Case Report

Congenital Self-Healing Reticulohistiocytosis Presenting as Hypopigmented Macules and Papules in a Neonate

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Congenital self-healing reticulohistiocytosis, also known as Hashimoto-Pritzker disease, is a rare variant of Langerhans cell histiocytosis. It is characterized by developing multiple or single red-brown papules, nodules or vesicles at birth or first few weeks of life and involutes spontaneously without systemic involvement of other organs. We report a rare case of female neonate presented with multiple white macules and papules on trunk. Langerhans cell histiocytosis is diagnosed by histological examination and immunohistochemical study. We remind dermatologists to be aware of this rare presentation and should put it into differential diagnosis lists when facing hypopigmented lesions in a neonate. (Dermatol Sinica 26: 80-84, 2008)

Key words: Langerhans cell histiocytosis, Congenital self-healing reticulohistiocytosis, Hashimoto-Pritzker disease, Hypopigmentation

INTRODUCTION

Congenital self-healing reticulohistiocytosis (CSHRH), a rare form of Langerhans cell histiocytosis, is typically presented at birth or in the newborn period with spontaneous regression. It is usually characterized by the eruption of multiple, disseminated, red-brown papules and nodules which may increase in size and number during the first few weeks of life without systemic signs. Skin lesions initially manifested by depigmented or hypopigmented macules and papule had never been described before. We present a case of female neonate presented with multiple white macules and papules on trunk, which may mimic other hypopigmented diseases clinically.

CASE REPORT

A 7-month-old girl in our clinic was noted to have progressive white cutaneous eruption on her back, abdomen and chest. The patient’s mother said the eruption had been present one month after birth and was asymptomatic. There were no preceding papules or vesicles before the eruption. The patient had an unremarkable past medical history and no family history of similar lesions. The baby was otherwise healthy without fever, weight loss, dyspnea, mucosal involvement, hepatosplenomegaly and lymphadenopathy. On physical examination, multiple small round scaleless white macules and papules were distributed on

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her back, abdomen and chest. (Fig. 1) There is no lesion on the extremities and head. The margins of the lesion is irregular. (Fig. 2)

A skin biopsy from the papule revealed hyperkeratosis, focal parakeratosis, mild acanthosis, and histiocytes in papillary dermis and in epidermis. Occasional eosinophils are also seen. (Fig. 3) Immunohistochemical stains showed that the histiocytes were positive for S-100 and CD1a (Fig. 4) and confirm the diagnosis of Langerhans cells histiocytosis.

The patient was referred to pediatric hematologist for further survey and revealed only cutaneous involvement. The skin lesions completely regressed spontaneously 3 months later and did not recur in one and half year follow-up.

DISCUSSION

Langerhans cell histiocytosis is a disease of unknown cause characterized by a clonal proliferation of S-100 and CD1a positive activated Langerhans cells. Four main forms were included: Letterer-Siwe disease, Hand-Schüller-Christian disease, eosinophilic granuloma, and Hashimoto-Pritzker disease (congenital self-healing reticulohistiocytosis). In neonates, cutaneous changes were the most common initial manifestation in Langerhans cell histiocytosis.

CSHRH is a rare variant of Langerhans cell histiocytosis, typically presents at birth or first few weeks of life and involutes spontaneously with no systemic involvement. It is characterized by multiple red, violaceous, or brownish, firm, painless papules and crusts scattered all over the face, scalp, trunk, limbs, palms and soles of the feet, and some patients may have a vesicular eruption, or sparse nodules.

Our patient presented skin eruptions without lesions of the mucous membrane and no systemic symptoms. Histopathologically, infiltrations of Langerhans cells in papillary dermis and in epidermis with occasional eosinophils were seen. The skin lesions disappeared spontaneously after 8 months and did not reappear during one and half year’s follow-up. In view of the histology, dermatological lesions and evolution, the diagnosis of CSHRH should be considered.

