

**Case Report**
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## Langerhans Cell Histiocytosis of the Temporal Bone in a Child with Central Diabetes Insipidus

Mariam LAGRINE<sup>1</sup>, Youness LABANI<sup>2</sup>, Karima EL FAKIRI<sup>1</sup>, Noureddine RADA<sup>1</sup>, Ghizlane DRAISS<sup>1</sup>, Hassan NOURI<sup>2</sup>, Abdelaziz RAJI<sup>2</sup>, Mohammed BOUSKRAOUI<sup>1</sup>

<sup>1</sup>Pediatric A Departement, Mohammed VI University Hospital Center, Marrakech, Morocco

<sup>2</sup>ENT-HNS Department, Mohammed VI University Hospital Center, Marrakech, Morocco

**ABSTRACT**

Langerhans cell histiocytosis is a rare clonal disease defined by an accumulation of dendritic cells with Langerhans cell immunological properties within different organs of the body. We intend to describe a new case of Langerhans cell histiocytosis of the temporal bone in a child with central diabetes insipidus. The child presented the polyuria-polydipsia syndrome for 6 months with Chronic bilateral otorrhea. On physical examination, there were bilateral retroauricular redness and induration. The otoscopy visualized inflammatory polyps filling all of the right and left external ear canal. Biological exploration confirmed the central diabetes insipidus. Magnetic resonance imaging of the brain showed the presence of pituitary infiltration. Computed tomography showed osseous lysis of the temporal bone bilaterally. A retroauricular biopsy was performed under general anesthesia. Histological and immunohistochemical evaluation confirmed the diagnosis of Langerhans cell histiocytosis.

**\*Corresponding author**

Mariam Lagrine. Pediatric A Departement, Mohammed VI University Hospital Center, Marrakech, Morocco, E-main: lagrine.mariam@gmail.com

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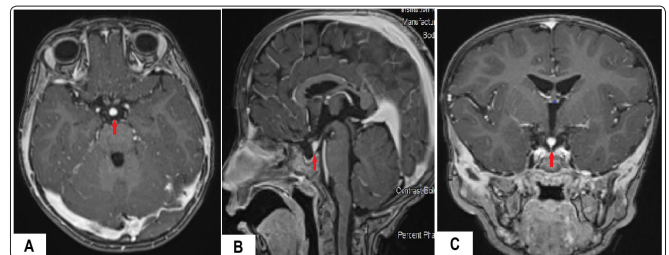
**Introduction**

Langerhans histiocytosis (LH) is a rare clonal pathology defined by an accumulation of dendritic cells with the immunological characteristics of Langerhans cell within different organs of the body [1]. Its clinical presentation is highly variable and can range from isolated, self-healing skin and bone lesions to life-threatening multisystem disease. We report a new case of LH in a 2-year-old child with central diabetes insipidus with bilateral temporal bone involvement.

**Case Report**

2-year-old child, with no particular pathological history, who has presented for 6 months a polyuropolydipsic syndrome with chronic bilateral otorrhea. Upon physical examination, the child presented bilateral retroauricular redness and induration. Otoscopy revealed the presence of inflammatory polyps filling both the right and left external ear canals. On laboratory testing, fasting blood glucose was normal at 0.8 g / L, urinary osmolality was reduced to 112 mOsm / L and blood sodium was normal at 140 mmol / L. Urinary osmolality remained low even after the fluid restriction test. Urinary osmolality increased to 326 mOsm / L after 4 hours of administration of 10 µg of Desmopressin (Minirin®) spray. The diagnosis of central diabetes insipidus was suspected. Brain magnetic resonance imaging (MRI) showed the presence of pituitary infiltration with enlargement of the pituitary stem enhancing to T1 sequences after injection of contrast agent (Figure 1). Temporal bone CT scan showed bone lysis involving the temporal bone bilaterally (Figure 2). A deep right

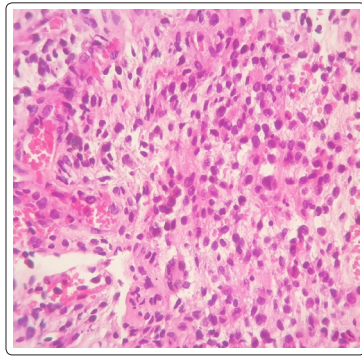
retroauricular biopsy was performed under general anesthesia. The histological study showed a fibrous tissue with inflammatory cells and histiocytic infiltration (Figure 3). Immunohistochemical study showed positive staining for PS100, CD1a and CD68. The diagnosis of LH was confirmed.



**Figure 1:** Brain MRI showing the pituitary infiltration (A: axial section, B: sagittal section, C: coronal section)



**Figure 2:** Temporal bone CT scan showed bone lysis involving the temporal bone bilaterally



**Figure 3:** Histological study showed a fibrous tissue with inflammatory cells and histiocytic infiltration

### Discussion

LH is a systemic disease that can affect several organs; preferably the bone, the skin, the central nervous system, the lungs, the liver and the spleen. LH can occur at any age. It mainly affects children between 1 and 4 years old. The incidence of the disease in children under 15 years of age is 2 to 9 cases per million per year, with a slight male predominance [2].

The clinical presentation of the disease is very variable and there are several clinical forms. LH was formerly called Histiocytosis X and included 3 clinical forms which are eosinophilic granuloma, Hand-Schüller Christian disease and Letterer-Siwe disease [3]. Currently, this nomenclature is abandoned; HL is therefore classified according to the number of lesions and organs affected. A distinction is made between the mono-tissue unifocal form, the mono-tissue multifocal form and the multi-tissue form [4]. Damage to certain organs such as the lung, liver, spleen and bone marrow worsens the prognosis [5].

Otologic involvement in LH varies from 4 to 61% depending on the series [6]. It is the consequence of the temporal bone involvement which is often unilateral but can be bilateral. The clinical presentation is similar to that of chronic otitis media or otitis externa. Radiologically, the CT scan of the temporal bone shows limited or diffuse bone lysis, preferentially affecting the mastoid, the middle ear and the outer ear.

Central diabetes insipidus is the most common endocrine manifestation; it is found in 15 to 50% of patients with LH with involvement of the hypothalamic-pituitary axis [7]. It results from a defective secretion of the antidiuretic hormone (ADH). An anterior pituitary hormone deficiency can also be found and which can be manifested by growth retardation, hypothyroidism and delayed puberty.

We report the case of a 2-year-old child with LH with bilateral temporal bone involvement associated with central pituitary involvement responsible for diabetes insipidus. Rare cases with similar presentation have been reported [8-10].

The diagnosis of LH requires the association of radioclinical presentation with a histological and immunohistochemical appearance compatible with the disease. The identification of characteristic cells and the presence of specific markers, such as CD1a and / or CD207, allow a definitive diagnosis of the disease [4].

### Conclusion

LH is a rare condition that manifests in different clinical forms. Its diagnosis is difficult and often carried out late. The clinician should become familiar with the different manifestations suggesting the disease such as the presence of central diabetes insipidus or bilateral temporal diffuse bone lysis.

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