

DOTS ON CANVAS: THE USEFULNESS OF WOOD LAMP EXAMINATION IN A CASE OF LINEAR AND WHORLED NEVOID HYPERMELANOSIS

LIUBA GARAGA*, OANA MIRELA TIUCĂ**, ANDREEA BEATRIX BĂLAN*, RALUCA-IOANA AILINCĂI*, SILVIU-HORIA MORARIU**

Summary

Introduction: Linear and whorled nevoid hyper-melanosis (LWNH) is a rare, asymptomatic condition mainly affecting the Blaschko lines.

Case report: We report the case of a 5-year-old female patient who presented with clinically ill-defined brown macules on the trunk and upper limbs. Wood lamp examination highlighted clearly demarcated lesions, while dermoscopy identified hyperpigmented areas arranged in a parallel manner. The diagnosis of LWNH was established, and the patient was reassured of the benign nature of the disease.

Conclusions: LWNH is a benign skin condition that can be associated with systemic comorbidities. Future promising therapeutic options are much-needed additions to this complex disease approach.

Keywords: hypermelanosis; Blaschko lines; the Wood lamp.

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Introduction

Linear and whorled nevoid hypermelanosis (LWNH) is a rare and sporadic pigmentation disorder defined by reticulate hyperpigmented macules that occur along Blaschko's lines [1]. Several cases of familial involvement have been reported [2,3]. Even though it is most often a benign, asymptomatic condition, some patients may associate extracutaneous changes, such as nervous, musculoskeletal, ophthalmic, or cardiovascular anomalies. Such cases can sometimes require a multidisciplinary approach. This paper aims to present the case of a patient diagnosed with LWNH and to summarize current clinical and therapeutical knowledge based on existing literature.

Case Report

A 5-year-old female patient with Fitzpatrick skin type IV and no significant medical history presented to the Dermatology Clinic of Mureș Clinical County Hospital complaining about "brown spots" on her trunk and right upper limb. Dermatological examination revealed asymptomatic light brown macules, measuring between 0.5 and 1.5 cm in size, uniformly pigmented, with irregular margins, and disposed along Blaschko's lines on the right upper limb and on the left abdominal flank (Figure 1). The lesions were not preceded by other inflammatory skin changes in the same location. The rash appeared one year prior, initially only on the trunk but has gradually expanded. The mucosas, hair, nails, palms,

* Dermatology Clinic, Mureș Clinical County Hospital.

** Department of Dermatology, "George Emil Palade" University of Medicine, Pharmacy, Science, and Technology of Târgu-Mureș, Romania.



Figure 1. Light brown macules arranged along Blaschko's lines.

soles, and face were spared. The lesions had a strong emotional impact on the patient's family due to their aspect and the fear that they would continue to spread. Skin examination of the patient's family members (grandparents, parents, and two siblings) didn't reveal any similar-looking lesions.

Wood lamp examination showed no fluorescence, but the macules, clinically presenting with poorly defined margins, appeared clearly demarcated, confirming the intraepidermal location of the pigment deposit. Clinically inapparent lesions were also identified (Figure 2). Polarized light dermoscopy revealed hyperpigmented areas arranged in a parallel manner (Figure 3).

Laboratory tests were within limits. The fungal examination was negative. The patient's parents refused a skin biopsy, fearing the residual

scar. Upon complete check-up, no systemic, ophthalmologic, or dentition abnormalities were detected.

A diagnosis of LWNH was established. Due to no significant past or present associated anomalies, both patient and family were reassured of the benign nature of the disease and informed that given that the disease does not usually affect the face, it has a smaller cosmetic burden. 755 nm Q switched laser treatment was advised but declined due to economic burden, and the patient's family agreed to periodical follow-up.

