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**Photo Vignette**

**Linear and whorled nevoid hypermelanosis with dermatoscopic features**

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**Abstract**

Linear and whorled nevoid hypermelanosis (LWNH) is a rare sporadic disorder, characterized by hyperpigmented, reticulated, streaky, and whorled macules along Blaschko lines. Herein we report a 33-year-old male presenting with linear and whorled nevoid hypermelanosis. There was no family history. Dermoscopy revealed a net like pattern. This case is being presented for its rarity and for its dermatoscopic pattern.

**Key words: Blaschko lines, Net like pattern, Reticulate hyperpigmentation, Dermoscopy**

**Introduction**

Linear and whorled nevoid hypermelanosis (LWNH) is a rare sporadic disorder, first described by Kalter et al [1] in 1988 and characterized by hyperpigmented, reticulated, streaky, and whorled macules along Blaschko lines, without atrophy or preceding inflammation. It commonly occurs in the first few weeks of life and hyperpigmentation tends to persist throughout life. This is thought to reflect an underlying mosaicism or chimerism, and several cytogenetic alterations have been described [2]. This entity has been rarely reported in the Indian literature. We are reporting a classical case with dermatoscopic features.

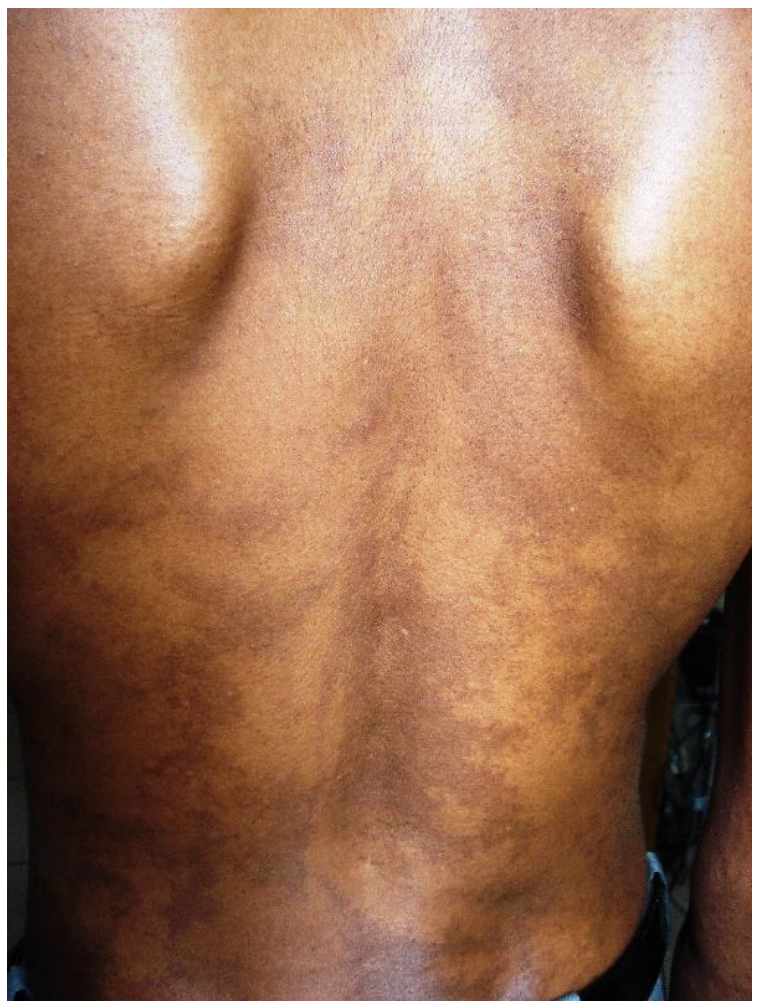
**Case synopsis**

A 33-year-old male patient born from a non consanguineous marriage and an uneventful delivery presented to the dermatology department with complaints of asymptomatic hyperpigmentation all over the body since 2 years of age. The pigmentation initially started on the trunk. then progressed to involve the extremities. The patient had known rheumatic heart disease. He was on penicillin prophylaxis until he attained 20 years of age. He had normal development milestones as per the history without dental, ophthalmologic. or neurologic abnormalities. There was no history of vesiculation or verrucous lesions in childhood. There were no family members with similar skin lesions. He had 3 normal siblings.

Cutaneous examination revealed multiple, light brown, hyperpigmented macules arranged in a linear and whorled pattern arranged along Blaschko lines on the abdomen, chest, back, thighs, and trunk. Palms, soles, genitalia, and mucous membranes were spared. Hair, nails, and teeth were normal. Dermoscopy (Digital dermascope with magnification 20x) revealed a net like pattern of pigmentation over both linear and whorled lesions. Skin biopsy was not done, as the patient refused.



