CASE REPORT

Benign Cephalic Histiocytosis: The face of a rare pathology

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SUMMARY

Benign cephalic histiocytosis (BCH) is a rare dermatological disease that primarily affects infants and young children, characterized by the appearance of multiple asymptomatic, flat, erythematous-yellow papules, often in the cephalic region. We present the case of a 1 year and 3 months old male patient with asymptomatic brown papules on the face. Following clinical-pathological correlation, a diagnosis of benign cephalic histiocytosis was established. During follow-up, stability and involution of the lesions were observed.

INTRODUCTION

Benign cephalic histiocytosis is an uncommon disease that primarily affects the craniofacial region of children.[1,5] It is a type of histiocytosis, a group of diseases that involve the abnormal proliferation of histiocytes in the soft tissues of the head and neck, which can cause facial and cranial deformities. Unlike other more serious forms of histiocytosis, such as Langerhans cell histiocytosis, BCH is not associated with malignancy or serious systemic involvement. The exact cause of BCH is still unknown, but it has been proposed that it may be linked to a transient inflammatory or immunological response.[3]

Benign cephalic histiocytosis (BCH) typically manifests in infants between 3 and 6 months of age. The characteristic skin lesions are erythematous papules measuring 2 to 6 mm in diameter, which may be accompanied by a scaly surface.[3,11] These papules usually appear on the scalp, forehead, cheeks, and upper trunk. Sometimes, they may extend to other areas of the body, although this is less common.[2]

An important feature of BCH is that, despite their alarming appearance, the lesions typically do not cause significant discomfort to the patient. They are not itchy or painful, which helps differentiate them from other dermatological conditions.[4]

CLINICAL CASE

A 1 year and 3 months old male patient, with no significant medical history; the mother reports that since the age of 7 months, he has had asymptomatic papular lesions on his face. Topical treatment has been applied

without improvement. Over the months, the number of lesions on the face has increased.

PHYSICAL EXAMINATION

Localized dermatosis on the face, characterized by multiple brown papules, approximately 2 mm in diameter, non-confluent (Figure 1). Upon dermatoscopy: lesions show a sunset pattern (Figure 2).

In the histopathological examination, a diffuse inflammatory infiltrate is observed in the upper and mid dermis, composed of elongated and spindle-shaped histiocytes that tend to converge, forming multinucleated cells. This infiltrate is accompanied by numerous eosinophils. There is also evidence of erythrocyte extravasation into the upper dermis. The infiltrate does not invade the epithelium or its appendages. The epidermis shows rectification of the ridge patterns and is covered by orthokeratosis at the crest (Figure 3).

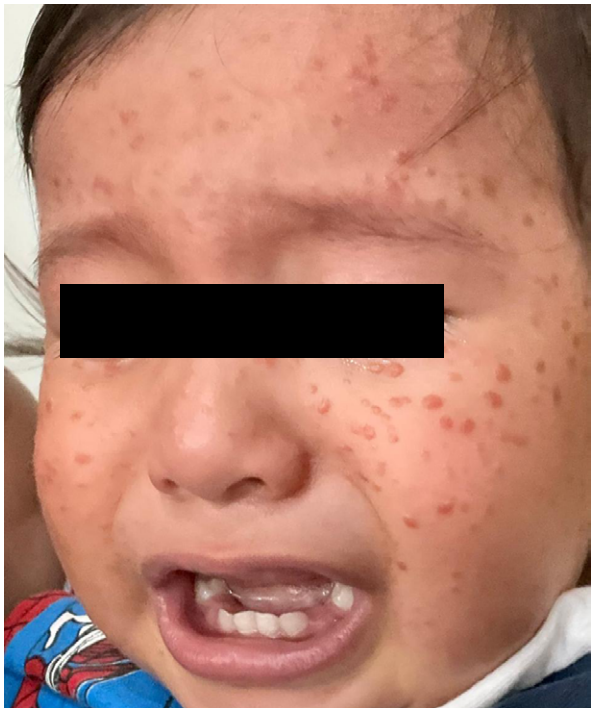

Figure 1. Brown papules.

