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Linear and whorled nevoid hypermelanosis in a Malaysian Chinese girl

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Abstract

Linear and whorled nevoid hypermelanosis is a rare sporadic skin disorder characterized by swirls and streaks of macular hyperpigmentation along lines of Blaschko. It is associated with extracutaneous features and has poor treatment outcome. A case of possible linear and whorled nevoid hypermelanosis is reported in a 5 year old Malaysian Chinese girl.

Introduction

Linear and whorled nevoid hypermelanosis is a rare skin disorder characterized by swirls and streaks of macular hyperpigmentation along the Blaschko line and usually appears within the first 2 years of life [1]. A case of possible linear and whorled nevoid hypermelanosis in a 5 years old Malaysian Chinese girl is reported.

Case synopsis

A 3 years old Chinese girl with normal developmental milestones was referred to us in 2006 for whorled and linear hyperpigmentation along the Blasckho's lines, predominantly left sided ***(Figure 1)***. She was born full term to a non-consanguineous parent and started to have skin lesions at 2 months of age. Retrospective review failed to elicit vesicular or verrucous lesions preceding the linear hyperpigmentation. She had normal developmental milestones without dental, ophthalmologic or neurological abnormalities. No family history was elicited and she had 2 normal male siblings (2). No skin biopsy was done. Her blood eosinophil count was normal.

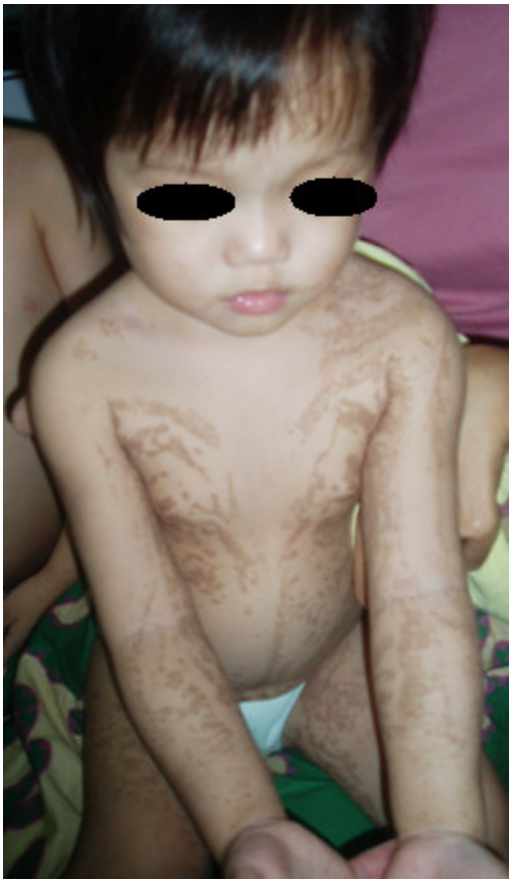

Fig 1: Linear and whorled pigmentation along the lines of Blaschko

Fig 2: Figure 2: The patient with her unaffected mother and 2 older brothers

Discussion

Linear and whorled nevoid hypermelanosis usually starts within the first few weeks of life with progression during the first 2-3 years of life before stabilizing [1]. Patients typically present with reticulate hyperpigmented macules that coalesce to form streaks and whorled hyperpigmentation following the lines of Blaschko [2]. The trunk, extremities, neck, face and genitalia are the typical sites affected [2,3,4]. The patient presented here showed the typical pattern and distribution of the skin lesions.

It is important to exclude other pigmentary disorders following the Blaschko lines before making a diagnosis of linear and whorled nevoid hypermelanosis. The differential diagnoses include incontinentia pigmenti, linear epidermal nevus, hypomelanosis of Ito and Goltz syndrome. Incontinentia pigmenti, an X linked dominant genodermatosis seen almost exclusively in females, usually presents with an initial inflammatory vesicular stage and later a verrucous proliferative stage before the hyperpigmented stage [5].. Linear epidermal nevus, on the other hand will present with prominently elevated and verrucous lesion with time after a hyperpigmented stage in infancy [1]. Hypomelanosis of Ito, as the name implies presents as linear hypopigmentation following the lines of Blaschko. Goltz syndrome or focal dermal hypoplasia, another X linked dominant disorder, commonly presents with reddish tan, atrophic, linear verrucous lesions along the lines of Blaschko. Telangiectases are commonly seen. The patient presented had only hyperpigmented lesions since 2 months old without any vesicular and verrucous lesions, suggesting a diagnosis of linear and whorled nevoid hypermelanosis.

The histopathological changes of linear and whorled nevoid hypermelanosis consists of diffuse moderate hyperpigmentation in the basal layer and lack of pigmentary incontinence in the dermis [4,6]. However, due to the parent refusal, a skin biopsy was not done in the patient presented.

Linear and whorled nevoid hypermelanosis can be associated with neuro-developmental and growth retardation, ocular anomalies, skeletal anomalies and congenital heart diseases [3,6]. It is also associated with chromosomal mosaicism [6]. In the patient presented, she did not have any extracutaneous presentation of the disorder. Chromosomal study was not done here.

No effective treatment is available for this disorder. Chemical peels [3] and depigmenting agents [6] have been tried without much success. Future laser techniques and cosmetic camouflage offer some promise.

References

1. Kalter DC, Griffiths WA, Atherton DJ. Linear and whorled nevoid hypermelanosis. J Am Acad Dermatol. 1988; 19(6): 1037- 1044. PMID: 3204178

2. Harre J, Millikan LE. Linear and whorled pigmentation. Int J Dermatol. 1994; 33: 529- 37. PMID: 7960343

3. Alvarez J, Peteiro C, Toribio J. Linear and whorled nevoid hypermelanosis. Pediatr

Dermatol. 1993; 10: 156- 8. PMID: 8346111

4. Yuksek J, Sezer E, Erbil AH, et al. Linear and whorled nevoid hypermelanosis. Dermatology Online Journal. 2007; 13 (3): 23. PMID: 18328217

5. Leong KF, Pubalan M, Yap FBB. Incontinentia pigmenti: report of 3 cases from Sarawak. Malaysian Journal of Dermatology. 2008; 21: 113- 5.

6. Hong SP, Ahn SY, Lee WS. Linear and whorled nevoid hypermelanosis: unique clinical presentations and their possible association with chromosomal abnormality inv (9). Arch Dermatol. 2008; 144(3): 415- 6. PMID: 18347306