World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Pediatric temporal intracerebral Langerhans cell histiocytosis: A case report

Rayhane Hamdaoui, Djimrabeye Alngar, Nourou Dine Adeniran Bankole *, EL Mehdi Hakkou, Adyl Melhaoui, Yassaad Oudrhiri, Mahjouba Boutarbouch and Abdessamad El Ouahabi

Neurosurgery Department of Hôpital des Spécialités de Rabat.

World Journal of Advanced Research and Reviews, 2022, 14(02), 225–229

Publication history: Received on 02 April 2022; revised on 10 May 2022; accepted on 12 May 2022

Article DOI: https://doi.org/10.30574/wjarr.2022.14.2.0412

Abstract

Langerhans cell histiocytosis (LCH) is a rare condition of unknown etiology characterized by infiltration of one or more organs by Langerhans-type dendritic cells, most often organized into granulomas. This entity was initially described in young adult children. We report an infant case of 4 years old boy with clinical particularities such as tinnitus, imbalance of movement coordination, and itching. The MRI revealed a temporal mass lesion. Surgical management was performed by direct excision. The tumor excision was subtotal with simple postoperative outcomes, and good clinical improvement at discharge. Then we address him to the pediatric oncology hospital center. We will present also the state of current knowledge concerning infant LCH and recent data to guide therapeutic management. LCH has an extremely polymorphic clinical presentation. Its management in children must be multidisciplinary between neurooncologists and pediatric neurosurgeons.

Keywords: Langerhans Cell Histiocytosis; Intracranial; Temporal Localization; Pediatric

1. Introduction

Langerhans cell histiocytosis (LCH) is a rare disease of unknown etiology [1, 2]. It is due to an abnormal proliferation of Langerhans cells in various tissues and organs (bones, skin, lymph nodes, etc.) [3]. the first clinical descriptions of Langerhans cell histiocytosis (LCH) date back to the beginning of the last century [4]. The authors report previously a spheno-temporal LCH pediatric case [2].

The way histiocytic disorders are classified and the names derived from them have not always been the same over time. Histiocytic disorders are rare disorders characterized by the accumulation of cells derived from macrophages, dendritic cells, or monocytes in various tissues in both children and adults [5, 6, 7, 8].

There have been over 100 different subtypes since the first classification in 1987, which classified histiocytosis into 3 categories: Langerhans cells (LC), non-LC related, and malignant histiocytes [9, 10, 11]. These major developments in the clinical field have been complemented by productive research concerning the fundamental nature of the histiocyte and its disorders [12]. Various hypotheses have been explored, including whether HCL is a clonal disorder [13], cytokine-mediated Langerhans cell proliferation, or a reactive process following viral infection [14, 15].

It is now accepted that the disease is a clonal pathology, involving somatic mutations of the proteins involved in the MAP kinase pathway [16].

We report a case of infant temporal LCH with a brief review of the literature regarding then management.

* Corresponding author: Nourou Dine Adeniran Bankole
Neurosurgery Department of Hôpital des Spécialités de Rabat.

2. Case description

2.1. Clinical Presentation

A 4-year-old infant was hospitalized in December 2021 at our department for hearing itching associated with tinnitus for 4 weeks without improvement, the evolution will be marked one week by a hyper painful temporal swelling of rapid evolution. 2 days earlier before his admission in our department, he presented balance disorders associated with increasing difficulties in performing fine movements with the appearance of diffuse pruritus.

The clinical examination at admission found a conscious patient with Glasgow Coma Scale (GCS) at 15, with painful temporal swelling, tinnitus, imbalance of movement coordination, and itching without sensorimotor deficit, any associated pyramidal or extrapyramidal syndrome. He presented also a soft and painless right temporal mass.

2.2. Diagnosis Assessment

The usual biological examinations showed an increased number of platelet (524000/dl of blood), and all the other parameters are normal.

The MRI revealed a temporal fossa process that involves the temporal bone (Figure1).

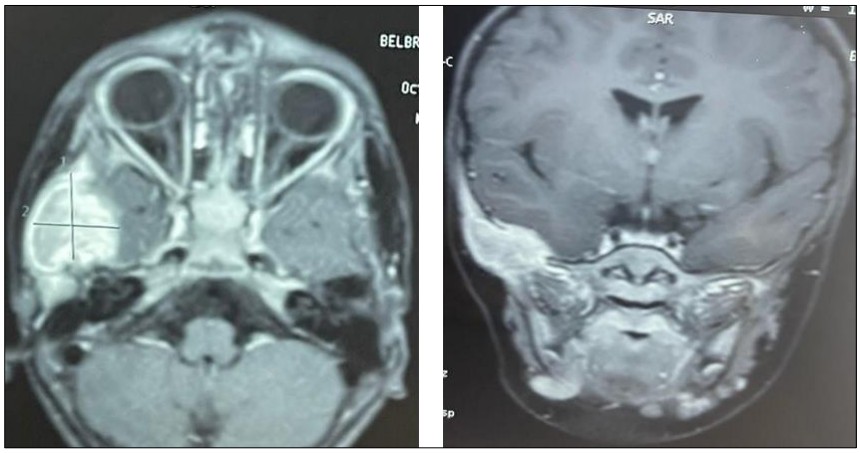

Figure 1 MRI T1 sequence with contrast in Axial view showing a temporal fossa mass, and the coronal view showing temporal bone involvement

