

Development of lentigines and melanocytic nevi within segmental nevus depigmentosus: A case report.

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ABSTRACT:

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Nevus depigmentosus (ND) is an uncommon congenital nonprogressive, hypopigmented macule or patch that remains stable in size and distribution throughout life, resulted from a defect of melanosomes transfer from melanocytes to keratinocytes. Multiple lentigines or pigmented nevi occurring within nevus depigmentosus are considered a rare phenomenon. Somatic revertant mosaicism may be the plausible explanation. We report a 12-year-old girl who presented with a hypopigmented patch on the right neck and shoulder since birth. There has been a progressively increasing in number of macules and patches that varied from light to dark brown within the area of leukoderma for six years. Histopathology from a dark brown spot showed elongated rete ridges with slightly enlarged nevus cells and mild lamellar fibrosis, which were consistent with dysplastic nevus.

Key word: Nevus depigmentosus, lentigines, melanocytic nevi

บทคัดย่อ:

เพชรรัตน์ นิตสารัตน์, จักรพงษ์ ขุนหะเสวี รายงานผู้ป่วยที่มีการเกิดกระและไฝบนปานขาว (NEVUS DEPIGMENTOSUS) วารสารโรคผิวหนัง 2557; 30: 136-140.

ปานขาว (nevus depigmentosus) เป็นปานสีขาวพบได้ตั้งแต่แรกเกิด พบบ่อยบริเวณลำตัว ขนาดของปานขาวนั้นมักจะคงที่ไม่ขยายขนาด สาเหตุเกิดจากความผิดปกติของการส่งต่อเม็ดสีจากเซลล์สร้างเม็ดสีไปยังเซลล์ผิวหนัง การเกิดกระหรือไฝบนปานสีขาวนั้นพบได้น้อยเชื่อว่าเกิดจาก somatic revertant mosaicism รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงอายุ 12 ปี ซึ่งมีปานสีขาวตั้งแต่เกิดบริเวณคอและไหล่ขวา เริ่มมีผื่นราบสีน้ำตาลอ่อนและเข้มขึ้นบนบริเวณปานสีขาวมากขึ้นเรื่อยๆ เป็นเวลา 6 ปีก่อนมาพบแพทย์ลักษณะทางพยาธิวิทยาของชิ้นเนื้อจุกน้ำตาลเข้มพบ dysplastic nevus ได้รับการวินิจฉัยว่ามีทั้งกระและไฝเกิดขึ้นบนปานสีขาว

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คำสำคัญ: ปานขาว, กระ, ไฝ

Case report

A 12-year-old Thai girl, otherwise healthy, presented with asymptomatic multiple light and dark brown macules and patches within a hypopigmented patch on the right neck extending to right shoulder for 6 years. The parents noted the hypopigmented lesion has been present since birth, gradually enlarged and remained stable by the age of 3. By the age of 6, multiple light brown macules and patches and two dark brown spots were noted within the leukoderma area with gradually increased in number. None of the brown macules lightened or darkened in response to sun exposure. Palm, soles, eyes and mucous membrane were spared. Psychomotor and musculoskeletal developments were normal. No family history of similar cutaneous findings or melanoma.

Physical examination revealed a well defined, serrated and irregularly bordered, hypopigmented patch,

measuring 16 x13 cm in size on the right neck and shoulder. Within the area of segmental leukoderma, several hyperpigmented macules that varied from light brown to dark brown measuring 1-5 mm in diameter and a light brown patch measuring 7x4 cm in size, with irregular border for most of the lesions were noted (Figure 1). They were not perifollicular. Under Wood's lamp examination, area of leukoderma revealed an off-white accentuation. No evidence of limb asymmetry, musculoskeletal or neurological abnormalities were detected.

Histological examination of a dark brown macule biopsied from the right shoulder showed elongated rete ridges with slightly enlarged nevus cells and mild lamellar fibrosis, which were consistent with dysplastic nevus (Figure 2).



Figure 1 Several light and dark brown macules and patches on a serrated leukoderma background on the right neck and shoulder.

