Benign Cephalic Histiocytosis

Benign Sefalik Histiyositozis

Ekrem GÜLER, MD,[a]
Özer ARICAN, MD,[b]
Mehmet DAVUTOĞLU, MD,[a]
Harun ÇIRALIK, MD[c]

Departments of [a]Pediatrics,
[c]Pathology, Faculty of Medicine,
Kahramanmaras Sutcuimam
University, Kahramanmaras
Department of [b]Dermatology,
Faculty of Medicine,
Trakya University, Edirne

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Yazışma Adresi/*Correspondence:*
Ozer ARICAN, MD
Trakya University, Faculty of Medicine,
Department of Dermatology, Edirne
TÜRKİYE/TURKEY
ozerari@hotmail.com

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Histiocytic skin diseases are very rare in children and are usually classified as either Langerhans cell histiocytosis (LCH) or non-LCH, based on the pathology of the proliferating histiocytes. Bening cepholic histiocytosis (BCH), juvenile xanthogranuloma (JXG), xanthoma disseminatum (XD), indeterminate cell histiocytosis, necrobiotic xanthogranuloma, and generalized eruptive histiocytomas (GEH) are the main types of non-LCH.[1]

BCH is a rare, benign, self-healing cutaneous condition of young children. The etiology of BCH is unknown.[1,2] Since its description by Gianotti et al in 1971, about 40 known cases have been described in the literature worldwide. The clinical findings consist of usually asymptomatic, small (2 to 8 mm in diameter), yellow-brownish macules and papules, slightly raised, round or oval, localized first on the upper part of the face, mainly around the eyelids, forehead, and cheeks. The lesions are present at ages ranging from 2-34 months, and there is an equal incidence in males and females. Serum lipids are normal and the mucous membranes, soles, palms and internal organs are not involved.[2,3]

By light microscopy, lesions of BCH show a well-circumscribed histiocytic infiltrate within the upper and middle dermis without epidermal invasion. Scattered or grouped lymphocytes and a few eosinophils are found among the histiocytes. Histiocytes can be uniform or pleomorphic, with oval to reniform nuclei and occasional prominent nucleoli. There is abundant pale to slightly amphophilic cytoplasm without cytoplasmic lipids. Foamy cells or Touton giant cells are not seen. On immunohistochemical staining, the Langerhans cell markers CD1a and S-100 protein are negative whereas the macrophage/histiocytic marker (CD68) is positive.[3,4]

The differential diagnosis of BCH with other diseases is sometimes difficult, particularly with plane warts, molluscum contagiosum (MC), Spitz nevi, JXG, LCH, GEH, XD, urticaria pigmentosa, and cutaneous sarcoidosis.[3,4]

MC, plain warts and Spitz nevi are easily distinguished on physical and histological examination. MC and warts are usually self-limited benign epidermal eruptions resulting from viral infections of the skin. The lesions of MC may be found anywhere on the body in children. At the top of each lesion, there is usually an opening through which a distinctive small, white cone can be seen. They often have an umbilicated central depression called a punctum. Plain warts may also occur anywhere but especially on the hands, limbs and face. The lesions are flesh colored or pigmented, well defined, very slightly raised, and flat-topped. Their surface is smooth or very slightly roughened. The lesions of MC and warts can spread in linear patterns by self-scratching (autoinoculation).[5] Spitz nevus is usually a reddish-brown, smooth surfaced and firm papule. It is typically solitary and it most often appears during the first two decades of life. A biopsy is usually obtained in such cases. Histologically, Spitz nevus is distinct and the presence of spindle and epithelial nevus cells are characterized.[1]

BCH can be differentiated from JXG, in which the nodules are pleomorphic and may be disseminated over the entire body and extra-cutaneous involvement can occur. Histologically, abundant foamy cells and Touton giant cells can be seen in JXG. These findings are not seen in BCH. LCH is distinguishable because its papular lesions spread to the scalp and the trunk. It may involve multiple organs including the eyes and bones and there may be other systemic symptoms such as fever and malaise. Langerhans cell histiocytes are stained positively for S100 and CD1a, and electron microscopy confirms the diagnosis (ultrastructurally Birbeck granules are present).[3,4] There was no extracutaneous involvement in our patient.

It is also difficult to make a differential diagnosis for GEH. It is distinguishable because of the age of onset (mainly adults) and the sites involved (mucosal lesions). It has a more extensive distri-

TABLO 1: Clinical and histopathological features of plain warts, molluscum contagiosum (MC), benign cephalic histiocytosis (BCH), juvenile xanthogranuloma (JXG), generalized eruptive histiocytomas (GEH) and Langerhans cell histiocytosis (LCH).

Disease	Plain warts	MC	BCH	JXG	GEH	LCH
Age of onset	Every age	Childhood, ubertal age	Infancy	Infancy	Mainly adults	1-5 years of age
Localization	Anywhere	Anywhere	Head, face, neck, upper trunk	Disseminated	Trunk, extremities, extensive	Scalp and the trunk
Lesions	Flesh colored o pigmented, well-defined, smooth surface	Umbilicated papules or nodules	Macules, papules	Papulonodular	Papules	Papules
Mucous membranes	No	No	No	No	Yes	No
Visceral involvement	No	No	No	Sometimes	No	Multiple organs
Histopathology	Intracytoplasmic eosinophilic keratohyalin-like granules	Hyaline acidophilic granular masses (molluscum bodies)	Histiocytic infiltrate, S-100 negative, CD 68 positive	Foam cells, Touton giant cells	Histiocytic infiltrate, S-100 negative	S-100 and CD 1a staining positive
Systemic symptom	No	No	No	No	No	No
Course	Self-healing	Self-healing	Self-healing	Self-healing	Self-healing	Self-healing

bution of lesions with occasional mucosal lesions. In histological examination, giant cells are not observed and foamy cells are sometimes seen. It is easy to make a differential diagnosis for XD. It was described predominantly in adult patients. Histologically, there are cytoplasmic lipids or giant cells in the biopsy specimen. Lesions of the nodular type of urticaria pigmentosa are distinguishable because of the wheal that follows rubbing (positive Darier's sign); histology will reveal a mast cell infiltrate. Cutaneous sarcoidosis can be differentiated histopathologically by its islands of epitheloid cells permeated and surrounded by reticulum fibers.[1]

Our patient did not demonstrate the clinical or histological features of the abovementioned diseases. The differential diagnosis of significant diseases from BCH is also summarized in Table 1.

We present here a 3-month-old girl with BCH. In our patient, the histopathological and clinical findings revealed the diagnosis of BCH. Spontaneous regression of the eruption is the rule in BCH. The lesions begin to regress after a variable length of time ranging from 8 to 48 months, and complete resolution may take up to 54-72 months.[1-3] To date she remains well, this is a further confirmation of the diagnosis of BCH.

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