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Case Report

Evaluation of Melanonychia as a Result of a Pediatric Case Evaluated in the Daily Clinic. Is More Frequent than we Can Think?

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Abstract

Melanonychia is defined as the pigmentation of the nails with shades ranging from light brown to black, secondary to the deposition of melanin in the nail plate. The pigment can come from exogenous or endogenous sources and cover the entire nail plate, a portion of it transversely, or more frequently as longitudinal bands. Melanonychia can be an early sign of nail melanoma, for which the appropriate approach is of great importance. Diagnostic aids such as dermatoscopy and histopathological study are useful to specify the etiology and define the treatment.

Keywords: *Evaluation of Melanonychia; Result of a Pediatric; Daily Clinic*

Introduction

The nail apparatus is made up of the nail plate, the proximal or eponychial fold, the distal or hyponychial fold, the lateral folds, the nail bed, and the matrix or germinative zone that is hidden in the proximal part of the nail.

The nail plate is made up of multiple layers of completely keratinized keratinocytes, 0.5 to 0.7 mm thick and pink in color.

The growth rate is continuous at 0.125-0.1 mml per day. If for any reason it stops, transverse bands called Beau's grooves are formed.

The nail plate is divided into 3 layers (Figure 1):

a) The dorsal or upper area, whose brightness is due to the cells of the dorsal matrix

b) The intermediate zone that comes from the distal portion of the ventral matrix

c) The deepest or ventral area that comes from the bed.

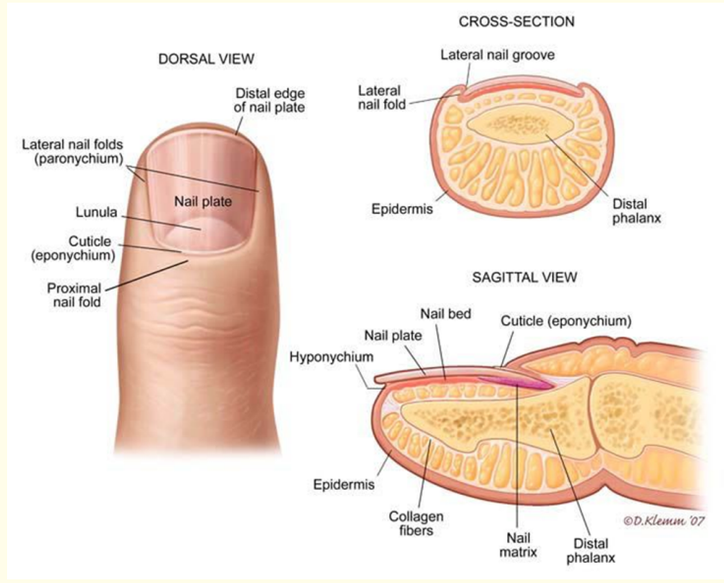

Figure 1

The bed is the main holding area of the sheet. Its dermis is highly vascularized and its capillaries are arranged longitudinally. The matrix is the germinative tissue of the nail, where the cells responsible for the onychization process (special keratinization of the lamina) are found [1].

Melanonychia (from the Greek black mélan and ónyx, nail) is the pigmentation of the nail plate with different shades of grey, brown or black. The most common clinical form is the linear distribution of the pigment along the long axis of the nail, which is called longitudinal melanonychia (ML) [2]. The origin of the pigment can be exogenous or endogenous, in the first group the causes include mycotic and bacterial infections, smoking and the presence of hemoglobin due to trauma (Figure 2). In the second it is the result of melanocyte activation and hyperplasia. The causes of LM are generally endogenous, in which the production of melanin in the nail matrix is favored; benign entities include nevi and lentigines of the nail matrix, autoimmune and iatrogenic diseases, and trauma [3]. Early-stage nail melanoma is a cause of longitudinal melanonychia.

