Circumscribed Nodular Lesion on the Face of an Infant

Clinical and Dermoscopic Findings
An 8-month-old male child born out of nonconsanguineous marriage with normal growth and developmental milestones presented with a 2-month history of an asymptomatic protuberant nodule over the right cheek. On examination, a well-defined, 2 × 2 × 3 cm solitary skin-colored, nontender, firm, nodule was present over the right mandibular region with areas of yellowish discoloration and telangiectasia present over the summit of the nodule and overlying scaling [Figure 1a]. The skin over the nodule was not pinchable, and the surrounding skin was normal. The oral cavity examination was normal. No regional lymphadenopathy was seen, and there were no other similar lesions anywhere in the body. Systemic examination was within normal limits. Dermoscopy of the lesion showed yellowish-brown orange areas, linear and curved telangiectatic vessels, a peripheral rim of scales with an upturned inner edge, and red structureless areas [Figure 1b, Dermlite DL4 10×, polarized].

Histological Findings
Histopathological examination revealed acanthotic epidermis and collection of histiocytes in the reticular dermis in sheets and nodules, admixed with few scattered foamy macrophages and multinucleated giant cells, neutrophils, and numerous eosinophils [Figure 2a]. The histiocytes showed round to oval nuclei with bland chromatin, small conspicuous nucleoli, and a moderate amount of cytoplasm [Figure 2b]. On immunohistochemistry, these cells were positive for CD68 [Figure 2c], S100, and CD163; and negative for CD1a, Langerin, and BRAF.

Diagnosis
Benign cephalic histiocytosis
Based on clinical features, dermoscopy, histopathology, and immunohistochemistry findings, diagnosis of benign cephalic histiocytosis (BCH) was made. Considering the self-limiting nature of the disease with an average age of regression of 50 months, the parents were counseled, and no active intervention was done in our patient.

Discussion
BCH is a rare, self-limiting, non-Langerhans cell histiocytosis of infants and young children. According to the revised classification by Emile et al.,[2] BCH falls under the xanthogranuloma family, C-group: cutaneous and mucocutaneous histiocytosis. Common morphological presentations of BCH include multiple skin-colored to yellowish papules in a cephalic distribution.[3] However, rarely, as in the index case, a solitary nodule may be a presenting feature. The differential diagnoses considered at presentation included juvenile xanthogranuloma (JXG), cutaneous lymphoma, cutaneous leishmaniasis, and giant molluscum contagiosum.

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JXG presents usually in early childhood as well-demarcated solitary or multiple, firm, rubbery papules or nodules, pink to red in color with a yellow tinge in early lesions, later acquiring yellow-brown hue with occasional telangiectasia. The dermoscopic finding of JXG has been described metaphorically as a “setting sun appearance.”[4,5] Histopathologically, JXG usually shows abundant foamy or lipid macrophages/histiocytes compared with BCH in which histiocytes with small conspicuous nucleoli and a moderate amount of cytoplasm are seen. Immunohistochemically, both entities are identical with CD68+, CD163+, Factor XIIIa+, and negative CD1a and S-100. However, S-100 positivity in BCH varies, and S-100-positive BCH and JXG have been reported.[6]

Cutaneous lymphomas of B-cell origin may present with large, firm, bluish, asymptomatic solitary or multiple nodules mostly in the head and neck region in the pediatric age group. The absence of blast cells that are positive for immature B-cell markers in the dermis and subcutis was against a diagnosis of cutaneous lymphoma.[7] Giant molluscum contagiosum may present as a solitary nodular lesion in an otherwise immunocompetent child.[8] However, the absence of central umbilication in the nodule, crown vessels on dermatoscopy, and intracytoplasmic inclusion bodies on histopathology excluded giant molluscum contagiosum. Cutaneous leishmaniasis typically starts over the exposed sites such as head and neck or extremities, as small papule at the site of inoculation, evolves into the nodule, and then ulcerates.[9] Dermoscopy of cutaneous leishmaniasis shows a white starburst-like pattern, teardrop-like structures, yellow tears, and salmon-colored ovoid structures.[9]

The dermoscopic features of BCH have hitherto been undescribed in the literature. Dermosscopic evaluation of the index case showed yellowish-orange areas representing the dermal accumulation of histiocytes, red structureless areas due to epidermal hemorrhage, and superficial telangiectatic vessels representing dilated blood vessels on histopathology. BCH has overlapping clinical and histological features with JXG, and reports of BCH transforming into JXG may suggest that BCH is an abortive form of JXG.[10] BCH runs a benign course with most of the eruptions regressing spontaneously by an average age of 50 months.[1]

**Photo consent**

The consent was given, and the procedures were followed.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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