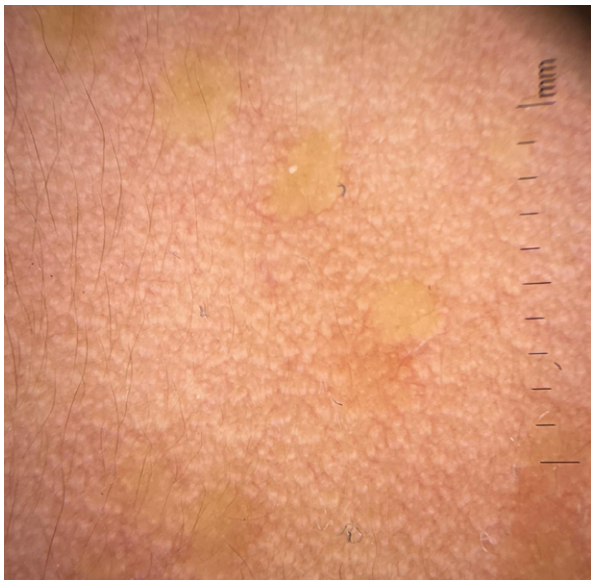

Figure 2. Lesions with a sunset pattern.

DISCUSSION

Gianotti et al. first described benign cephalic histiocytosis in 1971; it is a rare and underdiagnosed disease,[5] with just over 70 documented cases in the literature.[4] It predominates in males, with a mean age of 7 months at diagnosis. Fifty percent of cases occur before 6 months8, but it can occur up to 3 years of age, as seen in our patient who presented lesions at 7 months of age. The clinical manifestations, histopathological examination, and immunophenotype determine its diagnosis. Its main distribution area is the face, although some lesions can be found on the trunk, upper limbs, and buttocks.[4]

In 2016, a group of specialists divided histiocytic disorders into five groups based on their clinical, histological, and molecular characteristics due to advances in diagnostic methods. Table 1 shows the current classification of histiocytoses.[7]

The entities that cause non-Langerhans cell histiocytosis in the skin and mucous membranes are classified based on their clinical appearance, immunophenotype,

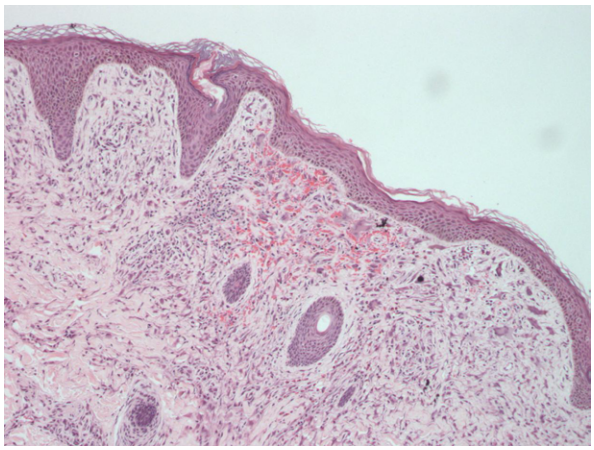

Figure 3. The epidermis shows orthokeratosis and slight hyperpigmentation of the basal layer; diffuse inflammatory infiltrate in the upper and mid dermis, composed of elongated and spindle-shaped histiocytes.

