations [7].

Langerhans cell histiocytosis (LCH) is a rare and complex disease with an etiology that remains largely poorly understood, with discrepancies in the medical literature. It encompasses three main entities classified according to severity and distinct clinical manifestations: Letterer-Siwe disease: This is the most severe form of LCH, characterized by multi-organ involvement affecting several organs. Patients often present with severe systemic symptoms such as fever, weight loss, rash, and vital organ dysfunction. Hand-Schüller-Christian disease: This form of LCH is characterized by bone lesions, usually accompanied by endocrine abnormalities such as diabetes insipidus [4].

Affected children may also present with cranial bone defects and exophthalmos. Eosinophilic granuloma (EG): Unlike the other two forms, EG typically manifests as isolated bone lesions without systemic involvement. However, these lesions can be severe and lead to functional complications. The precise etiology of LCH remains uncertain, but several hypotheses have been proposed. Environmental factors, such as viral infections (notably human herpesvirus 6) and bacteria, are believed to play a role in triggering it. Abnormalities of the immune system, as well as dysfunctions in intercellular communication between T cells and macrophages, may also contribute to the development of the disease. Cytokine-mediated processes involving factors such as tumor necrosis factor, IL-11, and leukemia inhibitory factor are also suspected. LCH can affect virtually any organ in the body, but it tends to manifest in tissues where Langerhans cells or their precursors are normally present [6].

The most frequently affected organs include bones, skin, the hypothalamo-pituitary axis, and the lungs. In bone lesions, they can be single or multiple, with a predominance observed in the dorsal vertebrae. Although spinal involvement is rare, it can occur and lead to neurological complications, although the development of spinal cord compression is unusual. Recently, the Writing Group of the Histiocyte Society proposed disease stratification based on the number and type of organs involved. This classification distinguishes single-system disease, where only one organ is affected, from multisystem disease, where multiple organs are involved. This approach allows for a better understanding of clinical variability and management of LCH [4].

Langerhans cell histiocytosis (LCH) often manifests with bone lesions, which may be asymptomatic in many cases. However, bone pain and the presence of a soft tissue mass may occur in some patients. The skull is the most commonly affected site, followed by long bones, flat bones, and vertebrae. Neurological symptoms are more common in adult patients, but pain and restricted movement can also occur in both adults and children. Additionally, in cases of cervical LCH, muscle symptoms such as restricted mobility or torticollis are more common. This variability in clinical presentation underscores the importance of careful evaluation and an individualized approach in diagnosis and management of the disease [3].

The most common radiographic features in spinal LCH are lytic lesions of the vertebra, which can often lead to vertebral collapse. (alterations in signal intensities with enhancement in vertebral bodies). MRI revealed a continuous epidural lesion with compression of the spinal cord. MRI shows low to intermediate signal on T1WI, hyperintense signal on T2WI, and marked homogeneous enhancement after gadolinium administration - Epidural LCH is generally indistinguishable from other spinal lesions such as infection, lymphoma, and metastases on MRI. Laboratory and clinical findings often lead to inaccurate differential diagnosis. Ultimately, MRI is performed to exclude a soft tissue mass, which would suggest a more aggressive cause of vertebral collapse. In bone LCH, complete bone radiography is the main examination. It typically shows single or multiple lytic lesions with round or oval shapes and well-defined borders, sometimes associated with peripheral condensation [5].

Bone involvement may be accompanied by cortical blowing and periosteal reaction or endosteal scalping, responsible for the "budding" appearance on CT or MRI and suggesting a differential diagnosis with Ewing's sarcoma, tuberculoma, or osteomyelitis. Also shows a destructive vertebral lesion respecting the posterior wall and intervertebral discs, responsible for the characteristic appearance of flattened vertebra (plana) which is considered a typical radiological feature in children. CT and MRI are used for better analysis of cortical erosion and tissue involvement. In the presence of neurological symptoms, MRI is indicated to investigate spinal cord and epidural involvement [2].

T99 scintigraphy is not specific but sometimes complementary. The contribution of positron emission tomography is poorly defined.

Classic radiographic findings include vertebral collapse, maintenance of disc spaces, absence of extra-spinal spread, and absence of soft tissue mass [1].

The definitive diagnosis of Langerhans cell histiocytosis (LCH) is based on an immunochemical study demonstrating positive staining of cells with CD1a or CD 207, while staining for S-100 protein lacks specificity. Although Birbeck granules on electron microscopy are highly specific, their use is limited due to cost and time constraints. Histological and immunophenotypic examination of a lesion biopsy revealing a Langerhans cell granuloma is essential for

diagnosis, with variations in appearance depending on the stage of the disease and the affected tissue [4].

Under the light microscope, Langerhans cells are recognizable by their convoluted nucleus, blade, and slightly eosinophilic cytoplasm, containing few or no phagocytic particles. Infiltration by lymphocytes and eosinophilic granulocytes forming pseudoabscesses is characteristic. These cells, large in size and with a reniform eccentric nucleus, are often associated with other inflammatory cells such as eosinophilic polymorphonuclear cells, lymphocytes, and macrophagic cells in bone and lymph node locations. Eosinophilic granuloma (EG) is the mildest variant of Langerhans cell histiocytosis (LCH), first introduced by Lichtenstein and Jaffe in 1940 [3].

Initially classified under the generic term histiocytosis X, it is now recognized as LCH. EG is characterized by abnormal proliferation of Langerhans cells, originating from myeloid dendritic cells rather than the skin, and constitutes the most common form of LCH.

Differential diagnosis includes a variety of different malignant tumors, including Ewing's sarcoma, osteosarcoma, leukemia, and lymphoma [9].

Management of vertebral Langerhans cell histiocytosis (LCH) requires a meticulous approach, emphasizing the importance of tissue biopsy before any therapeutic decision is made. Although bone lesions may sometimes resolve spontaneously, local injection of methylprednisolone provides symptomatic relief. Surgery is usually reserved for cases with neurological symptoms or functional deficits. Therapeutic strategies vary by risk, ranging from observation to chemotherapy, including options such as radiation, steroids, and surgery, tailored to individual presentation. Although rare in the cervical spine, LCH should be considered in children presenting with a flattened vertebra, requiring careful evaluation before ruling out other diagnoses. Despite debates over its efficacy, chemotherapy is often advocated, given the variability in disease progression. Vertebroplasty may be considered for pain relief and spinal stabilization, while surgery aims to prevent deformities and improve neurological deficits. Therapeutic options in adults remain controversial, with recommendations taking into account clinical presentation and disease extent [6].

The prognosis primarily depends on patient, disease, and treatment-related factors. Patients with single-system disease have a better prognosis than those with multisystem disease [8].

CONCLUSION

The case presented highlights the diagnostic and therapeutic challenges associated with Langerhans cell histiocytosis (LCH), a rare and complex disease. Despite its rarity, LCH can lead to serious complications, including bone lesions with the risk of spinal cord compression. In this case, a multidisciplinary approach including medical imaging, surgery, and pathology was crucial to reach the appropriate diagnosis and treatment. Histopathological confirmation of LCH by immunohistochemistry was essential to guide therapeutic management. Regression of dorsalgia after treatment underscores the importance of early and adequate intervention in managing this disease. However, despite advances in understanding LCH, challenges remain, particularly regarding its etiology and optimal therapeutic strategies. Further studies are needed to improve the management of this complex pathology and to better understand its underlying mechanisms. In conclusion, this case highlights the importance of a holistic and individualized approach for each patient with LCH, with long-term monitoring for detecting relapses and late complications.

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