2.2.1. Legends of Figures

Figure 1 MRI T1 sequence with contrast in the axial view shows a temporal fossa mass, and the coronal view shows temporal bone involvement

2.3. Management

The patient underwent craniotomy with the temporal approach for direct excision, and subtotal tumor resection was achieved. The histopathology confirmed a Langerhans cell histiocytosis. Then we addressed him at discharge under prednisolone 40 mg/day for two weeks to the neurooncology department for complementary chemotherapy management.

2.4. Outcomes and follow-up

The outcomes after surgery and at discharge were good, with no complications.

At 6 months of follow-up, the infant has good outcomes under chemotherapy with less pain, improvement of tinnitus, and imbalance of movement coordination.

3. Discussion

We report a pediatric case of central nervous system LCH with multidisciplinary management with a good outcome.

Langerhans cell histiocytosis is a disease in children and young adults. It is rare in adults [17, 18] and its etiology remains unknown (19, 20]. it is caused by the proliferation of a clone of Langerhans cells [21, 22].

Central Nervous System involvement in LCH is unusual but not uncommon. Magnetic resonance imaging (MRI) reveals neurodegenerative lesions in approximately 1% of patients with LCH [23]. Cerebral involvement in LCH may likely result from primary histiocyte proliferation and secondary atrophy, or from demyelination and gliosis of unknown origin [24].

Langerhans cells are known to be cutaneous dendritic histiocytes derived from bone marrow and immunoglobulin G and C3 receptors [24], they are not phagocytic and contain unique Birbeck granules [26]. Langerhans cells are the only extrathymic mononuclear cells to normally express thymocyte differentiation. A higher level of diagnostic confidence is warranted when light microscopic observations are supplemented by the presence of two of the following features: 1) positive staining for adenosine triphosphate; 2) S-100 protein; 3) α-D-mannosidase, or 4) characteristic finding of peanut lectin [27]. We confirmed histologically our case in this way."Definitive diagnosis" requires identification of Birbeck granules in lesion cells by electron microscopy or demonstration of T6 antigenic determinants on the surface of lesion cells [27].

The rule of thumb in the treatment of LCH is that the aggressiveness of the treatment should be proportional to the aggressiveness of the disease in each patient. Biopsy could also be achieved in a case where the excision cannot achieve safely [2].

In our current case here, we had acted aggressively in an emergency with a very favorable postoperative result. Surgery is the mainstay treatment when the lesion is accessible; there is seldom underlying dural penetration [2]. Reconstruction of the defect with autologous cranial bone can be easily performed. However, in our case the size of the defect was small, and no bone reconstruction was necessary. Chemotherapy Vinblastine 6 mg/m2/dose intravenous per week with corticosteroid therapies, is commonly used in pediatric cases and usually has good outcomes [2]. The role of radiotherapy is dubious and is generally reserved for cases of recurrent tumors [28]. The risk of toxicity is not negligible and must therefore be reserved in the event of recurrent disease [29].

Abbreviations

- MRI: Magnetic resonance Imaging
- LCH: Langerhans cell Histiocytosis
- CNS: Central Nervous System
- GCS: Glasgow Coma Scale

4. Conclusion

Intracranial LCH is still rare today, and research is still needed to optimize the diagnosis and treatment of the disease. Currently, the treatment is still based on a case-by-case basis, without any standard of treatment. Surgery is a goal standard when the process is accessible for excision. However, a multidisciplinary approach between neuro-oncologist and neurosurgeons should be helpful taking good decisions case by case.

Compliance with ethical standards

Disclosure of conflict of interest

The authors did not receive any funding for the preparation of this case report.

Statement of informed consent

Informed consent and verbal permission were obtained from the patient and her family 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] Satter EK, High WA. Langerhans cell histiocytosis: a review of the current recommendations of the Histiocyte Society. Pediatr Dermatol. 2008; 25: 291–5.

[2] Sayore, C. , Bankole, N. , Rifi, L. , Cherradi, N. , Melhaoui, A. and Elouahabi, A. (2020) Spheno-Temporal Langerhans Cell Histiocytosis: A Case Report. Open Journal of Modern Neurosurgery, 10, 260-266. doi: 10.4236/ojmn.2020.102028.

[3] Lallemant B, Fayoux P, Nelken B, Leroy X, Vaneecloo FM. Du diagnostic a` la prise en charge des localisations ORL de l'histiocytose langerhansienne chez l'enfant. Ann Otolaryngol Chir Cervicofac. 2003; 120: 30–9.

[4] Coppes-Zantinga A, Egeler RM. The Langerhans cell histiocytosis X files revealed. Brit J Hematol. 2002; 116: 3-9.

[5] E Caroli, L Ferrante. Intradural eosinophilic granuloma with intraparenchymal invasion: a new growth pattern, Zentralbl Neurochir. 2007; 68: 79–82.

