

# Linear and whorled nevoid hypermelanosis

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Sir,

Two girls aged 9 and 7 years consulted for asymptomatic, hyperpigmented lesions on the upper and lower limbs following Blaschko's lines evolving since childhood not having changed over the years, with no notion of a similar case of pigmentary disorder in the family or a history of verrucous or hyperpigmented lesions. A dermatological examination revealed multiple brown macules arranged in linear striations following Blaschko's lines on the lower and upper limbs (Figs. 1a and 1b) and trunk (Fig. 2) sparing the mucous membranes and the palmo-plantar region. The rest of the somatic examination, notably neurological, cardiac, dental, and ophthalmological, was unremarkable. Histologic examination of biopsy specimens revealed a hyperpigmented basal cell layer with an increased density of melanocytes.

Linear and whorled nevoid hypermelanosis (LWNH), also named linear nevoid hyperpigmentation, reticulated hyperpigmentation of Naito and Uyeno, and zebra-like hyperpigmentation in scrolls and striae [1]. It is a rare disorder of skin pigmentation characterized by swirls and whorls of reticulated hyperpigmented macules following Blaschko's lines. These lesions usually occur within several weeks of birth and stabilize rapidly over time [2]. However, late forms may be described [3,4]. It may be associated with extracutaneous anomalies including central nervous system disease, cardiac malformations, and psychomotor retardation.

It poses a differential diagnosis with several pathologies, including incontinentia pigmenti, which presents a vesicular and verrucous stage before the pigmentary appearance. Café au lait macules of McCune–Albright



**Figure 1:** (a and b) Hyperpigmented macules in a linear striated or whorled distribution on the limbs in both girls.



**Figure 2:** Linear and whorled nevoid hypermelanosis on the trunk.

syndrome may be confusing disorders with an early onset and hyperpigmentation along Blaschko's lines. Ito's hypomelanosis, Conradi–Hünemann–Happle syndrome. There is no current treatment for LWNH, yet camouflage cosmetics may be helpful [5].

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## Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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