



(CASE REPORT)



LANGERHANS CELL NEUROHISTIOCYTOSIS: A CASE REPORT

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ABSTRACT

Langerhans cell histiocytosis is a rare multisystem disease. Involvement of the central nervous system (neuro-LCH) is observed in 10% of cases and is characterized by the abnormal proliferation of histiocytes within the central nervous system. It is a rare disease with an etiology and pathophysiology that remain largely unknown. Lesions primarily affect the hypothalamic-pituitary axis—often causing diabetes insipidus—though they may also involve the cerebellum or brainstem.

We report the case of a patient hospitalized in the neurosurgery department at the Rabat Specialized Hospital for the purpose of establishing a histological diagnosis and receiving therapeutic treatment.

Keywords: Langerhans cell, Neurohistiocytosis, Osteolytic skull lesions, Brain MRI, Button sequestrum.

1 INTRODUCTION

Langerhans histiocytosis (HL) is a rare disorder characterized by the clonal proliferation and accumulation of dendritic cells exhibiting the phenotype of Langerhans cells. Central nervous system (CNS) involvement, often described as neurohistiocytosis of Langerhans, represents one of the most serious and complex manifestations of this disease, although it can also affect various organs such as the bones, skin, lungs, and liver.

Approximately 5 to 10% of individuals with Langerhans histiocytosis experience neurological symptoms. The identification of somatic mutations in the MAP kinase pathway has profoundly transformed the pathophysiological understanding of the disease, shifting its status from a reactive immune disorder to a myeloid neoplasm.

Clinically, neurohistiocytosis frequently presents with diabetes insipidus, multifactorial endocrine deficiencies, and debilitating cerebellar and cognitive impairments. Diagnosis relies on a combination of clinical findings, magnetic resonance imaging (MRI), and histological or molecular confirmation.

1.1 Objective

To describe the clinical manifestations, neuroradiological features, treatment approaches, and clinical course of patients with central nervous system Langerhans cell histiocytosis.

2 CASE REPORT

We report the case of a healthy 11-year-old female patient with no significant medical history, who was being followed at the neurosurgery department of the Rabat Specialty Hospital for isolated, intermittent headaches occurring in the absence of fever, dermatological lesions, endocrine disorders, or other associated neurological symptoms. Brain MRI revealed two lytic bone lesions in the right cranial vault—one frontal (17x17 mm) and one parietal (27x10 mm)—disrupting both the inner and outer tables; the lesions showed T1-hypointensity and T2-hyperintensity, enhanced following gadolinium administration, and were associated with meningeal thickening overlying the lesions, without parenchymal extension. This clinical presentation initially suggested cerebral Langerhans cell histiocytosis. Curettage of the bone lesions and a surgical biopsy confirmed the diagnosis of Langerhans cell histiocytosis.

The patient continued regular follow-up at the neurosurgery clinic of the Rabat Specialty Hospital; given that she was paucisymptomatic and a follow-up brain CT scan three months post-biopsy showed regression of the bone abnormalities, resolution of dural enhancement, and no signs of active disease, a conservative management approach (monitoring) was adopted.