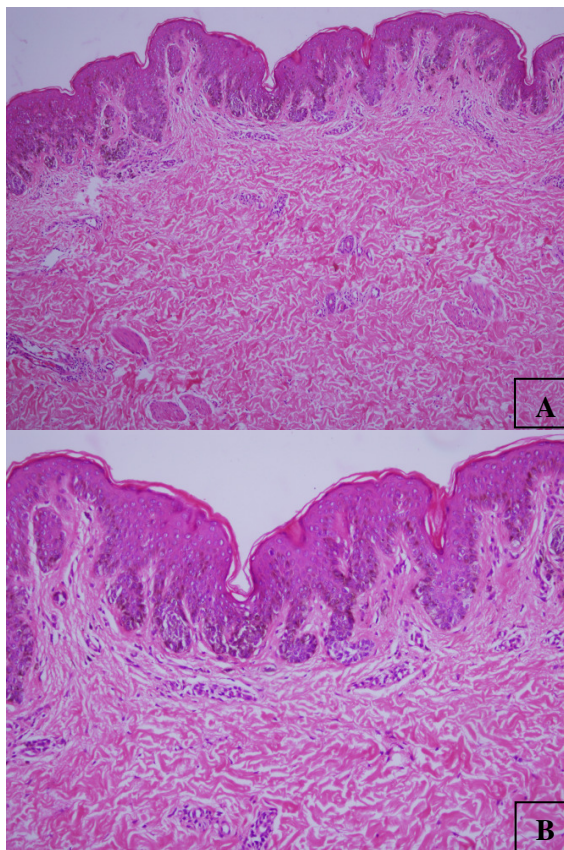


Figure 2 Dysplastic nevus (A: H&E10X, B: H&E20X)

Discussion

Nevus depigmentosus (ND) is an uncommon congenital nonfamilial cutaneous hypomelanosis, first reported by Lesser in 1884¹, which characterized by a nonprogressive hypopigmented macule or patch with circumscribed irregular and serrated border. The lesion remains stable both in size and distribution throughout life. Localized, segmental or systematized pattern can be observed. ND occurs sporadically with no known patterns of inheritance or familial tendency². The pathogenesis of ND remains uncertain. The defect of melanosome transfer from melanocytes to keratinocytes^{1,2} and degeneration of melanosomes in keratinocytes may explain the pathogenesis³.

The clinical diagnosis based on Coupe's criteria⁴ proposed in 1976 includes (1) leukoderma is present at birth or early in life; (2) leukoderma remains stable throughout life; (3) no change in texture or sensation in the lesions; and (4) no hyperpigmented border around the affected area. Under Wood's lamp examination, the lesion shows an off-white accentuation. In contrast with ND, chalky-white accentuation is observed in vitiligo^{1,3}. Histological finding on lesional skin shows a marked reduction in the density of the melanosomes, but variable results in the number of melanocytes³. PUVA, excimer laser, and surgical grafting methods have been tried to repigment the lesions of ND. Repigmentation with blister roof grafting and cultured epidermal grafting has also been reported.

Recently, there have been eight reported cases of multiple lentigines within ND⁵ and three cases with melanocytic nevi within ND^{6,7}. Therefore, we presented a case of both lentigines and melanocytic nevi within segmental ND. This is a rare phenomenon. Bologna et al.⁸ considered to be the reversion of a mutation in one of the genes involved in pigmentation. Somatic revertant mosaicism has recently explained the phenomenon of congenital reticular ichthyosiform erythroderma, which is the coexistence of cells carrying inherited genetic mutations with cells that have undergone spontaneous changes that correct the mutant phenotype^{9,10}. Inactive cells may become functional to produce hypomelanotic contraction, which presents as a form of “repigmentation”^{6,9}. Bardazzi et al.¹¹ suggested that the melanocytes in ND can move from a functionally defective stage to an active stage that overproduces melanin granules in the lentigines after repeated exposure to ultraviolet B phototherapy. Therefore, functionally defective melanocytes can differentiate into melanocytic nevus cells in the ND. Hyperpigmented spot has also been observed within the hypopigmented patches of piebaldism which probably induced by sunlight, as a result of accelerated melanin production of the melanocytes that had been inactive¹².

Melanin serves as a photoprotection against UV radiation. The development of dysplastic nevus within the hypopigmented lesion in this patient may be caused by DNA damage from the functional alteration of melanocytes⁶. The prevalence of atypical nevus in the

general population ranging from 2% to 8% in different studies, but with a history of melanoma the frequency of dysplastic nevi is higher, ranging from 34% to 59%¹². Only about 20% to 30% of melanomas arise in a pre-existing nevus and atypical melanocytic nevi.

In conclusion, we present a case of both multiple lentigines and melanocytic nevi within segmental nevus depigmentosus in a Thai patient. The dysplastic nevus was developed within the lesion of ND, therefore every 3-12 month follow up is recommended in this patient^{13,14}. Any increasing in number or clinical atypia of nevi should seek for attention. Excision is recommended for lesion that is worrisome for melanoma. Avoidance of sun exposure is also encouraged as a preventive measure for developing pigmented nevi.

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