Benign Cephalic Histiocytosis in an Infant

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A 17-month-old boy was referred to our pediatric dermatology clinic for evaluation of a facial rash that had been present since age 6 months. The patient's mother reported that the eruption had initially started as papules on the right cheek and subsequently spread to the left **(Figure 1)**, with new papules appearing gradually over time.

The eruption was asymptomatic, unchanged with daily moisturizers, and seemed to become more noticeable when the patient was irritated or crying. There was no family history of similar skin eruptions.

Physical examination

Upon examination, multiple pink-orange, dermal papules with increased vascularity were seen on the bilateral cheeks, forehead, and left upper eyelid. The patient's trunk, extremities, oral mucosa, and acral surfaces were spared.

A punch biopsy of a papule on the left lateral cheek revealed an infiltrate of histiocytes and a few eosinophils in the superficial dermis **(Figures 2 and 3)**. Clinical and histopathologic findings were consistent with a diagnosis of benign cephalic histiocytosis.

Discussion

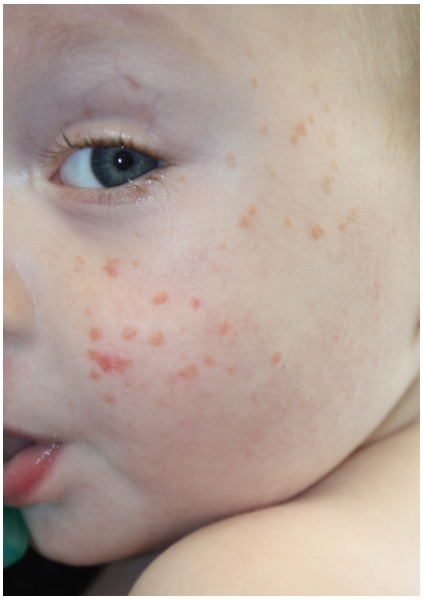

Figure 1. Patient's face with scattered rash.

First described in 1971 by Gianotti and colleagues, benign cephalic histiocytosis, or papillary histiocytosis of the head, is a rare variant of non-Langerhans cell histiocytosis, typically occurring in infants and young children.[1] The average age of onset is 15 months, and there exists an equal distribution among boys and girls.[2]

Benign cephalic histiocytosis presents as asymptomatic, small, yellow to red-brown macules and papules located on the head and neck. Lesions can spread to the trunk and extremities, and one study suggests the eruption may involve noncephalic skin more often than previously thought.[3] The disorder is limited to the skin, sparing the mucosal membranes and internal organs. The eruption resolves spontaneously over an average of 50 months, sometimes leaving behind hyperpigmentation or small, atrophic scarring.[2,4] Although benign cephalic histiocytosis is not thought to have systemic involvement, 2 reports of coexisting systemic illness, with dia-

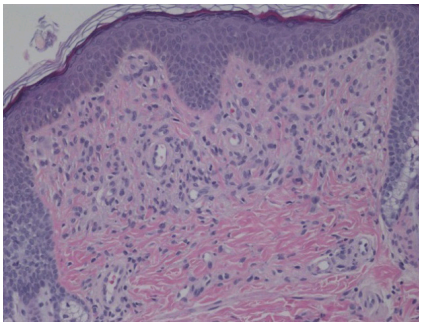

Figure 2. Results of a skin biopsy showed increased histiocytes in the papillary dermis (hematoxylin and eosin stain at 200× magnification).

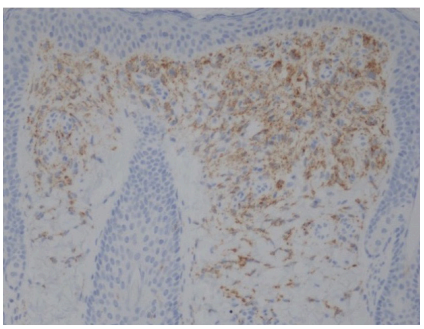

Figure 3. The histiocytes were diffusely positive for CD68 (CD68 Immunohistochemical stain, 200× magnification).

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betes mellitus and diabetes insipidus in separate instances, have been reported.[5,6] However, these associations were thought to be coincidental.

The diagnosis is mostly clinical but should be confirmed with histopathology and immunohistochemical staining.[4] Histology demonstrated a histiocytic infiltrate in the superficial to midreticular dermis with characteristic staining pattern.[2-4] The histiocytes stain positive for factor XIIIa, HAM-56, fascin, and CD68 and stain negative for CD1a, langerin, and S100.[7-9]

Benign cephalic histiocytosis can be mistaken for juvenile xanthogranluoma, which was considered in our patient's case. Juvenile xanthogranluoma is often more widespread and can have extracutaneous involvement, specifically ocular, which is not seen in patients with benign cephalic histiocytosis.[2-4] Benign cephalic histiocytosis can also be distinguished from juvenile xanthogranluoma on histology.

Generalized eruptive histiocytosis and Langerhans cell histiocytosis should also be considered in the differential diagnosis. Generalized eruptive histiocytosis is histologically similar to benign cephalic histiocytosis but is seen mostly in adults and can have mucosal involvement.[2-4] Langerhans cell histiocytosis differs from benign cephalic histiocytosis with a scaled, crusted, or sometimes petechial eruption often on flexor, scalp, or intertriginous surfaces and has the possibility of visceral involvement.[2] Histology and immunostaining of Langerhans cell histiocytosis is distinctive.[2,8,9]

Conclusions

Benign cephalic histiocytosis is a rare disease with an estimated 60 cases reported in the English-language literature.[3,4] Despite its rarity, benign cephalic histiocytosis is an important disease to recognize when considering histiocytic skin lesions or other mimickers. Given its extremely low prevalence, it is our hope that sharing this case assists clinicians in recognizing benign cephalic histiocytosis and avoid unnecessary, aggressive treatment or workup for this self-limiting, benign condition.

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