Spheno-Temporal Langerhans Cell Histiocytosis: A Case Report

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Abstract

Langerhans cell histiocytosis (LCH) is a proliferation of Langerhans cells with inflammatory cells, characterized by the activation of histiocytic cells or their proliferation that affects structures of the human body, including the temporal bone which localization is rare in children. The objective of this study is to illustrate the clinical presentation, management and prognosis of this disease, and to compare our case with previously reported series. We report the case of a young 4-year-old patient with a history of otorrhea and recurrent otalgia, with left axillary non-blowing exophthalmia progressive since 5 months associated with indurated left temporal swelling and also an indurated swelling of posterior face of the left shoulder on pathological fracture. A cerebro-orbital MRI performed at home showed an extra-conical process with extension at the temporal fossa. The patient, then, underwent an open-air biopsy of the temporal mass and the pathological analysis returned to favor Langerhans cell histiocytosis. The diagnosis was made by immunohistochemical findings of the S-100 protein and/or the CD1 antigen. The child was treated by chemotherapy with a good outcome. Our results concord with literature. The prognosis is different and better in children with unifocal bone disease than those with a multifocal disease with a survival rate of 65% - 100%.

Keywords

Langerhans Cell, Histiocytosis, Temporal, Sphenoid, Exophthalmia

1. Introduction

Langerhans cell histiocytosis (LCH), previously called histiocytosis X, was first described by Alfred Hand, Jr. in 1893 and described a spectrum of proliferation
of Langerhans cells and were developed as unisystem disease (unifocal or multifocal) or multisystem disease [1]. Langerhans cell histiocytosis (LCH) is a proliferation of Langerhans cells with inflammatory cells. It is characterized by the activation of histiocytic cells or their proliferation that affects structures of the human body, including the temporal bone. LCH usually occurs in young children with a peak at the age of 1-4 years. The spheno-temporal localization is rare in children [2]. About 50%-80% of pediatric LCH is found in the head and neck regions. The temporal bone invasion is approximately 15% to 60% of cases [3]. Otologic presentations are common and are dominated by otalgia, otorrhea, mastoid swelling and are very similar to cholesteatoma, otitis media, otitis externa. That usually delayed the diagnosis [4].

2. Reported Case

We report the case of a young 4-year-old Moroccan patient with a history of otorrhea and recurrent otalgia treated by antibiotics without improvement. Then, he presented left axillary non-blowing progressive exophthalmia since 05 months associated with indurated left temporal swelling and also an indurated swelling of posterior face of the left shoulder on pathological fracture. The patient was clinically conscient; no deficit with Karnofsky performance score at 80. Free-field pure-tone audiometry showed moderate conductive deafness and there was no pathological nystagmus. A cerebro-orbital MRI performed showed an extra-conical process with extension at the temporal fossa (Figures 1-3).

An X-Ray performs on the left shoulder indurated shows a lytic lesion of the upper 1/3 of the humerus (Figure 4).

The patient, then, underwent an open-air biopsy of the temporal mass.

The pathological analysis returned to favor Langerhans cell histiocytosis. The diagnosis was made by hemolysin-Eosin stained with magnification that
Figure 2. MRI T1 sequence sagittal view with contrast showing extraconical process with exophthalmia.

Figure 3. MRI T1 sequence axial view showing extraconical process with exophthalmia Grade II.

Figure 4. Shoulder X-ray: presenting an lytic lesion 1/3 upper humerus with periostal reaction, showing then pathological humerus fracture. Done before shoulder’s pain and fever. Lung was normal.
showed large cells with abundant eosinophilic cytoplasm. The nuclei of these cells are irregular with prominent folds and grooves and fine chromatin (Figure 5). The immunohistochemical findings includes the expression of CD1a; S100 protein (Figure 6 and Figure 7).

The patient was treated par corticosteroid prednisone at dose of 1 mg/kg for three months followed by chemotherapy Vinblastine 6 mg/m²/dose intraveinous per week with a good outcome (Table 1).

The follow up was excellent after 3 years.