Discussions

LWNH is a rare, sporadic, and benign clinical entity characterized by the appearance of hyperpigmented macules that follow Blaschko's



Figure 2. Hyperpigmentation on Wood's lamp examination.

lines [1]. It was first reported as such by Kalter et al. in 1988 [4]. The umbrella term "cutaneous pigmentary mosaicism" is sometimes used [5,6] and includes similar diseases, such as progressive cribriform and zosteriform hyperpigmentation, zebra-like hyperpigmentation in whorls and streaks [7,8], and reticulate hyperpigmentation of Iijima. To date, less than 100 cases of LWNH have been reported worldwide and only 60 cases were published in the Pubmed database.

Genetic mosaicism is the most likely cause of such a condition. Skin pigmentation mosaicism involves the concomitant proliferation and migration of two distinct types of melanocytic populations with different potentials for pigment production [6]. Mosaic trisomy of chromosomes 7, 14, 18, or 20 can be linked to LWNH, as can the inversion of chromosome 9 [9,10]. Family involvement, although very rare, has also been

described [2,3]. In 2011, Metta et al. described LWNH in three successive generations [3].

Usually, the lesions appear in the first weeks of life. They expand and become darker in color for 2-3 years, after which the rash stabilizes [11]. Clinically, two forms of LWNH are described: with unilateral involvement, not associated with systemic lesions, or with bilateral involvement, frequently accompanied by skeletal (brachydactyly, facial and limb asymmetry), cardiovascular (ventricular septal defect, tetralogy of Fallot), and neurological abnormalities (microcephaly, convulsions, mental retardation) [12]. Dermoscopic features are linear or circular streak-like pigmentation arranged in a parallel manner [13]. Wood lamp examination is particularly of use in appreciating disease extension.

Differential diagnoses should take into account other diseases that follow Blaschko's

