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Case Report

Linear and whorled nevoid hypermelanosis with depigmented macules

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Abstract Linear and whorled nevoid hypermelanosis (LWNH) is a benign pigmentary disorder characterized by hyperpigmented macules in a linear, whorled or streaky configuration, following the lines of Blaschko on the trunk and limbs. The soles, palms, face, and mucous membranes are spared. Depigmented macules and cafe au lait macules (CALMs) occurring in association with the whorls are not seen. Here, we report a case of LWNH with depigmentation and CALMs.

Key words
LWNH, CALMs, Linear and whorled nevoid hypermelanosis.

Case report

A 27-year-old male presented to the outpatient department with chief complaints of asymptomatic, dark colour skin lesions over the body since 9 years of age. The lesions first appeared as a few hyperpigmented streaks on the upper back and progressed to involve the trunk and upper limbs. Since the last 6 months, depigmented macules started appearing on the upper back and progressed to involve arms and thighs. There was no history of warty lesions or blisters prior to the appearance of these lesions. There was no history of neurological, skeletal, and ocular abnormalities.

Systemic examination was unremarkable. Cutaneous examination revealed hyperpigmented macules in a linear, streaky pattern forming whorls. Pigmented streaks displayed a 'V' shaped pattern over spine, 'S' shape or whorled pattern over the anterior and lateral aspect of the trunk and linear arrangement over the extremities. Few depigmented macules are seen on the upper back, arm and thigh. Association of cafe au lait macules (CALMs) is seen on the right shoulder blade.

Routine haematological and biochemical tests revealed no abnormalities. Histopathological evaluation of the pigmented macule on the back revealed mild elongation of rete ridges and hyperpigmentation of the basal keratinocyte with vacuolization of melanocytes.

Discussion

Linear and whorled nevoid hypermelanosis (LWNH) is a rare sporadic disorder of pigmentation characterized by hyperpigmented macules in a linear or whorled configuration. It was first described by Kalter *et al*. as a hyperpigmentation composed of homogenous colored macules forming reticulated configurations.[1]

Lesions are distributed mainly on the trunk and extremities, sparing the palms, soles, and mucosae. The usual age of the onset of hyperpigmentation is within the first few weeks

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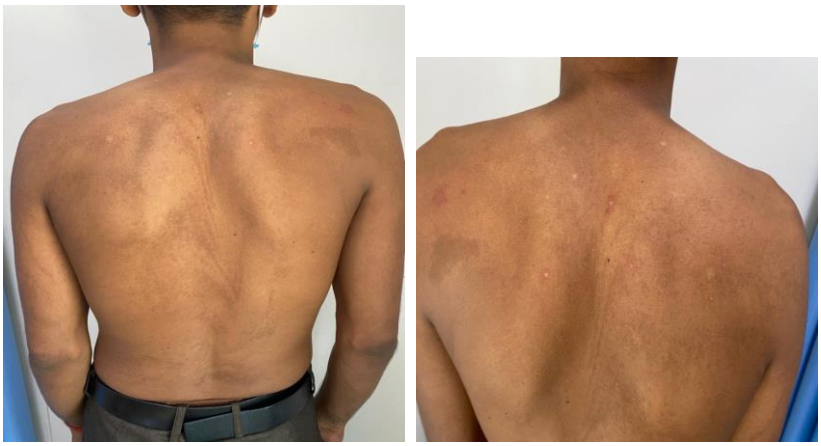

Figure 1 Linear and whorled nevoid hypermelanosis with Depigmented macules seen on the back of patient.

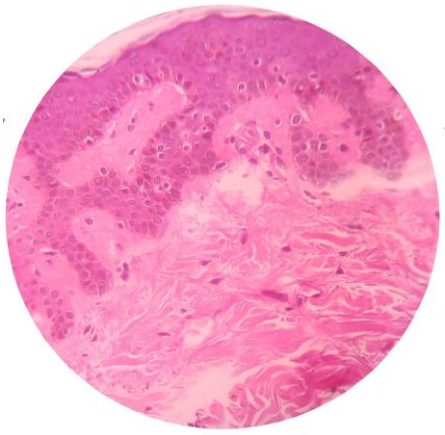

Figure 2 Histopathlogy (H&E, 40X) showing elongation of rete ridges and hyperpigmentation of the basal keratinocyte

of life, which continues to progress for a year or two before stabilization. There is no preceding inflammation or palpable lesions. Clinically, reticulated hyperpigmented macules coalescing to form streaks and whorled areas are seen over the trunk, extremities, and neck following the lines of Blaschko. Usually affected individuals have no accompanying extra dermal features. Few case reports have shown associated with cardiovascular, neurological, and musculoskeletal abnormalities.

Our patient presented with hyperpigmented lesions since childhood without any vesicular and verrucous lesions. The clinical picture was suggestive of LWNH. There were associated depigmented macules and CALMs with no systemic involvement.

Histopathological findings revealed skin biopsy revealed mild elongation of rete ridges and hyperpigmentation of the basal keratinocyte. Vacuolization of melanocytes can be seen.

Dermoscopy examinations showed a linear or circular arrangement of streak-like pigmentations arranged in a parallel manner.

Differential diagnosis of LWNH include incontinentia pigmenti, linear epidermal nevus, and hypomelanosis of Ito. Incontinentia pigmenti is an X linked dominant genodermatosis occurring almost entirely in females. It presents with an initial inflammatory vesicular stage and later a verrucous proliferative stage before the hyperpigmented stage. Epidermal nevi often become noticeable during infancy as streaks oriented along the lines of Blaschko, but with time they become papillomatous and hyperkeratotic. Hypomelanosis of Ito is typified by whorled and streaked bilaterally asymmetric leukoderma resembling "marble cake" or the reverse pattern of late hypermelanosis in incontinentia pigmenti.

Depigmented macules are usually not seen occurring in association with whorls.

In our case, depigmented macules were seen scattered on the upper back, arm, and thigh not following any distribution pattern .There was no associated with itching or scaling. No clinical signs of koebner phenomenon or any leukotrichia were visible. Explanation for the appearance is unclear. Literature search reveals unclassified or poorly classified generalized vitiligoid conditions like ‘Punctate vitiligo’.[2] It

refers to pea-sized depigmented macules that may involve any area of the body. If lesions do not coexist with classical vitligo, the term 'leukoderma punctata' is used. Other distinct conditions that may be difficult to distinguish from vitiligo clinically include idiopathic guttate melanosis and progressive macular hypomelanosis.[2,3]

Similar case was reported by Sinha *et al.* in which multiple pinpoint depigmented macules were seen along the areas of streaky macular pigmentation i.e. following lines of Blaschko.[4]

Explanation of the appearance of depigmented macules was unclear but they suggested "Mixed vitiligo of Blaschko lines". In this entity there is presence of segmental and non-segmental vitiligo in Blaschko linear pattern.[5]

The present case has been reported for its classical presentation, rarity, and unusual findings.

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