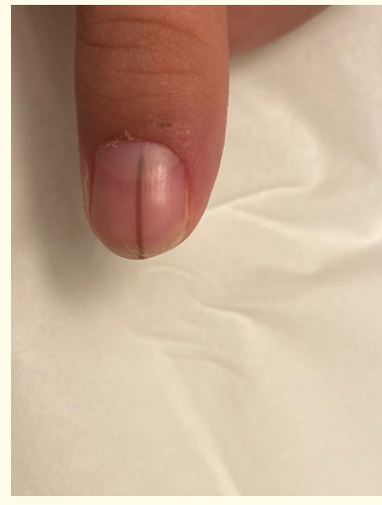

Figure 2

To establish a correct diagnosis, it is necessary to emphasize family history of melanoma, physical activities, occupational exposure, systemic diseases, medications and the time of evolution [2,3].

Clinical aspects

The clinical approach to LM consists of evaluating the distribution of the pigment (homogeneous or non-homogeneous), the color and the bands (diameter) [3,4].

For the study of melanonychia, the ABCDEF mnemonic has been proposed in search of nail melanoma data, which is considerably different from the ABCDE mnemonic for the study of nevi. The letter A encompasses age (age), African Americans, Asians and Native Americans; B is assigned for borders of 3 mm or more that can have shades of brown to black; C considers changes in the nail band; D is to specify the most affected digit(s); E describes extension to the cuticle or nail fold, and F refers to personal or family history of dysplastic nevi or melanoma [5]. The ABCDEF mnemonic does not improve the sensitivity or specificity of the diagnosis of melanonychia.

Findings suggestive but not pathognomonic of nail melanoma

Hutchinson's pseudosign: corresponds to the extension of the melanin pigment to the skin adjacent to the nail plate. When pigmentation is observed at the subcuticular level at the expense of the presence of pigment in the underlying nail plate, it is called Hutchinson's pseudo-sign (Figure 3) and is a visual phenomenon due to the transparency of the cuticle [3,4]. Table 1 shows the conditions that can produce this sign.

Figure 3

Addison's disease
AIDS
Bowen's disease
Drug-induced pigmentation
Laugier's syndrome
Malnutrition
Nail matrix nevus
Peutz-Jeghers syndrome
Racial pigmentation (phototypes V and VI)
Radiotherapy
Trauma

Table 1. *Causes of Hutchinson's pseudosign.*

The melanin in the nail plate comes from the melanocytes in the matrix. In an adult, the nail matrix has around 200 melanocytes per mm2 as opposed to the 1150/mm^2 found in the epidermis. From a histological point of view, we can divide LM caused by melanocyte activation and melanocyte hyperplasia.

Longitudinal melanonychia due to melanocyte activation

Melanocytic activation is responsible for 73% of LMs [2].

Racial

This is the physiological pigmentation that is detected in the nails of individuals with dark phototypes (IV to VI). It appears in up to 80% of African-Americans, 30% of Japanese, and 50% of Hispanics [7,8]. Racial melanonychia manifests gradually, usually in proportion to chronological aging. It is more evident in fingers subjected to constant trauma [2].

Traumatic

Secondary to onychotillomania or chronic friction. Chronic friction trauma occurs primarily in the toenails, which are subject to constant friction from footwear. They are generally symmetrical and affect the lateral aspects of the first, fourth and fifth fingers. Onychotillomania is more frequent in the second finger of both hands [7].

Inflammatory

It is usually not very evident in early stages. It is associated with abnormalities of the nail plate surface and does not have a characteristic clinical appearance or dermatoscopic signs. It can be seen in cases of pustular psoriasis, lichen planus, amyloidosis, and chronic radiodermatitis [2,6].

Associated with endocrine pathology

Some of the entities that have been described in association with LM are: Addison's disease, Cushing's syndrome, hyperthyroidism and acromegaly. Physiologically, longitudinal melanonychia can be seen during pregnancy due to endocrine changes [2,7,9].

Associated with HIV

It is possible to detect LM in HIV-positive patients and unrelated to treatment with zidovudine in one or several nails with or without association with spots on the palms, soles and mucous membranes [6].

