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Orbital Involvement Revealing Histiocytosis In A Child: A Rare Case Report

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ABSTRACT

Histiocytosis is a rare disease primarily affecting children, with ophthalmological involvement in 20% of cases. We report a 2-year-old with unilateral exophthalmos and a right temporal mass, diagnosed as Langerhans cell histiocytosis. After initial chemotherapy complications, treatment with Vemurafenib led to a favorable response. Early diagnosis and tailored treatment are crucial for managing this unpredictable condition

Introduction:

Histiocytosis is a rare disease most often affecting small children, sometimes young adults, the cause of which is unknown. Ophthalmological involvement has been observed in 20% of cases (1). However, it is not known whether this condition is malignant. Its evolution is unpredictable, and spontaneous regression may be observed after a simple biopsy, among other procedures.

Observation:

A 2-year-old child with no previous history presented to the ophthalmological emergency department with unilateral axial exophthalmos that was not reducible, accompanied by a mass in the right temporal fossa that had appeared for one month. Ophthalmological examination revealed preserved visual acuity. Anterior segment and fundus examinations were normal. Palpation revealed an orbital swelling at the superior-external angle of the left orbit, which was painless and not very mobile. Exophthalmos measured 22 mm on the left side, with downward and inward deviation. The lymph nodes were free. The rest of the ophthalmological and general examinations were normal. Orbito-cerebral CT showed a right frontal lesional process with bone lysis corresponding to stage 1 exophthalmos. Magnetic resonance imaging confirmed a right frontal lytic process with orbital and soft-tissue extension, possibly related to a secondary localization of a neuroblastoma or primary tumor, to be compared with histological data. A biopsy was performed for anatomopathological and immunohistochemical study, and revealed Langerhansian histiocytosis. The extension work-up did not reveal any other tumour locations. In collaboration with pediatric oncologists, the child was treated with induction chemotherapy with 4 courses of vinblastine 1 week apart. During treatment, the child developed infections that did not respond to treatment, prompting the pediatricians to replace the chemotherapy with Vemurafenib

(Zelboraf). The evolution was favorable, with a good response to treatment. The child is still under treatment.