[6] S Husain, HM Alkhalidi, E Raddaoui, A 38-year old woman with a dural based lesion, Brain Pathol. 2012; 22: 433–437.

[7] L Mihova, M Yaneva, M Sopadjieva, V Andreev, I Nejtcheva. Primary solitary eosinophilic granuloma of the brain and postoperative radiotherapy: a clinical case, J buon. 2007; 12: 125–128.

[8] Müller CS, Janssen E, Schmaltz R, Körner H, Vogt T, Pföhler C. Multisystemic Langerhans cell histiocytosis presenting as chronic scalp eczema: clinical management and current concepts. J Clin Oncol. 2011 Jun 20;29(18):e539-42. doi: 10.1200/JCO.2010.33.9127.

[9] S Noh, DS Kim, J Kim, SH Kim. Langerhans cell histiocytosis in endoscopic biopsy: marked pinching artifacts by endoscopy, Brain Tumor Pathol. 2011; 28: 285–289.

[10] Perren F, Fankhauser L, Thiévent B, Pache JC, Delavelle J, Rochat T, Landis T, Chizzolini C. Late adult onset of Langerhans cell histiocytosis mimicking glioblastoma multiforme. J Neurol Sci. 2011 Feb 15;301(1-2):96-9. doi: 10.1016/j.jns.2010.11.006

[11] KD Weaver, D Armao, JM Wiley, MG Ewend. Histiocytic lesion mimicking intrinsic brainstem neoplasm, Case report. J Neurosurg. 1999; 91: 1037–1040.

[12] Willman CL, Busque L, Griffith BB, et al. Langerhans'-cell histiocytosis (histiocytosis X): a clonal proliferative disease. N Engl J Med. 1994; 331: 154-60.

[13] Yu RC, Chu C, Buluwela L, Chu AC. Clonal proliferation of Langerhans cells in Langerhans cell histiocytosis. Lancet. 1994; 343: 767-8.

[14] McClaln K, Jin H, Gresik V, Favara B. Langerhans cell histiocytosis: lack of a viral etiology. Am J Hematol. 1994; 47: 16- 20.

[15] Leahy MA, Krejci SM, Friednash M, et al. Human herpesvirus 6 is present in lesions of Langerhans cell histiocytosis. J Invest Dermatol. 1993; 101: 642-5.

[16] Badalian-Very G, Vergilio JA, Degar BA, et al. Recurrent BRAF mutations in Langerhans cell histiocytosis. Blood. 2010; 116: 1919–23.

[17] Donadieu J, Thomas C, Emile JF, Teillac-Hamel D, Landman-Parker J, Aubier F, et al. Histiocytose langerhansienne mise au point. Med thérapeutique. 1996; 2: 441-51.

[18] The French Langherans Cell Histiocytosis Study Group. A multicentre retrospective retrospective survey of Langherans's cell histiocsis: 348 cases observed between 1983 and 1993. Arch Dis Child. 1996; 75: 17-24.

[19] Emile JF, Brousse N. Histiocytoses: données physiopathogéniques récentes. Rev Med Interne. 1996; 17: 891-2.

[20] Donadieu J, Thomas C, Emile JF, Teillac-Hamel D, Landman-Parker J, Aubier F, et al. Histiocytose langerhansienne mise au point. Med thérapeutique. 1996; 2: 441-51.

[21] Willman CL, Busque L, Griffith BB, et al. Langerhans'-cell histiocytosis (histiocytosis X)—A clonal proliferative disease. N Engl J Med. 1994; 331: 154–160.

[22] Egeler RM, D'Angio GJ. Langerhans cell histiocytosis. J Pediatr. 1995; 127: 1–11.

[23] Grois NG, Favara BE, Mostbeck GH, et al. Central nervous system disease in Langerhans cell histiocytosis. Hematol Oncol Clin North Am. 1998; 12: 287–305.

[24] Burn DJ, Watson JD, Roddie M, et al. Langerhans' cell histiocytosis and the nervous system. J Neurol. 1992; 239: 345–350.

[25] Barthez MA, Araujo E, Donadieu J. Langerhans cell histiocytosis and the central nervous system in childhood: Evolution and prognostic factors. Results of a collaborative study. J Child Neurol. 2000; 15: 150–156.

[26] Katz SI, Tamaki K, Sachs DH: Epidermal Langerhans cells are derived from cells originating in bone marrow. Nature. 1979; 282: 324–326.

[27] Schuler G, Steinman RM: Murine epidermal Langerhans cells mature into potent immunostimulatory dendritic cells in vitro. J Exp Med. 1985; 161: 526–546.

[28] Perren L, Fankhauser B, Thievent JC, Pache J, Delavelle T. Rochat, et al., Late adult-onset of Langerhans cell histiocytosis mimicking glioblastoma multiforme, J Neurol Sci. 30.

[29] Park SH, Park J, Hwang JH, et al. Eosinophilic granuloma of the skull: a retrospective analysis. Pediatr Neurosurg. 2007; 43: 97-101.