Due to nutritional deficiencies

Vitamin B12 and folate deficiency can cause longitudinal melanonychia with shades of dark blue and black. Similar pigmentation can also be seen on the knuckles and distal phalanges [6].

Syndromic

In Laugier Hunziker syndrome, the LM usually has a maximum diameter of 2 mm in single or double bands, which accompanies the other characteristic findings of the entity: idiopathic mucocutaneous hyperpigmentation, lenticular pigmentation in the gums, oral mucosa, lips and anogenital [6].

Drug-induced

Some can generate longitudinal and less frequently transverse bands of melanonychia in the hands and feet, affecting all or some of the fingers.

The hue can vary from brown to black. It is a common effect of antineoplastic drugs, which activate melanocytes in the nail matrix (Figure 4).

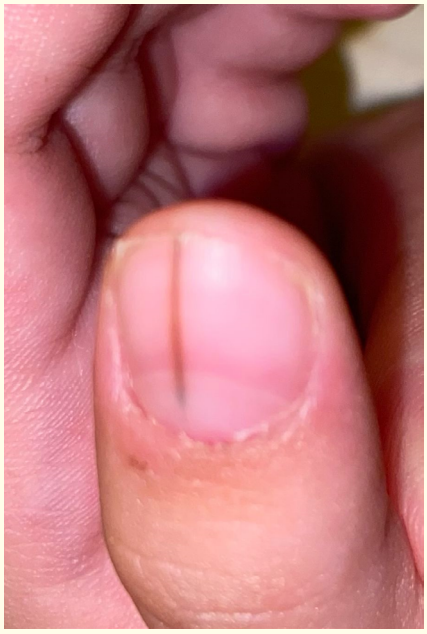

Figure 4

Transverse melanonychia can be seen in patients treated with psoralens, infliximab, and zidovudine. It is secondary to intermittent production of melanin. There is a reported case of transverse melanonychia due to melanotan, a synthetic drug analogous to melanocyte-stimulating hormone that was originally designed as a photoprotector for patients with erythropoietic porphyria and actinic dermatoses.

Some drugs such as clofazimine generate melanonychia after being eliminated through the nail matrix and being deposited in the nail plate [10,11].

Tetracyclines and antimalarials generate melanonychia by pigmentation at the level of the dermis. In these cases, the growth of the nail plate does not generate changes in the clinical appearance and is associated with pigmentation in other tissues [12].

There are cases with Hutchinson's pseudosign signal associated with minocycline and amlodipine [13,14]

table 2 lists the drugs that have been associated with the presence of longitudinal melanonychia.

Bleomycin
Busulfan
Cyclophosphamide
Daunorubicin
Doxorubicin
Etoposide
5-fluoroacyl
Hydroxyurea
Melphalan
Methotrexate
ACTH
Chloroquine
Clofazamine
Fluconazole,
Ketoconazole
Gold salts
Ibuprofen
Lamivudine
Mepacrine
Mercury
Phenytoin
Phenothiazides
Psoralens
Steroids
Sulfonamides
Timolol
Zidovudine

Table 2: *Medications that can cause melanonychia.*

In general, drug-associated melanonychia is reversible once the offending drug is discontinued; however, clinical improvement is observed after months [10,11].

Associated with skin tumors: some non-melanocytic tumors of the nail apparatus that can manifest with LM include onychomatricoma, Bowen's disease and basal cell carcinoma [15,16].

Longitudinal melanonychia caused by proliferation of melanocytes

In these cases, the nail changes are the result of the local increase in the number of melanocytes.

Melanocytic lentigines and nevi (both acquired and congenital) are the cause of 12% of LMs in adults [2]. In both cases, the dermatoscopic findings are suggestive of benignity.

Subungual melanoma: begins as longitudinal melanonychia in between 38% and 76% of cases and is accompanied by nail dystrophy, periungual pigmentation, and ulceration. It predominates in adults with a mean age of 60 years. Its prognosis depends on when the diagnosis is suspected, and 5-year survival ranges from 16% to 76% (7.17).

Longitudinal melanonychia due to non-melanic pigment

Exogenous: includes silver nitrate, ethacridine lactate, henna tattoos, and nicotine [6].

