

Benign Cephalic Histiocytosis

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Abstract

Observation: Histiocytosis is the name of a group of diseases affecting skin and visceral organs with a pathogenesis not fully understood. Histiocytosis classified in two groups based on pathological examination: Langerhans cell histiocytosis(LCH) and non-LCH. Benign cephalic histiocytosis(BCH) belongs to non-LCH group and it is a rare skin disorder characterized by yellow, red-to-brown multiple small papules occurred on scalp, face, neck and superior part of body. Histopathological examination of BCH shows histiocytes at superior and mid dermis without any epidermotropism. Our case was a girl at 5 months of age and yellow-red papules with a diameter of 0,3-0,6 were present on the face and auricle, predominantly on the scalp. BCH diagnosis was made by clinical and histopathological examination

Introduction

Benign cephalic histiocytosis(BCH) is a rare skin disorder belonging to non-LCH group and affecting infants and children without involving visceral organs except rare cases with not fully understood reason. Firstly, Gianotiet al. described this disorder in 1971 [1]. BCH is characterized by yellow, red-to-brown multiple small papules localized on superior part of body and head and neck. This disorder is asymptomatic and self-limiting. Histopathological ultrastructure examination shows histiocytes proliferating at superior and mid dermis and comma-shaped bodies and desmosomelike structures [2]. This histological appearance is not characteristic for BCH. These signs can be also found in other disorders of non-LCH group such as juvenalexanthogranuloma(JXG) and generalized eruptive histiocytoma (GEH) [3]. Differential diagnosis of these disorders is based on ad-

ditional histological and clinical signs. In this case, BCH was clinico-pathologically diagnosed in a girl at 5 months of age.

Case Report

A girl at 5 months of age presented to our clinic with yellow-red papules with a diameter of 0,3-0,6 on scalp, face and auricle. Patient history showed that these lesions were initiated on scalp 2 months before. Infant development was normal. Weight and height were within normal percentile. Physical examination revealed lesions localized predominantly on the scalp. There was also a papule on right auricle (Figures 1a, b, c and d). No lesion was found on the body and upper and lower extremities. Lymphadenopathy and organomegaly were not detected. Routine blood tests, hematological and serum chemistry parameters were within normal limits. There was no anomaly on blood smear test. Transfontanel USG showed normal structures.



Figures 1 a ,b, c, and d. Papulonodules localized predominantly on the scalp and auricle

Histological examination of biopsy material showed normal epidermis and diffuse histiocytes infiltration in dermis. There were partly eosinophils in dermis. Mitotic figures, multinuclear giant cells and cytoplasmic lipid were not present. Immunohistochemical analysis was negative for S100 and CD1a staining and positive for CD68(KP1/KIMP) staining. This staining pattern is typical for non-LCH. According to clinico-pathological findings, benign cephalic histiocytosis, a type of non-LCH, was diagnosed in patient.

Patient was monitored without treatment, as BCH does not require any treatment and regresses spontaneously.

Discussion

BCH is a rare disorder belonging to non-LCH group and its etiology is not fully understood. Clinical localization is limited to head and neck, except rare cases and it is characterized by self-limited eruptions. There eruptions are macules and papules with a diameter of 1-8 mm and the color was from yellow to red-brown. Majority of cases are asymptomatic. In certain rare cases, visceral organ involvement has been reported. This disorder is usually seen in infants at 2 to 34 months of age (mean, 15 months). Girls and boys are equally affected. Self-regression of lesions is seen between 8 and 48 months. Scars do not develop on lesion localization, however hyperpigmentation may sustain for long period [2].

Histopathological examination shows an infiltrate of histiocytic cells at papillary and reticular dermis. These cells do not contain cytoplasmic lipid and they have large cytoplasm, polymorphic nucleus and pale chromatin. Lymphocytes and rarely eosinophils also may accompany. At electron microscopy, the occurrence rate of coma shaped bodies with incytoplasm of histiocytosis cells is 5% to 30%. Birbeck granules seen in Langerhans cells are not present in these cells. Immunohistochemical examination shows negative S100 and CD1a, which are positive in Langerhans cells. However these cells have positive factor 13a and CD68 [4].

In differential diagnosis of BCH, JXG and GEH are at the top of list. Besides them, differential diagnosis can be also considered for verruca plana, urticaria pigmentosa, lichenoid sarcoidosis and multiple Spitz nevus [5]. Verruca plana, urticaria pigmentosa and multiple Spitz nevus are easily differentiated by histological examination. However in JXG, the disorder is not limited to head and neck and it can be spread to all body. Also extra-cutaneous involvement is more frequent. GEH is more frequent in adults and involves large body area including mucosal tissues. Clinical and histological differentiation of LCH is easier. In LCH, S100 and CD1a are positive. Birbeck granules can be seen in cell cytoplasm [6].

Recent studies and case reports suggest that BCH can clinically and histopathologically progress to JXG and GEH [7]. In addition, there are case reports indicating the regression of JXG and GEH to BCH. In their study, Gianotti et al. indicated that histopathological examination in 50% of cases couldn't precisely differentiate from early non-xanthomatous JXG and GEH [8]. The relationship between BCH and JXG and GEH is not clear. Therefore, certain authors consider BCH as a localized form of JXG and GEH, not a separate disorder. In conclusion, more studies and scientific knowledge is needed in order to understand the relationship between disorders of non-LCH group and their pathogenesis.

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