**Figure 1.** Linear and whorled hyperpigmentary lesions on the chest and abdomen

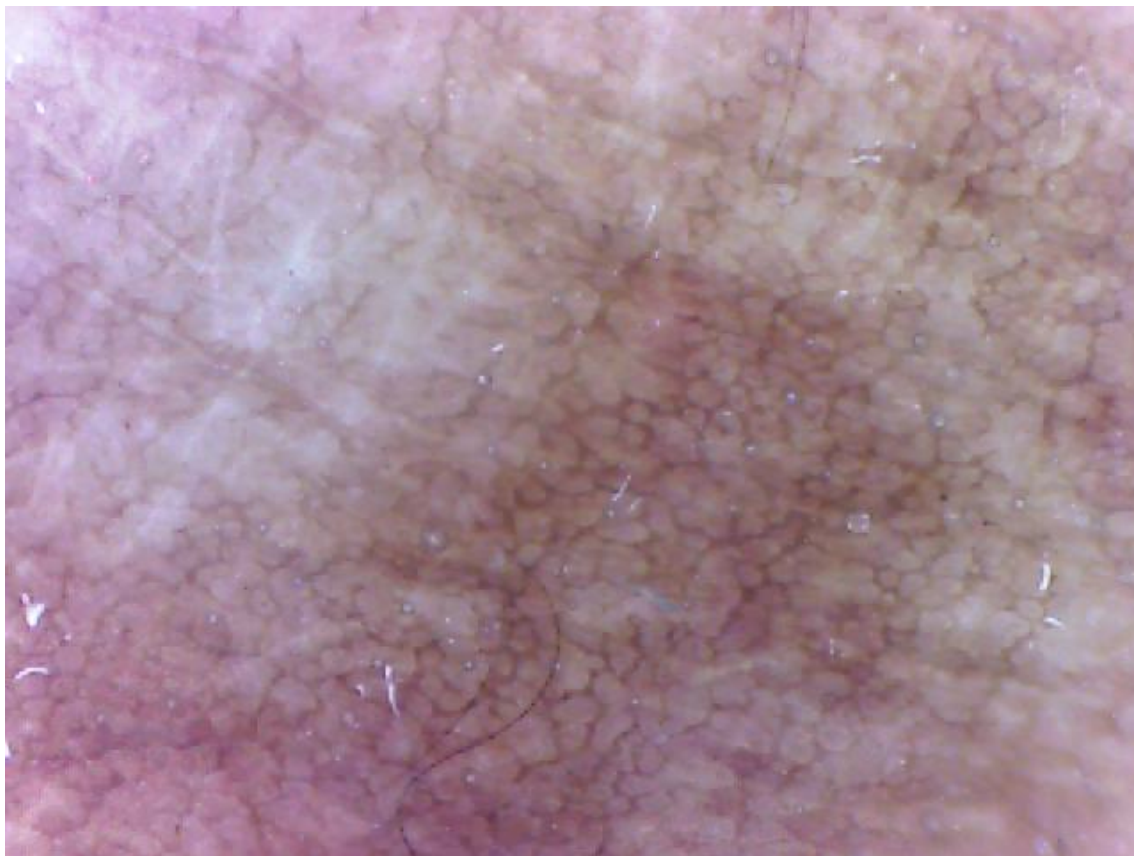


**Figure 2.** Hyperpigmented lesions along the line of Blascko on the back



**Figure 3.** Net like pattern on dermatoscopy of linear lesions





**Figure 4.** Net like pattern on dermatoscopy of whorled lesions

## Discussion

Kalter et al. first described LWNH as hyperpigmentation composed of homogenous colored 1 to 5 mm macules forming reticulated configurations. Streaks and whorls asymmetrically follow Blaschko lines. Onset is usually within a few weeks of age. There is no preceding inflammatory event or palpable lesion. It gradually spreads during the first 1 to 2 years of life with subsequent stabilization. The macules may become less prominent with age in some patients. Mucous membranes, eyes, palms, and soles are spared [1]. LWNH generally occurs sporadically, however, familial occurrence is also noted [3,4]

The conditions in the differential diagnoses for our patient included incontinentia pigmenti, linear epidermal nevus, and hypomelanosis of Ito. Incontinentia pigmenti is an X linked dominant genodermatosis occurring almost entirely in females. It usually 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 the late hypermelanosis in incontinentia pigmenti [1]. Our patient presented with only hyperpigmented lesions since childhood without any vesicular and verrucous lesions. The clinical picture was suggestive of LMNH.

Histopathological examination reveals increased pigmentation of the basal layer and prominence or vacuolation of melanocytes with no pigment incontinence or increase in dermal melanophages [1].

Ertam et al [5] discussed a case of LMNH with dermatoscopic findings. Their dermatoscopic examination of the lesions revealed linear or circular arrangement of streak-like pigmentations arranged in a parallel manner, following the lines of Blaschko. They named it "parallel pattern". Whorled-like configuration of the lesions on the epigastrium showed partially curved, circular, and/or linear streak-like brown and dark brown pigmentation in a parallel pattern. On the popliteal region, the same curved and/or linear streak-like pigmented macules were vertical to skin profiles, but parallel to each other. However, they noted that overall alignment was along the lines of Blaschko. Incontinentia pigmenti shows blue-gray dots, which is the dermatoscopic evidence of pigmentary incontinence [5]. In the present case, a net-like pattern was observed on both linear and whorled lesions. The absence of blue-gray dots ruled out incontinentia pigmenti. A net pattern is also regularly present in Becker nevus and, exceptionally, in hyperpigmented nevus, café-au-lait spot, and in hyperpigmented normal skin [6].

The present case has been reported for its classical presentation, rarity, and dermatoscopic features..

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