CASE REPORT

Benign Cephalic Histiocytosis: A Rare Dermatological Entity in the Paediatric Population

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Summary

Histiocytic proliferation of the skin can be categorised into Langerhan and non-Langerhan types. Benign cephalic histiocytosis (BCH) is a rare cutaneous non-Langerhan histiocytosis typically affecting infants and young children. We report a case of benign cephalic histiocytosis in a boy who, at seven months of age, presented with multiple yellowish papular eruptions on his face. Over the course of 18 months, the lesion spread to his trunk and upper extremities, sparing the mucous membranes, palms and soles. There was no systemic involvement identified. A histopathologic examination of the skin lesion showed diffuse infiltration of histiocytes within the superficial dermis intermingled with scattered eosinophils and small lymphocytes. Immunohistochemical studies showed that the histiocytes were diffusely positive for CD68. Langerhan markers CD1a and S100 were negative. The correct distinction between BCH and other histiocytic proliferations of the skin is important because BCH has a self-limiting clinical course with a tendency of spontaneous remission.

Key words: *Benign cephalic histiocytosis (BCH), histiocytic proliferation, cutaneous histiocytosis*

Introduction

Cutaneous histiocytic proliferation is typically categorised into Langerhan and non-Langerhan types. Benign cephalic histiocytosis (BCH) are rare, non-Langerhan, self-limiting cutaneous lesions in children. Sixty cases of BCH have been reported in English literatures. [1] To the best of our knowledge, this study is the first published occurrence of BCH in Malaysia.

Case Report

A two-year-old boy exhibited multiple skin rashes, appearing on his face at seven months old. Gradually, over 18 months, the rash spread to his trunk and bilateral upper limbs. There was no history of bone pain, increased urination or constitutional symptoms.

A clinical examination showed multiple papular lesions of varying sizes, measuring up to 5 mm in diameter (Figure 1). These papules ranged from yellow-orange to brown, predominantly on the

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head, neck, trunk and proximal upper extremities (Figure 1). There was an absence of organomegaly and lymphadenopathy, and the serum lipid profile and full blood count were normal. Previous clinical differential diagnoses included benign cephalic histiocytosis (BCH), juvenile xanthogranuloma (JXG), and generalized eruptive histiocytosis (GEH).

Figure 1. (a) – (b) Multiple yellow-orange to brownish papules predominantly over the face, extending to trunks

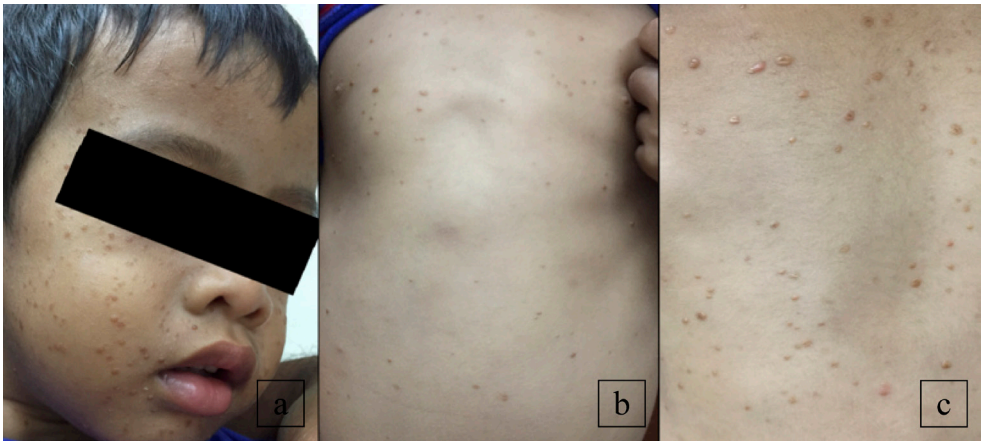
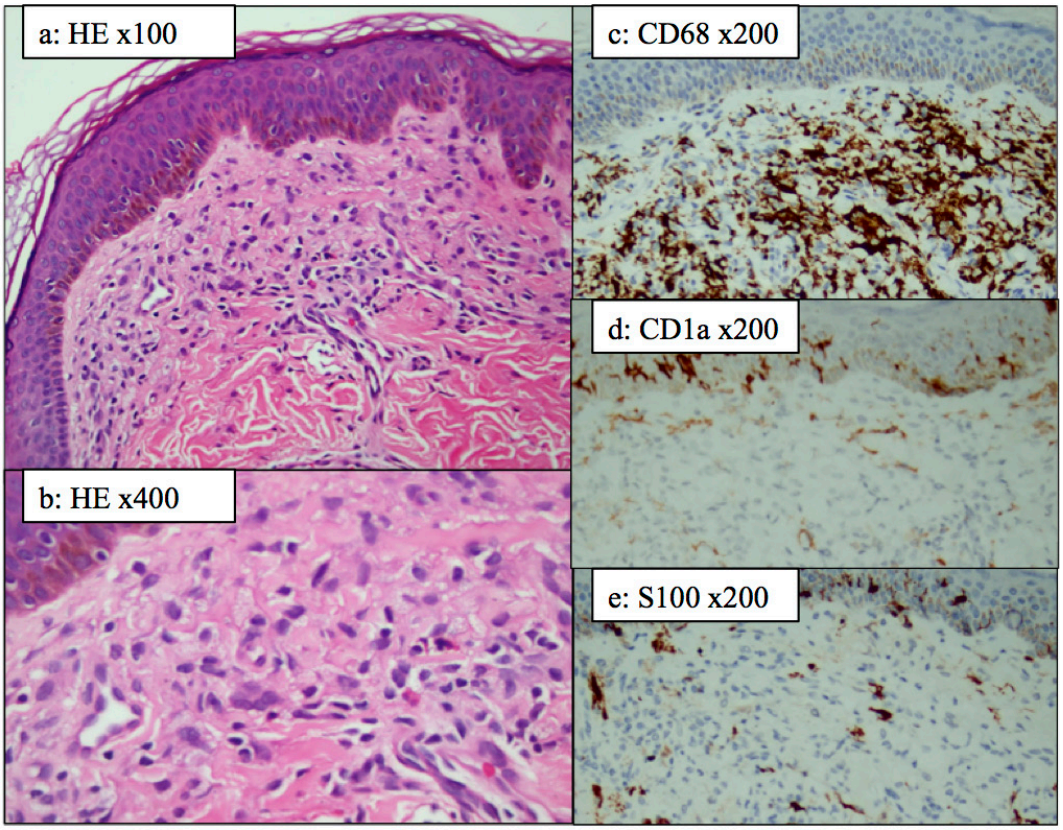