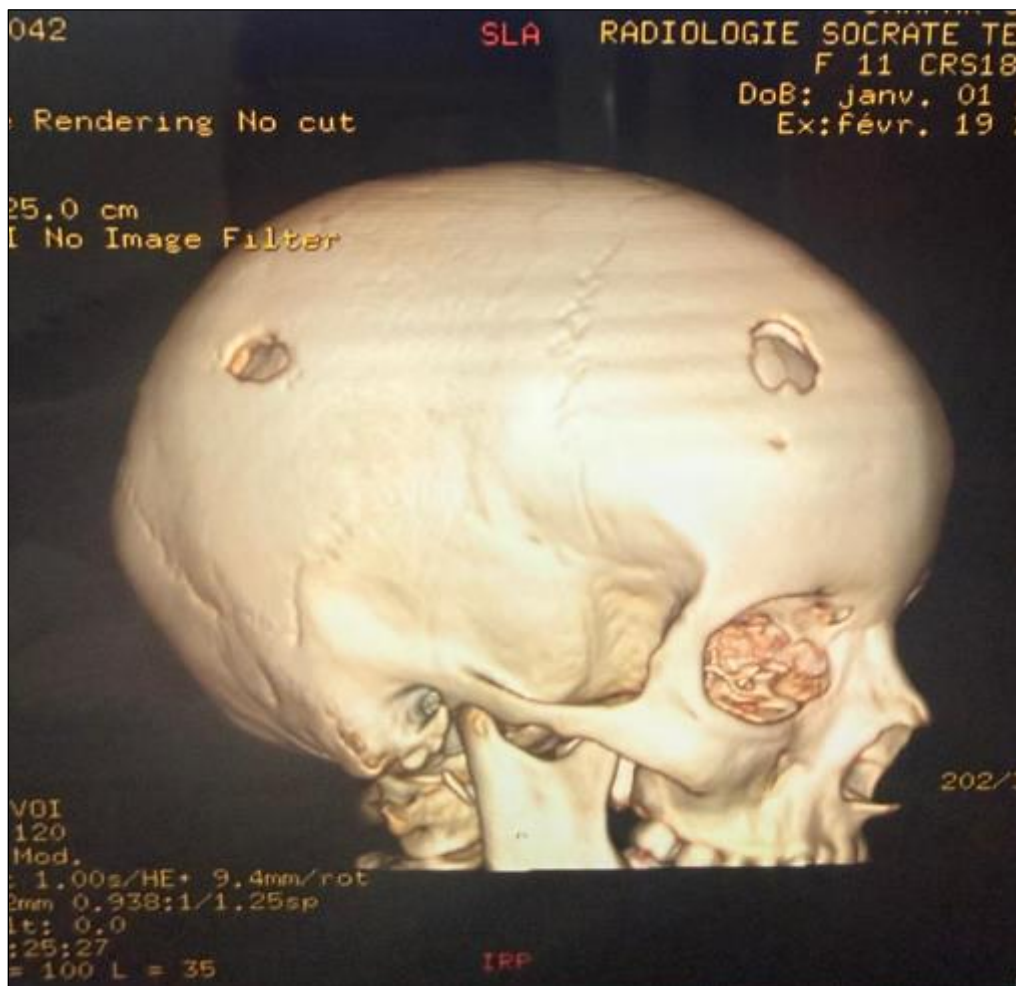


Figure 1: 3D bone-window CT image showing two lytic bone lesions in the cranial vault: a 17x17 mm lesion in the frontal bone and a 27x10 mm lesion in the parietal bone.

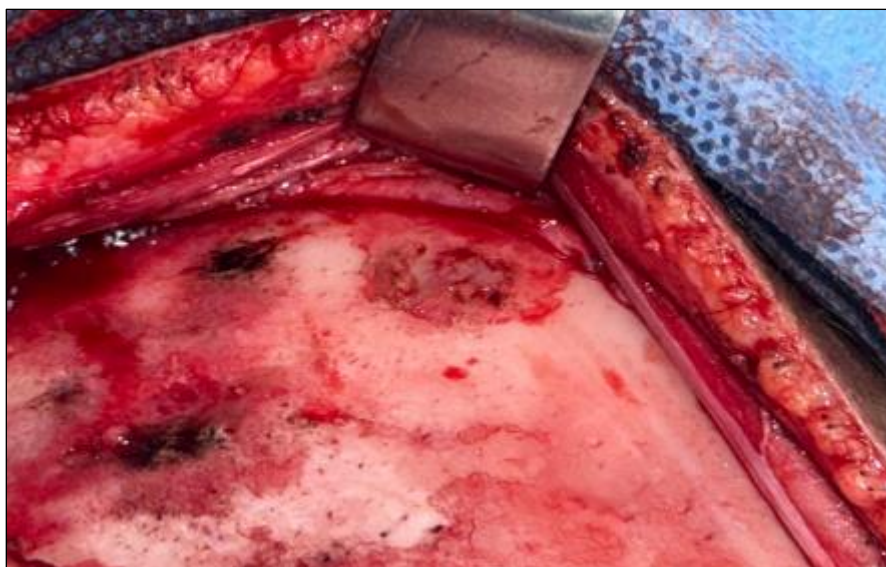


Figure 2: Intraoperative image showing the right frontal lesion, suggestive of cerebral Langerhans cell histiocytosis with overlying meningeal thickening

3 DISCUSSION

Neuro-Langerhans cell histiocytosis (LCH) is a rare manifestation of Langerhans cell histiocytosis, an inflammatory and neoplastic disease. Bone involvement is the most common clinical presentation, with the skull being the preferred site for lesions, particularly in children and young adults. However, extension into the central nervous system remains exceptional and presents specific diagnostic and therapeutic challenges [1].

In this context, osteolytic cranial lesions illustrate the well-known predilection of LCH for flat bones. They result from bone destruction secondary to infiltration by Langerhans cells, accompanied by a local inflammatory response. The most frequent sites involve the parietal, frontal, and temporal bones, with potential extension to the skull base or the orbit. Imaging typically reveals well-defined osteolytic lesions—sometimes featuring a central bone fragment, known as a "button sequestrum"—without significant periosteal reaction [2-3].