Figure 1 : orbital swelling at the superior-external angle of the left orbit,

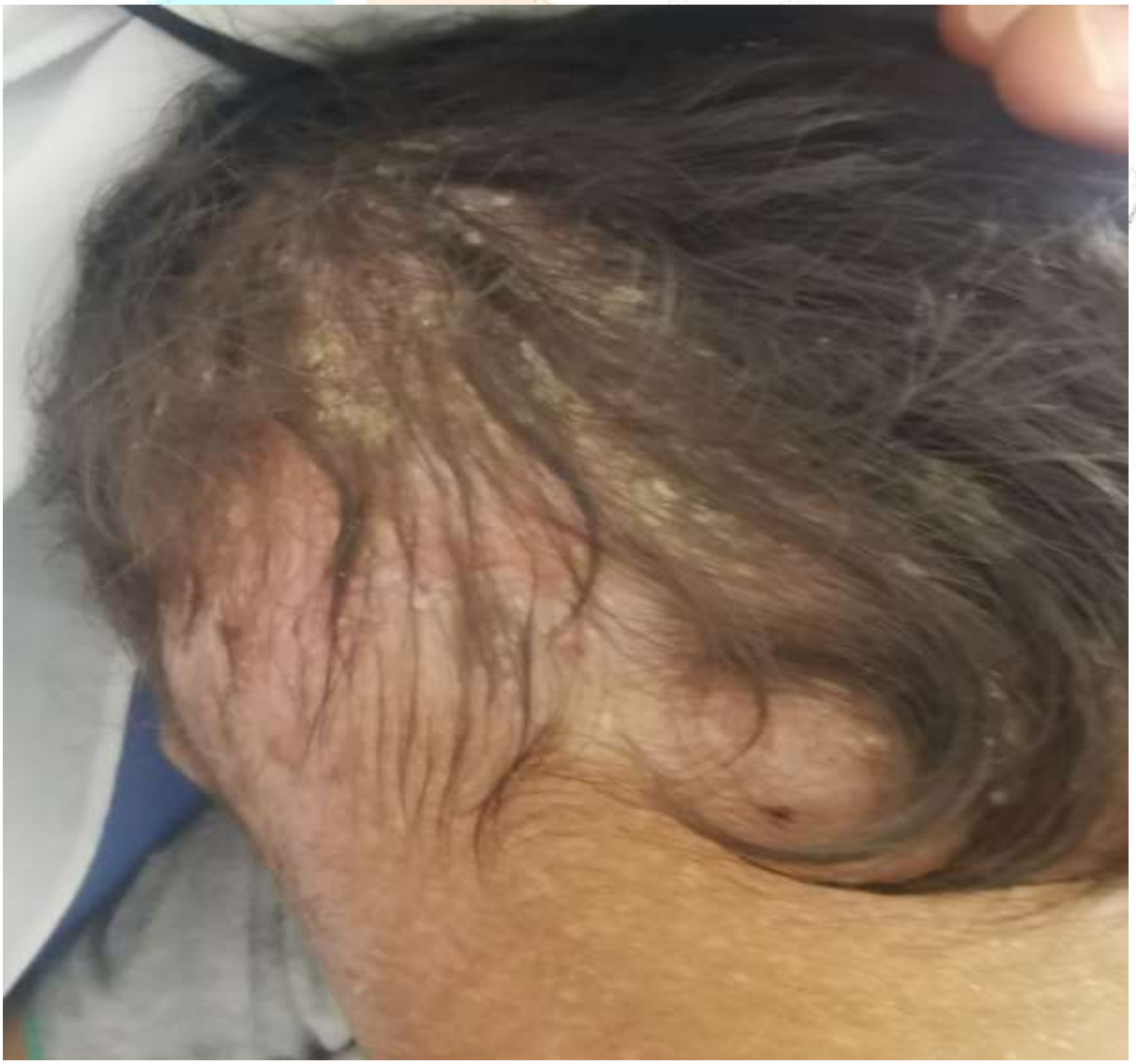

Figure 2 : histiocytosis "cradle cap" scalp lesions

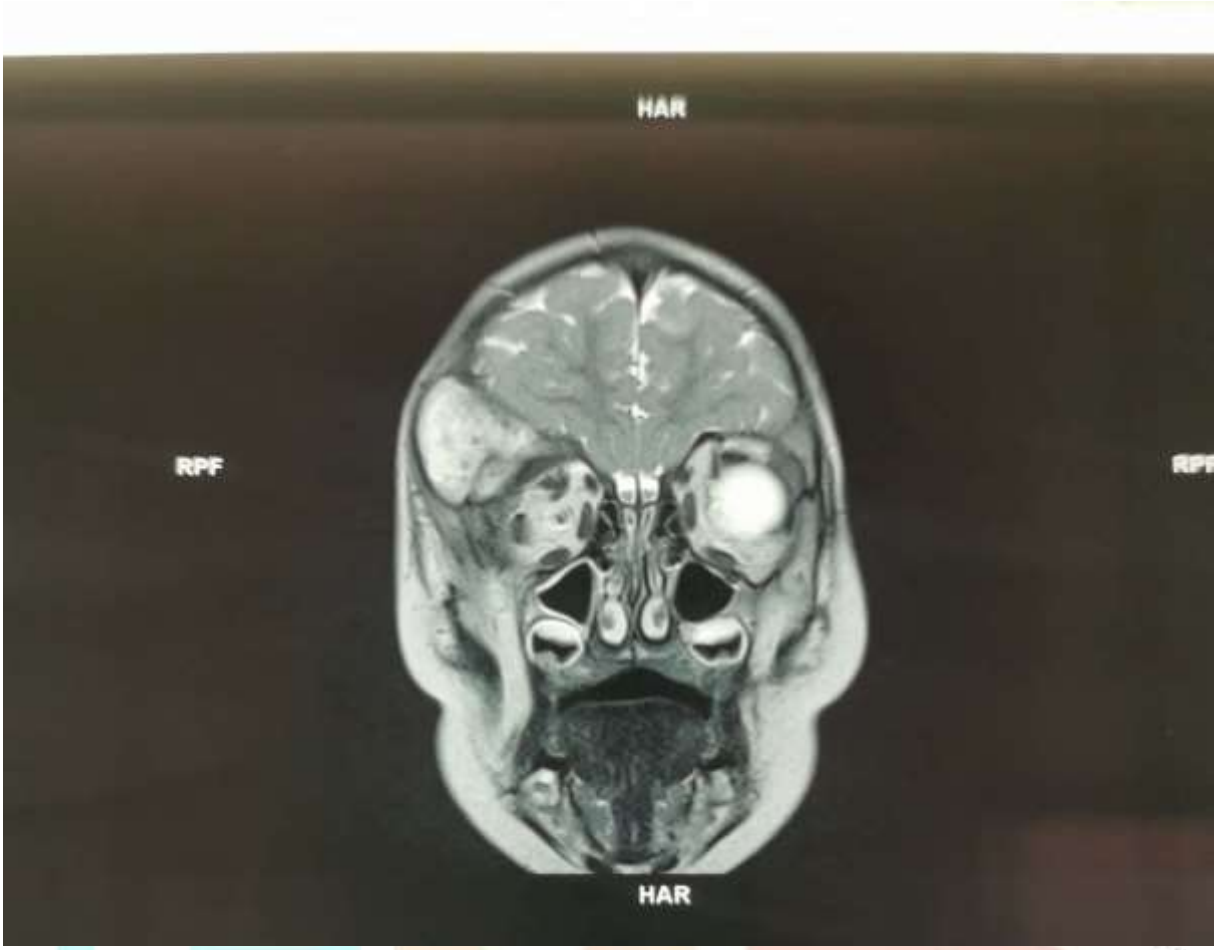

Figure 3 : CT showed a right frontal lesional process with bone lysis corresponding