Figure 5. H.E. G × 40 Coloration hematoxylin-Eosin-stained magnification 40 showing cells with abundant eosinophilic cytoplasm and irregular nuclei.

Figure 6. Immunomarquage with anti-PS100 antibody, The LCH cells are immunoreactive for antibodies directed against S100 protein.

Figure 7. Immunomarquage with anti-CD1a antibody the LCH cells are immunoreactive for antibodies directed against CD1a.
Table 1. Patient clinical characteristics.

| Age-sex | origin | symptoms | Histopathological analysis | Treatment |
|---------|--------|----------|---------------------------|-----------|
| 4 years old Male Moroccan | Otorrhea Recurrent otalgia Left non-blowing exophthamia Left temporal indurated swelling Pathological shoulder fracture | Langherhans cell histiocytosis with S-100 protein CD1a Antigen | Biopsy Chemotherapy |

3. Discussion

Langerhans’ cell histiocytosis formerly known as histiocytosis X includes the disorders (eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease) that express the heterogeneous clinical manifestations of the disease.

The etiology and pathogenesis are still unknown.

The clinical appearance of LCH is dependent on the location of the lesion. Due to the abnormal accumulation of histiocytes, every organ could be affected, such as the central nervous system, skin, bone, bone marrow, lung, liver, spleen and lymph nodes, and may cause associated signs and symptoms [5]. In our case left temporal bone sweeling with extension in the temporal fossa associated with the third upper humerus location.

Histopathologically, despite the relatively benign-looking histologic features, the histiocytic infiltration consists predominantly of a clonal proliferation of pathologic Langerhans cells like tissue macrophages rather than the typical dendritic shape of Langerhans cells in the skin. Also in the Immuno-chemistry analysis, neuronal marker S100 and CD1a which is specific for Langerhans cells (and thymocytes) and is not expressed by macrophages are positive, that helps to confirm the diagnosis [6].

The treatment for LCH includes local excision of the lesion, chemotherapy, steroid therapy, radiation, the use of anti-CD1a monoclonal antibodies and hormone replacement therapy in for patients with hormone deficiencies [7] [8].

Biopsy and curettage for solitary orbital lesions is recommended by most in the literature, but remission after biopsy alone has been reported by Maccheron et al. [9] [10].

Chemotherapy might be considered for orbital lesions with dura involvement like in our case.

Surgery is considered in aggressive LCH of the orbit and skull base with intracranial extension and cranial nerve involvement and completed by chemotherapy and radiation [10].

In addition to the use of systemic corticosteroids, cytostatic therapies like 2-chlorodeoxiademosin (2CdA-cladribine), cyclosporine, thalidomide and myeloablative therapies were prescribed [11] [12]. Single system disease can be treated with local injectable intratumoral steroids [13] [14]. Following surgery,
numerous investigators recommend low-dose radiotherapy (10 - 20 Gy) in cases in which the tissue involved cannot be completely excised [14].

Recurrences are treated by low dose radiation therapy in most of the cases, but still have side-effects or long-term sequelae such as secondary neoplasms [15].

Our results concord with other studies, the prognosis of unifocal bone disease is excellent and children with a multifocal disease have a survival rate of 65% - 100%.

4. Conclusion

Langerhans cell histiocytosis is a rare disease. If the temporal mass is associated with chronic otitis and otorrhea, that contributes to be a high-index proliferation. A biopsy must be done in the presence of a temporal bone lytic lesion. The diagnoses are supported by the presence of S100 and CD1a in the immunohistochemical analysis. Chemotherapy brings good results and it is the reference therapeutic modality.

Conflicts of Interest

All the authors declare that they have no competing financial, professional, or personal interests that might have influenced the presentation of this work.