Table 1. Current Classification of Histiocytosis According to the Histiocyte Society:[10]

GROUP I	LANGERHANS GROUP	Langerhans cell histiocytosis Indeterminate cell histiocytosis Erdheim-Chester disease Mixed Langerhans cell histiocytosis and Erdheim-Chester disease
GROUP C	NON-LANGERHANS CUTANEOUS AND MUCOCUTANEOUS HISTIOCYTOSIS	Isolated cutaneous With systemic component
GROUP M	MALIGNANT HISTIOCYTOSIS	Primary Secondary
GROUP R	DORFMAN DISEASE	Familial Classical (nodal) Extranodal Associated with neoplasms Associated with immune diseases
GROUP H	HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS AND MACROPHAGE ACTIVATION SYNDROME	Primary Secondary Of unknown origin

and whether or not there is systemic involvement.[2] Recently, benign cephalic histiocytosis (BCH) has been classified as a juvenile xanthogranuloma (within Group C of non-Langerhans cell histiocytosis), along with other entities without systemic involvement.[5]

BCH is a clinical diagnosis based on the observation of lesions and the patient's medical history. Despite its rarity and similarity of manifestations with other skin conditions, a skin biopsy is often necessary. BCH is self-limiting, resolving spontaneously within a few months to a year without the need for specific treatment. The prognosis is excellent, with no long-term complications or significant recurrences.[9] In our case, the diagnosis of BCH was based on clinical criteria, including the onset of the disease in early childhood, the typical clinical morphology of the lesions, and their predominant distribution on the face, along with histopathological study.

CONSENT

Informed consent was obtained from the patient's representative for the publication of the data and photos in this article.

CONCLUSION

Benign cephalic histiocytosis is a rare but important dermatological entity to recognize in pediatric practice, with a good prognosis. Its benign and self-limiting presentation distinguishes it from other forms of histiocytosis and skin diseases in childhood. An accurate diagnosis and a clear understanding of its natural course can provide reassurance to parents and prevent unnecessary interventions. As further research is conducted, it is hoped that knowledge about the etiology and optimal management of this interesting condition will expand.

REFERENCES

1. A.F. Abreu, e. (2024). Benign cephalic histiocytosis: exuberant manifestation in an infant. Anais Brasileiros de Dermatologia, 451-454. doi: https://doi.org/10.1016/j.abd.2022.11.008
2. Bertino L, P. F. (2022). Benign cephalic histiocytosis with extra-facial manifestations. Journal of Paediatrics and Child Health [Internet]. doi: https://doi.org/10.1111/jpc.
3. Castillo, B. e. (2022). HISTIOCITOSIS CEFÁLICA BENIGNA. A PROPÓSITO DE UN CASO. ARCHIVOS DE PATOLOGIA - ISSN: 2660 -7611. doi: https://doi.org/10.17811/ap.v4i1.20843
4. De Avó HS, Y. S. (2020). Benign cephalic histiocytosis: a case report of unusual presentation with initial appearance of extrafacial lesions. International Journal of Dermatology [Internet]. doi: https://doi.org/10.1111/ijd.15058
5. Díaz, E. S. (2022). Histiocitosis cefálica benigna simulando una mastocitosis. Actas Dermo-sifiliograficas, 195-198. doi: DOI: 10.1016/j.ad.2020.05.014
6. Ekinci AP, B. N. (2017). Novel Clinical Observations on Benign Cephalic Histiocytosis in a Large Series. Pediatric Dermatology [Internet]. doi: Available from: https://doi.org/10.1111/pde.13153
7. Emile JF, A. O. (2016). Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. Blood, 2672-81.
8. Fraitag S, E. J. (2021). Cutaneous histiocytoses in children. Histopathology [Internet], 196-215. doi: https://doi.org/10.1111/his.14569
9. Monir RL, M. K. (2023). Red-brown papules in a 13-month-old. Pediatric Dermatology [Internet], 201-3. doi: https://doi.org/10.1111/pde.15137
10. Morán Edna, e. (2018). Histiocitosis de células de Langerhans: nuevos conceptos moleculares y clínicos. Dermatología Cosmética, Médica y Quirúrgica, 36-44.
11. Polat Ekinci A, B. N. (2017). Novel Clinical Observations on Benign Cephalic Histiocytosis in a Large Series. Pediatric Dermatology [Internet], 392-397. doi: 10.1111/pde.13153.