Noteworthily, our patient initially manifested with multiple small round white macules and papules on her back, abdomen and chest, which have never been reported before. Although firm, red-brown, painless, papulonodules or vesicles and crusts followed by residual hypopigmented or hyperpigmented scar had been mentioned in previous article, in our patient, according to her mother’s description, no such previous episode was noted. Besides, skin biopsy of the hypopigmented papule revealed active disease activity, instead of residual scar or only pigment changes. However, the possibility of unawareness of previous skin lesions by her families could not be totally excluded.

There are several differential diagnoses for multiple hypopigmented macules and papules in a neonate, such as clear cell papulosis, pityriasis versicolor, nevus depigmentosus, vitiligo, pityriasis alba, post inflammatory hypopigmentation, hypomelanosis of Ito, and incontinentia pigmenti.

Clear cell papulosis are hypopigmented macules or barely palpable papules following the milk line on anterior trunk. They only rarely developed on back. Pityriasis versicolor can be excluded by potassium hydroxide examination. Nevus depigmentosus is generally known to be a congenital, nonfamilial disorder characterized by hypopigmented lesion that remains stable in size and distribution throughout life. Most patients with nevus depigmentosus had one lesion or at least less than 10 lesions. Vitiligo is an acquired disorder with uniformly milk- or chalk-white color with discrete margin. The natural course of vitiligo is unpredictable, but often shows abrupt onset, followed by progression for a time, and then a period of stability. Pityriasis alba is characterized by
the presence of ill-defined, hypopigmented, round-to-oval, scaling patches on the face, upper arms, neck, or shoulders. The legs and trunk are less commonly involved. Post-inflammatory hypopigmentation can be excluded by previous history of previous inflammatory conditions or medication. Hypomelanosis of Ito shows a familial tendency and is usually associated with systemic abnormalities. It is characterized by hypopigmented patches in streaks and whorls. Incontinentia pigmenti is an X-linked dominant neurocutaneous syndrome with frequent systemic involvement. Typically, the cutaneous findings generally progress through 4 distinct characteristic stages. By histopathological examinations, diagnosis of Langerhans cell histiocytosis could be easily made.

In conclusion, due to the diversity of the clinical manifestation and the morphological similarity to other entities, cutaneous Langerhans cell histiocytosis could be a diagnostic challenge. We report a rare case of congenital self-healing reticulohistiocytosis manifested.

Fig. 1
Progressive white cutaneous lesions were noted on back, abdomen and chest.
There was no lesion on extremities and head.

Fig. 2
Close-up view revealed multiple small round scaleless white macules and papules with irregular margin.

Fig. 3
Skin biopsy showed hyperkeratosis, focal parakeratosis, mild acanthosis, and histiocytes in papillary dermis and in epidermis. Occasional eosinophils were also seen in other area. (H&E, original magnification x400)

Fig. 4
Immunohistochemical stains showed that the histiocytes were positive for CD1a (left) and S-100 (right). (Original magnification x200)
by multiple small round white macules and papules on trunk in a neonate. We remind dermatologists to be aware of this rare presentation and when multiple hypopigmented or depigmented macules or papules were noted in a neonate. Langerhans cell histiocytosis should be put into differential diagnosis lists with systemic survey and long-term follow up if pathology confirms the diagnosis.

REFERENCES
以白色斑疹及丘疹為表現的先天性自癒性網狀組織細胞增生症

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先天性自癒性網狀組織細胞增生症是蘭格罕氏組織細胞增生症中罕見的一種型態。其特徵是在剛出生或出生數週內在皮膚產生多個或單一紅棕色丘疹、腫塊或水疱而不侵犯其他器官或系統，且會自行痊癒。我們報告一位女嬰以白色斑疹及丘疹呈現於軀幹為表徵。藉由病理檢查及免疫組織學檢查證實為蘭格罕氏組織細胞增生症。我們提醒皮膚科醫師注意先天性自癒性網狀組織細胞增生症之罕見表現方式：當嬰兒身上呈現白色病灶時，應將本病列入鑑別診斷。（中華皮誌：26: 80-84, 2008）