Discussion:

Histiocytosis is a group of diseases characterized by abnormal proliferation of histiocytic cells, which derive from monocyte-macrophages. Langerhans cell histiocytosis (HCL) is the most common form of this disease. Manifestations of orbital involvement may include an orbital mass, periorbital edema, exophthalmos or visual disturbances (2).Langerhans histiocytosis (HL), also known as histiocytosis X (HX), is a proliferative disease of the reticulohistiocytic system. The etiopathogenesis of HL remains unknown, although certain cytokines and triggering factors, notably viral, have been suggested(2). LH occurs mainly in children and young adults, with a peak frequency between the ages of 1 and 4, and a male predominance (3), as in our patient.

The most frequent form is a single bone localization, called eosinophilic granuloma, which generally has a good prognosis(4).

Orbital involvement is part of the Hand-Schüller-Christian syndrome, which affects the skull base, pituitary gland and orbital apex. Approximately 20% of HL cases present with ophthalmological manifestations, eosinophilic granuloma of the frontal bone being the most common form(5). Computed tomography (CT scan) and magnetic resonance imaging (MRI) are used to assess lesions. Typically, osteolytic lesions appear without associated reactive calcifications. Diagnosis of certainty is based on histopathological examination, demonstrating Birbeck granules or CD1a antigens. Positive immunohistochemistry for S-100 protein and anti-CD1a antibodies confirms the diagnosis(6).

Treatment varies according to clinical form and age. Simple surveillance may be sufficient for non-vital localized forms and those that regress spontaneously. Surgery is indicated for accessible bony localizations, but extensive curettage can lead to problems of reossification(7). Radiotherapy can be used to treat multiple or inoperable bone lesions(8). Intra-lesional injection of methylprednisolone has proved successful in the treatment of orbital eosinophilic granulomas(9). Chemotherapy, often combined with

systemic corticosteroid therapy, is considered for disseminated forms or those with ocular compression. Bone marrow transplants and immunotherapy are reserved for resistant forms(10).

Conclusion:

Histiocytosis, particularly Langerhans cell histiocytosis, can present with orbital involvement in children, manifesting as exophthalmos and orbital masses. Early diagnosis through imaging and histopathological examination is crucial. Treatment options, including chemotherapy and targeted therapy, can lead to favorable outcomes. Ongoing monitoring and multidisciplinary collaboration are essential for managing this unpredictable disease.

REFERENCES

1. HARRIS G.J. – Eosinophilic granuloma of the orbit: a paradox of aggressive destruction responsive to minimal intervention.Trans Am Ophthalmol Soc, 2003; 101:93-105.
2. Yagci B, Varan A, Caglar M, Söylemezoğlu F, Sungur A, Orhan D, Yalçin B, Akyüz C, Kutluk T, Büyükpamukçu M. Langerhans cell histiocytosis: Retrospective analysis of 217 cases in a single center. *Pediatr Hematol Oncol* 2008; **25**: 399–408.
3. Vosoghi H, Rodriguez-Galindo C, Wilson MW. Orbital involvement in langerhans cell histiocytosis. Ophthal Plast Reconstr Surg. 2009;25(6):430–3.
4. Zausinger S.Muller A.Bise K.Klauss V.Eosinophilic granuloma of the orbit in an adult woman. Acta Neurochir (Wien). 2000; 142: 215-217
5. Elena Sieni, Carmen Barba, Marzia Mortilla, et al. Early Diagnosis and Monitoring of Neurodegenerative Langerhans Cell Histiocytosis [J]. PLOS one. 2015,0131635.
6. Herwig MC, Wojno T, Zhang Q, Grossniklaus HE. Langerhans cell histiocytosis of the orbit: five clinicopathologic cases and review of the literature [J]. Surv Ophthalmol. 2013;58(4):330–40.
7. Maccheron L.J.McNab A.A.Elder J.et al.Ocular adnexal Langerhans cell histiocytosis: clinical features and management.Orbit. 2006; 25: 169-177
8. Woo K.I.Harris G.J.Eosinophilic granuloma of the orbit: understanding the paradox of aggressive destruction responsive to minimal intervention. Ophthal Plast Reconstr Surg. 2003; 19: 429-439
9.
10. Guo Y, Wang L, Ma R, et al. JiangTang XiaoKe granule attenuates cathepsin K expression and improves IGF-1 expression in the bone of high fat diet induced KK-ay diabetic mice. Life Sci. 2016;48:24–30.