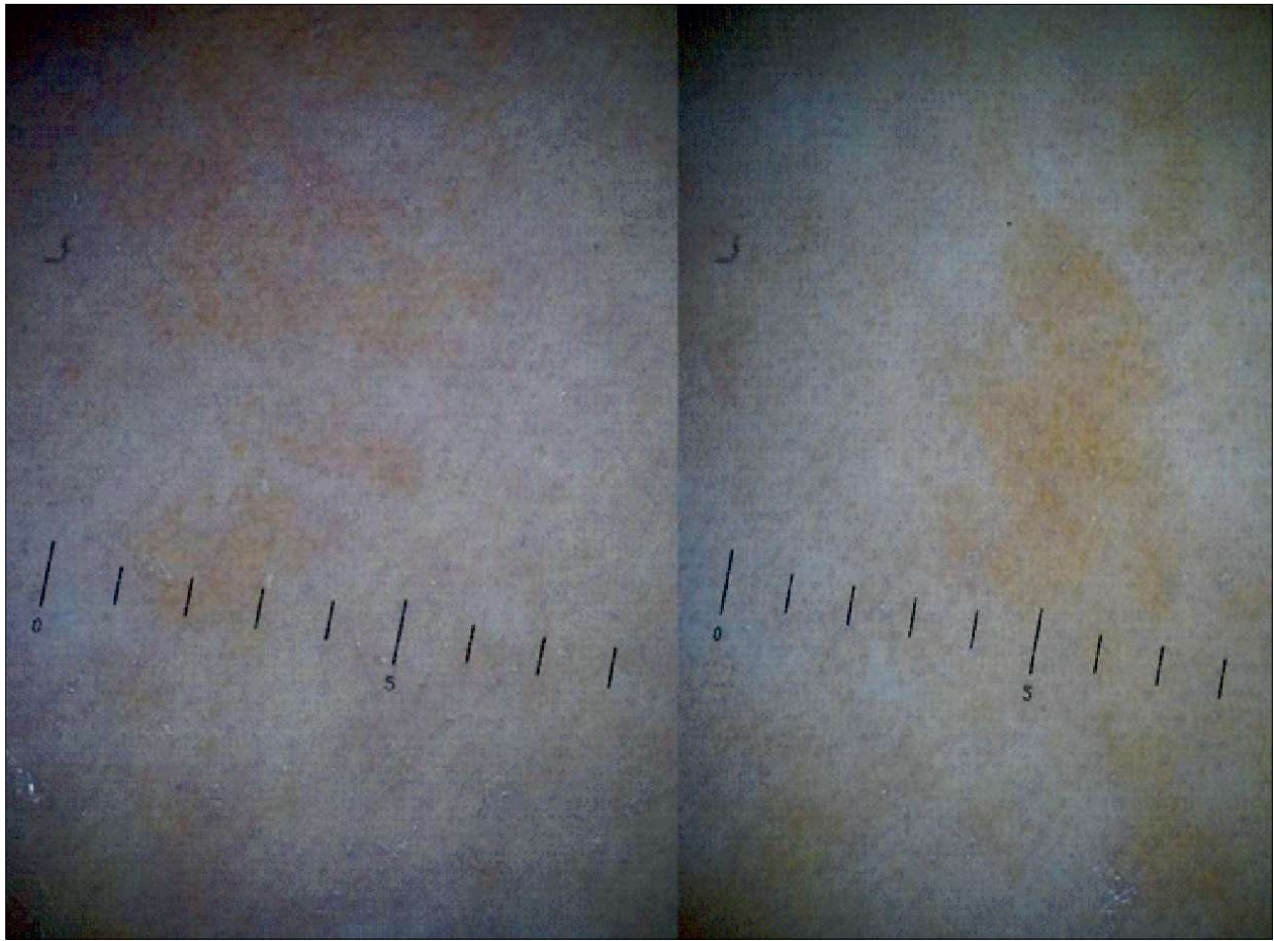


Figure 3. Dermoscopy image- linear streak-like pigmentation arranged in a parallel manner.

lines. The third stage of incontinentia pigmenti (IP) is preceded by a vesicular stage and then a verrucous stage, which should be evidenced by anamnesis. However, diagnostic difficulties can occur because the first two stages of IP may be missing [14,15]. In extensive cases of hypomelanosis of Ito (HI), determining whether the abnormality is hyperpigmentation or hypopigmentation may be difficult, and the appearance of patient lesions may be confused with that of LWNH [3]. In such cases, dermoscopic and Wood lamp examinations are useful in highlighting the hypopigmented macules. In the early stages, epidermal nevi may appear as hyperpigmented macules along Blaschko's lines. An important differentiation criterion refers to the moment of appearance because often epidermal nevi are present at birth, and shortly, their surface becomes papillomatous

or warty [16], while in LWNH, skin texture is not altered.

The peculiarity of the described case lies in the presence of a bilateral asymmetric rash without systemic involvement; the appearance of lesions of HNLC was at an older age compared to cases previously described in the literature. Wood lamp skin examination is of importance in appreciating the real extension of the lesions.

Therapeutic options in LWNH are limited. Chemical peels, such as 2% hydroquinone [17] or 70% glycolic acid in combination with 35% trichloroacetic acid [18], have no significant results. Laser treatment is a promising option, but it's still under investigation. In a study that enrolled 4 children aged 6-18 years with LWNH, QS lasers 755 nm and 532 nm were found to be effective. QS 755 nm laser was more effective in cutaneous phototypes III and IV, while QS 532

nm showed better results in patients with lighter skin. Only one patient reported recurrence after two years of follow-up (30% of the affected area was pigmented). Mild pain during the procedure, transient purpura at the site of laser action, mild and transient hypopigmentation were reported as side effects [20]. A recent report describes the successful use of a QS 694 nm ruby laser for the treatment of LWNH on the face of a 4-year-old boy without recurrence after a 3-month follow-up [9]. Camouflage makeup should also be considered [21].

Conclusions

LWNH is a benign skin condition that can be associated with systemic comorbidities and thus may require a multidisciplinary approach. Much is still incompletely known about this disease, but the patient's reassurance of the benign course of evolution is necessary. Future promising therapeutic options, especially laser therapy, represent much-needed additions to this complex disease approach.

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Conflict of interest
NONE DECLARED

Correspondance address: Oana Mirela Tiucă
"George Emil Palade" University of Medicine, Pharmacy, Science, and Technology of Târgu-Mureş,
Romania
e-mail: oanaa.marginean@yahoo.com