References

[1] Margo, C.E. and Goldman, D.R. (2008) Langerhans Cell Histiocytosis. Survey of Ophthalmology, 53, 332-358. https://doi.org/10.1016/j.survophthal.2008.04.007
[2] Hamre, M., Hedberg, J., Buckley, J., et al. (1997) Langerhans Cell Histiocytosis: An Exploratory Epidemiologic Study of 177 Cases. Medical and Pediatric Oncology, 28, 92-97. https://doi.org/10.1002/(SICI)1096-911X(199702)28:2<92::AID-MPO2>3.0.CO;2-N
[3] D’Ambrosio, N., Soohoo, S., Warshall, C., Johnson, A. and Karimi, S. (2008) Craniofacial and Intracranial Manifestations of Langerhans Cell Histiocytosis: Report of Findings in 100 Cases. American Journal of Roentgenology, 191, 589-597. https://doi.org/10.2214/AJR.07.3573
[4] Modest, M.C., Garcia, J.J., Arndt, C.S. and Carlson, M.L. (2016) Langerhans Cell Histiocytosis of the Temporal Bone: A Review of 29 Cases at a Single Center. Laryngoscope, 126, 1899-1904. https://doi.org/10.1002/lary.25773
[5] The Writing Group of the Histiocyte Society (1987) Histiocytosis Syndromes in Children. The Lancet, 1, 208-209.
[6] Risdall, R.J., Dehner, L.P., Duray, P., Kobrinsky, N., Robison, L. and Nesbit, M.E. (1983) Histiocytosis X Langerhans’ Cell Histiocytosis. Prognostic Role of Histopathology. Archives of Pathology & Laboratory Medicine, 107, 59-63.
[7] Allen, C.E. and McClain, K.L. (2007) Langerhans Cell Histiocytosis: A Review of Past, Current and Future Therapies. Drugs Today, 43, 627-643. https://doi.org/10.1358/dot.2007.43.9.1088823
[8] Allen, C.E., Ladisch, S. and McClain, K.L. (2015) How I Treat Langerhans Cell Histiocytosis. Blood, 126, 26-35. https://doi.org/10.1182/blood-2014-12-569301
[9] Jubran, R.F., Marachelian, A., Dorey, F. and Malogolowkin, M. (2005) Predictors of
Outcome in Children with Langerhans Cell Histiocytosis. *Pediatric Blood & Cancer, 45*, 37-42. [https://doi.org/10.1002/pbc.20364](https://doi.org/10.1002/pbc.20364)

[10] Maccheron, L.J., McNab, A.A., Elder, J., *et al.* (2006) Ocular Adnexal Langerhans Cell Histiocytosis Clinical Features and Management. *Orbit, 25*, 169-177. [https://doi.org/10.1080/01676830600669486](https://doi.org/10.1080/01676830600669486)

[11] Bayazit, Y., Sirikci, A., Bayaram, M., Kanlikama, M., Demir, A. and Bakir, K. (2001) Eosinophilic Granuloma of the Temporal Bone. *Auris Nasus Larynx, 28*, 99-102. [https://doi.org/10.1016/S0385-8146(00)00078-X](https://doi.org/10.1016/S0385-8146(00)00078-X)

[12] Martini, A., Aimoni, C., Trevisani, M. and Marangoni, P. (2000) Langerhans’ Cell Histiocytosis: Report of a Case with Temporal Localization. *International Journal of Pediatric Otorhinolaryngology, 55*, 51-56. [https://doi.org/10.1016/S0165-5876(00)00374-8](https://doi.org/10.1016/S0165-5876(00)00374-8)

[13] Humberlo de Barros, F., Granjeiro, R.C. and de Negreiros, J. (2009) Langerhans Cell Histiocytosis in Otorhinolaryngology. *International Archives of Otorhinolaryngology, 13*, 444-449.

[14] Dornfeld, S., Winkler, C., Dörr, W. and Herrmann, T. (1998) The Radiotherapy of Histiocytosis X. A Case Report of an Eosinophilic Granuloma in Adulthood and a Review of the Literature. *Strahlentherapie und Onkolgie, 174*, 534-535. (In German) [https://doi.org/10.1007/BF03038987](https://doi.org/10.1007/BF03038987)

[15] Holbach, L.M., Colombo, F., Heckmann, J.G., Strauss, C. and Dobig, C. (2000) Langerhans-Cell Histiocytosis of the Orbit, Diagnosis, Treatment and Outcome in three Patients—Children and Adults. *Klinische Monatsblätter für Augenheilkunde, 217*, 370-373.