Figure 2. (a) Diffuse histiocytic infiltrate at the upper dermis. **(b)** The histiocytes displayed oval kidney-shaped nuclei and ill-defined pale cytoplasm. **(c)** The histiocytes strongly expressed CD68 and **(d & e)** were negative for Langerhan markers CD1a and S100 protein.



Histopathological examination of the skin biopsy (Figure 2: a & b) revealed diffuse infiltrate of histiocytes within the superficial epidermis, with scattered, small lymphocytes and rare eosinophils. Nuclear atypia was minimal, with no mitotic activity. There were no cytoplasmic lipids or Touton cells, which supported the diagnosis of BCH instead of JXG. The histiocytic expression of CD68 and the absence of Langerhan marker CD1a and S100 protein were also in keeping with BCH (Figure 2: c-e).

A diagnosis of BCH was made based on clinical history, morphology and immunohistochemistry.

The parents were informed of the benign condition, requiring no active treatment. At a six-month follow up, signs of spontaneous regression were noted over the face and trunk.

Discussion

BCH is characterised by eruptions of asymptomatic, red-brown, maculopapular lesions measuring from 2 mm to 8 mm in diameter[2]. In more than 90% of cases, the initial eruption appears on the face and spreads to other body parts with predilection to the head, neck and upper extremities.[2,3] BCH is strictly limited to the skin[3], with no effect on mucocutaneous sites or deep organs.[2]

The age of onset ranges from two to 66 months (average 15 months).[3] With a natural self-healing course, spontaneous regression begins as early as eight months from the onset but may take up to 48 months to disappear[3]. Complete regression has been noted at an average of 50 months.[3] Signs of spontaneous regression observed in our patient further confirmed the diagnosis of BCH.

Two important differential diagnoses of dermal non-Langerhan histiocytosis are juvenile xanthogranuloma (JXG) and generalized eruptive histiocytosis (GEH).[1,2] JXG typically affects young children and exhibits similar papular lesions affecting the head and neck region, but it may occur on extremities and trunk.[4] JXG is also associated with rare extracutaneous involvement, particularly in the eye, which may lead to secondary glaucoma and blindness.[4] Internal organs may also be affected.[4] GEH is more common among adults and has a more widespread and symmetrical skin distribution with occasional mucosal involvement.[5] There were overlapping clinical features of JXG and BCH observed in our patient, but extracutaneous involvement was absent. Lesions were not only confined to the head and neck but also spread to the trunk and upper extremities. Histopathological examination confirmed the diagnosis of BCH.

BCH, JXG and GEH at early nonxanthomatous stages cannot be differentiated based on histology or immunohistochemistry alone.[2] All three conditions show dense dermal infiltrations of histiocytes, which are negative for Langerhan cell markers.[2,3] Morphological features favouring JXG are the presence of foamy macrophages and Touton giant cells.[2] The absence of Touton giant cells and foamy histiocytes are essential in excluding JXG in this case, as the other morphological features can be similar to BCH.

A diagnosis of BCH implicates a benign, self-limiting condition, requiring no treatment[2]. However, an association with diabetes insipidus has been reported in one child,[6] and there have also been reports of possible progression to JXG.[7,8]

Conclusion

This case illustrates classic clinical and histopathological presentations of BCH. Proper attention to history, examination of findings, histological examination, immunohistochemistry and follow up are important for a correct diagnosis.

Declaration of Conflict of Interest

None declared

Acknowledgement

We would like to thank the Director-General of Health Malaysia for his permission to publish this study. This research was approved by the Medical Research and Ethics Committee, Ministry of Health Malaysia (NMRR-19-7-45787).

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