

# Case Letter

## Large Plaque-Type Benign Cephalic Histiocytosis Showing Rapid Aggravation Following Vaccination

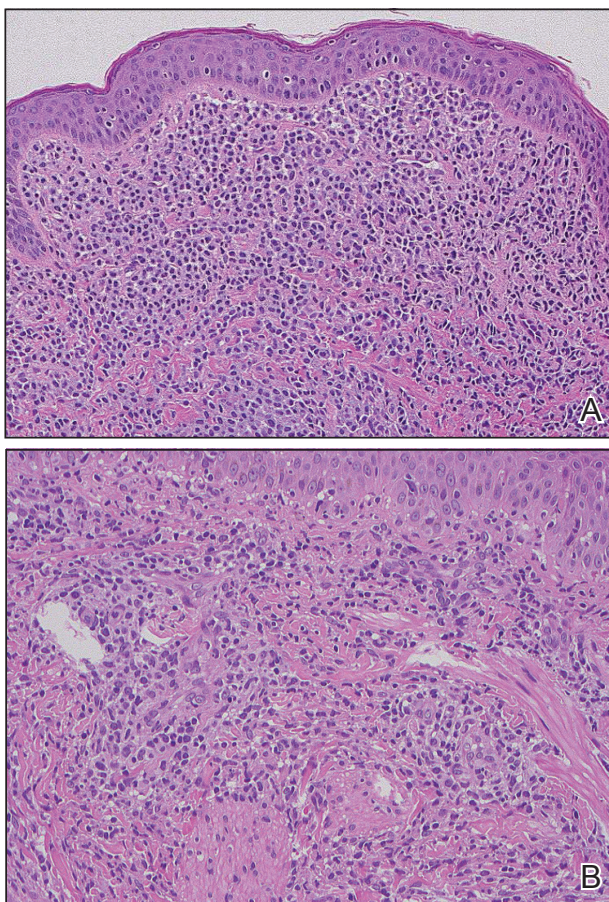
To the Editor:

Benign cephalic histiocytosis (BCH) is a rare benign dermatosis in which self-healing papular eruptions develop. This condition is classified as non-X histiocytosis and behaves as a benign histiocytic proliferation. Since the first report by Gianotti et al<sup>1</sup> in 1971, the etiology and pathophysiology of this condition have not been elucidated. The clinical and histologic features of BCH overlap with juvenile xanthogranuloma (JXG) and generalized eruptive histiocytosis.<sup>2</sup> Rodriguez-Jurado et al<sup>3</sup> reported that BCH showed clinical evolution to JXG with varicella-zoster infection, suggesting that BCH is part of a spectrum of diseases that non-Langerhans cell histiocytosis encompasses. We present a case of plaque-type BCH in an infant.

An 11-month-old male infant presented with asymptomatic, ill-defined, infiltrated plaques on the cheeks of 1 month's duration (Figure 1). Similar red plaques were present on both ears, the left forearm, and the right middle finger. One month following the initial onset as tiny red papules, the lesions had become larger and more prominent after an episode of fever (temperature, 39°C) and immunization against rubella virus and *Haemophilus influenzae* type b. No therapeutic effect was observed with the application of a topical steroid at the time of presentation. A skin biopsy specimen was taken from a flat plaque on the right cheek. Histopathologic findings revealed dense cell infiltrates predominated by histiocytic cells with some vacuolization in the papillary and upper reticular dermis (Figure 2). Neither foamy histiocytic cells nor giant cells were seen. Immunohistochemical analysis revealed that the cells were CD68<sup>+</sup>



**Figure 1.** Asymptomatic red, infiltrated, flat plaques on the left cheek.

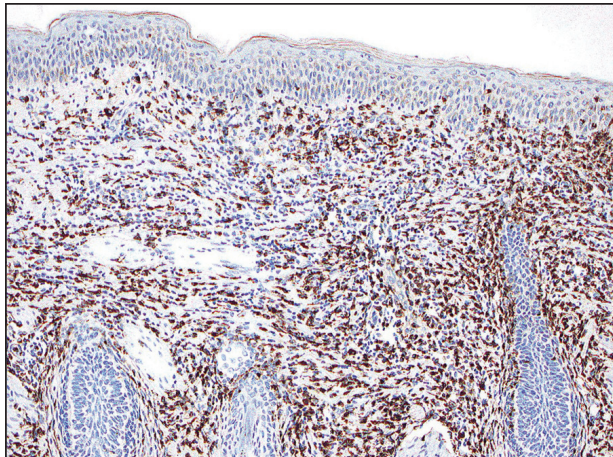


**Figure 2.** A biopsy revealed an infiltrate of histiocytes with vacuolization of cells in the papillary and upper reticular dermis (A and B) (both H&E, original magnifications  $\times 50$  and  $\times 200$ ).

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The authors report no conflict of interest.

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**Figure 3.** Immunohistochemistry revealed histiocytic infiltrate cells that were CD68<sup>+</sup> (original magnification  $\times 50$ ).

(Figure 3) and negative for S-100. Following 2 months of observation without any treatment, the plaques on the ears, left forearm, and right middle finger had begun to spontaneously subside. Overall, the remaining lesions showed complete remission by 21 months of age.

Benign cephalic histiocytosis commonly presents as smooth, round, or flat-topped papules with a diameter of 1 to 8 mm and color ranging from tan-yellow to brown-red.<sup>2</sup> The etiology of BCH is unknown. Plaque-type BCH represented by this patient is a rare feature, suggesting sarcoidosis, pernio, or Sjögren syndrome because the lesions are comprised of infiltrated erythematous plaques rather than papules. Rodriguez-Jurado et al<sup>3</sup> described a transitional form of BCH to JXG in a 2-year-old girl with facial plaques after an episode of varicella-zoster virus infection. Vasconcelos et al<sup>4</sup> also indicated that

JXG is associated with cytomegalovirus infection. These findings support the notion that BCH and non-Langerhans cell histiocytosis are on the same spectrum of diseases, representing a reactive histiocytic process to an infectious agent.

The difference in clinical manifestation of each non-Langerhans cell histiocytosis subgroup may reflect the degree or stage of infection. T lymphocytes may release different cytokines and activate macrophage migration when sensitized by a virus or vaccine. As a result, BCH may show various clinical features even when confirmed by histological analysis. Our patient might have had an early stage of BCH that would transform into JXG.

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