Neurological involvement may occur either through the direct extension of bone lesions into intracranial structures or as a distinct neurodegenerative process. Clinical manifestations are varied and include headaches, focal neurological deficits, seizures, cranial nerve involvement, cerebellar ataxia, or diabetes insipidus when the hypothalamic-pituitary axis is affected [4-5]. Brain MRI is the gold-standard examination for assessing neurological involvement; it allows for the precise characterization of the relationship between the lesions and the dura mater, brain parenchyma, and midline structures. A definitive diagnosis is established through histopathological examination, supplemented by immunohistochemistry [6-8].

Therapeutic management depends on the extent of the disease. A solitary bone lesion may be treated with surgical excision, sometimes combined with curettage. Multifocal or multisystemic forms require systemic chemotherapy, primarily based on vinblastine, in conjunction with corticosteroid therapy. The prognosis depends mainly on whether the disease is unifocal or multisystemic, as well as on whether there is involvement of "risk organs" [9-12].

4 CONCLUSION

Langerhans cell histiocytosis involving osteolytic skull lesions is a rare but characteristic presentation of the disease. Diagnosis relies on correlating clinical, radiological, histopathological, and immunohistochemical findings. A multidisciplinary approach involving neurosurgeons, radiologists, pathologists, oncologists, and endocrinologists is essential to optimize the prognosis and minimize long-term neurological sequelae.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent and verbal permission were obtained from the patient before the submission of this article. In addition, this article follows both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals.

REFERENCES

- [1] Fraitag, S. (2010) Histiocytoses Langerhansiennes. *Annales de Dermatologie et de Vénéréologie*, 137, 163-66. <https://doi.org/10.1016/j.annder.2009.12.009>
- [2] Veyssier-Belot, C. (2009) Histiocytoses Langerhansiennes. *EMC—Traité de médecine AKOS*, 3, 1-5. [https://doi.org/10.1016/S1634-6939\(09\)49777-1](https://doi.org/10.1016/S1634-6939(09)49777-1)
- [3] Ajja, A. and Ammor, R. (2015) Histiocytose Cérébrale. *Pan African Medical Journal*, 22, 345.
- [4] Tabarin, A., Corcuff, J.B., Dautheribes, M., Merlio, J.P., Cochet, C., Maire, J.P., Louail, C. and Roger, P. (1991) Histiocytosis X of the Hypothalamus. *Journal of Endocrinological Investigation*, 14, 139-145. <https://doi.org/10.1007/BF03350286>
- [5] Fournier, C. and Scherpereel, A. (2004) Histiocytose X Pulmonaire et Hypothalamo- Hypophysaire, diagnostiquée par biopsie cérébrale stéréotaxique [Endocrine and Pulmonary Histiocytosis: A Case Diagnosed through Stereotoxic Biopsy]. *Revue des Maladies Respiratoires*, 21, 141-145. [https://doi.org/10.1016/S0761-8425\(04\)71246-2](https://doi.org/10.1016/S0761-8425(04)71246-2).
- [6] Nguyen, K. and Tazi, A. (2006) Histiocytose langerhansienne de l'adulte. *Encyclopédie Orphanet*.
- [7] Donadieu, J. (2003) Histiocytose Langerhansienne. *Encyclopédie Orphanet*.
- [8] Donadieu, J., Guyot-Goubin, A., Clavel, J. and Thomas, C. (2008) Présentation clinique et épidémiologie de l'histiocytose langerhansienne chez l'enfant. *Archives de Pédiatrie*, 15, 520-522. [https://doi.org/10.1016/S0929-693X\(08\)71819-0](https://doi.org/10.1016/S0929-693X(08)71819-0)
- [9] Imashuku S, Okazaki N, Nakayama M, Fujita N, Fukuyama T, Koike K, et al. Treatment of neurodegenerative CNS disease in Langerhans cell histiocytosis with a combination of intravenous immunoglobulin and chemotherapy. *Pediatr Blood Cancer* 2008;50:308–11.
- [10] Imashuku S, Fujita N, Shioda Y, Noma H, Seto S, Minato T, et al. Follow-up of pediatric patients treated by IVIG for Langerhans cell histiocytosis (LCH)-related neurodegenerative CNS disease. *Int J Hematol* 2015;101: 191–7.
- [11] Chohan G, Barnett Y, Gibson J, Reddel SW, Barnett MH. Langerhans cell histiocytosis with refractory central nervous system involvement responsive to infliximab. *J Neurol Neurosurg Psychiatry* 2012;83:573–5.
- [12] Pardanani A, Phyliky RL, Li CY, Tefferi A. 2Chlorodeoxyadenosine therapy for disseminated Langerhans cell histiocytosis. *Mayo Clin Proc* 2003;78:301–6.