Subungual hematoma

Reddish or reddish-black pigmentation is observed, which depends on the time of evolution. Dermatoscopically, globules in shades ranging from red to black are distinguished along the lateral margins of the pigment, which is homogeneous. The hematoma moves away from the proximal nail fold in proportion to the growth of the nail. Some skin tumors can generate subungual hematomas, so their presence does not rule out the existence of a melanoma [4,6,7].

Onychomycosis

LM due to onychomycosis is produced mostly by dematiaceous fungi and by Trichophyton rubrum variety nigricans, which produce melanin as a protection mechanism against environmental stress and enzymatic hydrolysis. May be accompanied by nail dystrophy. Typically, melanonychia bands are wider distally [18].

Dermatoscopy

Nail plate dermatoscopy is a non-invasive method that helps in the differential diagnosis of LM. If we find a pigment band composed of multiple thin and homogeneous gray lines, they suggest melanocytic activation. Instead, a brown band composed of multiple lines is secondary to melanocyte hyperplasia, as seen in lentigo or melanoma. These lines can be regular or irregular depending on parallelism, spacing, width, and color. If it is irregular, malignancy must be suspected [6].

Dermatoscopy of the matrix and nail bed

It is an intraoperative procedure indicated when the nail biopsy is being performed and allows distinguishing aspects that are not visible when the nail plate is interposed. In this case, a dermatoscope with polarized light is more useful because it does not require contact with the lesion. Four patterns have been described: regular gray in case of hypermelanosis, regular brown in benign melanocytic hyperplasia, regular with globules and spots in case of a melanocytic nevus, and irregular in melanoma [7,19].

Biopsy

The indications for nail biopsy in longitudinal melanonychia are

1. Isolated pigmented band on a single finger that develops during the fourth and sixth decades of life.
2. Rapid onset pigmentation.
3. Acquired pigmentation of the thumb, index finger, or first toe.

4. Any suspicious lesion in a patient with a history of melanoma.

5. Hutchinson's pseudosign (2.6).

It can be taken from the nail plate, nail bed, or matrix depending on the clinical features of melanonychia. In case of pigment deposit, it is recommended that the biopsy only include the nail plate and can be performed with a punch or obtaining part of the distal portion of it. When it comes to LM, it is convenient to take a longitudinal lateral en bloc biopsy. In pigmented tumors, it is suggested that it includes the matrix, the bed or the entire nail unit.

Nail plate biopsy consists of cutting a 3-mm-wide distal fragment of the plate.

After removing the lamina in the nail bed biopsy, a longitudinal spindle excision of 3 mm width is performed; The double punch biopsy has also been described, with the first punch a part of the nail plate is removed and with a second it is covered up to the nail matrix.

In the matrix biopsy, a flap is excised from the proximal nail fold, leaving it uncovered, then the procedure is started in a transverse, fusiform or crescent shape, avoiding the proximal area and suturing the defect.

The lateral longitudinal biopsy is a longitudinal resection of the matrix, nail folds, bed and hyponychium through the fusiform incision in the most distal lateral fold passing through the lateral nail groove and medially through the lamina reaching the hyponychium. The tissue is dissected en bloc, freeing the bed, and the lateral nail fold is sutured to the nail bed using monofilament and synthetic suture material.

Tangential biopsy has recently been suggested, which is similar to matrix biopsy, but takes a minimal amount of tissue (less than 1 mm thick), which carries a small risk of nail dystrophy [2,20,21].

Treatment

It depends on the etiology. In benign lesions, vigilance is recommended. When surgery is chosen, the patient should be informed of the possibility of nail plate dystrophy. In nevi and lentigines, tangential biopsy is a good option because it allows the lesion to be removed with fewer adverse effects [2,5].

In the case of melanoma in situ, a conservative surgical approach is chosen, trying to preserve the phalanx to achieve a better functional and cosmetic result without affecting the prognosis. However, for invasive melanomas, amputation is required trying to preserve functionality [22].

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