

CASE REPORT

Melanonychia Striata Longitudinalis: A case report with brief review of literature

AlMutairi A, Sadiq Mahdi, MD

Department of Internal Medicine, Salmaniya Medical Complex, Bahrain

ABSTRACT

Melanonychia striata longitudinalis is a pigmented (dark brown-to-black) band running along the long axis of the nail. This abnormal color of the nail plate is caused by a focal increase of melanin because of increased number and/or function of normal or abnormal melanocytes in the nail matrix. Pigmented streaks in nails due to melanocytic hyperplasia are said to be so exceedingly common in black persons, as to be considered normal and the phenomenon is benign in the majority of cases. Longitudinal melanonychia presents a difficult clinical challenge because subungual melanoma must always be included in the differential diagnosis and because the cause of longitudinal melanonychia is usually not apparent. In our patient reported here, a 25-year-old male patient with brownish bands over nail plates of both thumb nails. The diagnosis was made by characteristic dermoscopic feature of presence of brownish symmetrical linear parallel lines in nail plates.

INTRODUCTION

A 25 - year - old male patient, presented with a longitudinal, asymptomatic, brownish discoloration on the nail plate of both thumb nails. The lesion exhibited a sudden onset and a progressive course with a linear alteration since it was initially noticed 4 years prior (Fig. 1-A&B). There was no pigmentation of the oral mucosa and/or any other abnormal pigmented lesion anywhere else in the body. The patient was otherwise well and

there was no history of skin or systemic illness, trauma or any medication. No lymphadenopathy, hormonal abnormality, or other adventitious influence was detected. He was having no family history of such type of abnormal pigmentation or melanoma.

The diagnostic challenge here was to exclude the most serious disease of the nail unit melanoma, primarily presenting with melanonychia. Nail plate dermoscopy showed thin, brownish, regu-



Fig. 1 A&B A longitudinal, brownish discoloration in nail plate both thumb nails.

Correspondence: Dr. Sadiq Mahdi, Department of Internal Medicine, Salmaniya Medical Complex, Bahrain Manama,

lar, parallel lines which suggested longitudinal melanonychia (Fig. 2). The patient continues to be monitored for further progression.

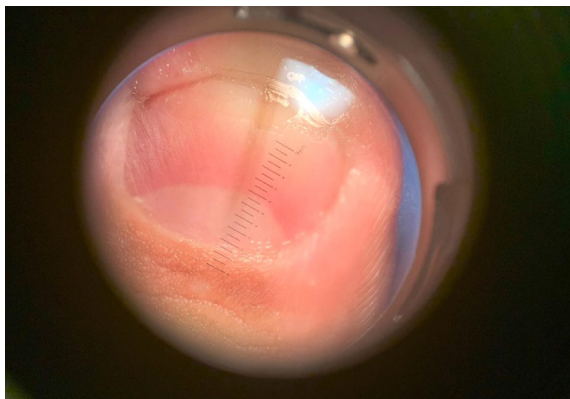


Fig. 2 Nail plate dermoscopy shows brownish, thin, regular, parallel lines.

FINAL DIAGNOSIS

- Melanonychia striata longitudinalis

DISCUSSION

Melanonychia also called “melanonychia striata”, refers to the Greek word “Melas” meaning black (or brown color) and “Onyx” meaning nail. It is characterized by brown black discoloration of the nail plate and the pigment referred to be conventionally melanin. It may involve single or multiple nails, both in finger and toes. It is due to increased activity of melanocytes or melanocytic hyperplasia in the nail matrix, with increased melanin deposition in the nail plate.¹ The most common type of longitudinal melanonychia due to melanocytic activation is ethnic melanonychia, which typically presents in dark-skinned individuals as multiple bands affecting multiple nails. Other causes of melanocytic activation include pregnancy, chronic local trauma or inflammation, systemic conditions, and drugs. Nail lentigo, nail melanocytic nevus, and nail melanoma are causes of longitudinal melanonychia due to melanocytic hyperplasia.²

Melanin within the nail plate is usually produced by nail matrix melanocytes.³ Active melanocytes transfer melanin-rich melanosomes by way of dendrites to differentiating nail matrix-derived onychocytes.⁴ This process most commonly results in a longitudinal band of melanonychia, but total or transverse melanonychia may also occur, although rarely.⁵ Melanonychia usually originates in the distal nail matrix.⁶ Most melanocytes in the proximal nail matrix lie dormant in the lower 2-4 germinative cell layers. While active (as well as dormant) melanocytes exist in the 1st and 2nd germinative layers of the distal nail matrix.⁷ The etiologies of melanonychia may be divided into 2 broad categories: melanocytic activation and melanocytic hyperplasia.

Kawamura et al. reported melanonychia in 11.4% cases, with highest prevalence in people aged 21-26 years and Tasaki et al. found the prevalence to be 20% in males and 23% in females. The number of nails involved and the width of the pigmented bands differ according to causative factors. While drug exposure, dermatological diseases, and racial pigmentation typically involve multiple nails, lentigines and nail matrix nevus are monodactylic (involving single nail/digit).^{9,10}

Racial melanonychia is commonly encountered in skin phototypes IV, V, and VI. That is, darkly pigmented races including Blacks, Asians, Middle-East, and Hispanics. The incidence reportedly varies from 1% in whites, 10%–20% in Japanese and Asians, and 77-100% in African Americans. Racial melanonychia is more common in fingers (thumb, index finger), generally involves multiple nails and the band width increases with age.¹¹

Longitudinal melanonychia may also be associ-

ated with pregnancy and generally involves several fingers and/or toe nails. It is thought to result from the activation of nail matrix melanocytes and may resolve or persist following delivery.¹² Bacterial melanonychia is frequently caused by gram negative pathogens; *Pseudomonas aeruginosa*, *Klebsiella* and *Proteus*. The risk factors include wet work and an immunocompromised state. The pattern of nail involvement can be longitudinal streaks with wider proximal edge or diffuse, which starts from the junction of proximal and lateral nail folds and spreads with irregular medial border.¹³

Fungal melanonychia may be caused by both dematiaceous and nondematiaceous fungi; most common being *Trichophyton rubrum* and *Scytalidium dimidiatum*. The involved nail typically has brown to black bands which are better seen with dermoscopy, and subungual hyperkeratosis with or without periungual inflammation. The pattern of nail involvement may indicate the causative agent. Longitudinal melanonychia is more common with dermatophytes like *Trichophyton rubrum* while diffuse pigmentation is seen with molds such as *Scytalidium*, *Aspergillus niger*.¹⁴

HIV positive patients may show diffuse melanonychia or multiple longitudinal or transverse bands in multiple fingers and toenails, often associated with hyperpigmentation of palms, soles and mucous membranes. Longitudinal melanonychia can also be seen with anti retroviral drugs like zidovudine.¹⁵

Inflammatory skin conditions like psoriasis, lichen planus, amyloidosis, chronic radio-dermatitis, and chronic paronychia in and around nail unit leads to melanocytic activation resulting in melanonychia. Longitudinal melanonychia has

also been seen in connective tissue diseases like systemic sclerosis and SLE.^{16,17} Longitudinal melanonychia can also occur due to melanocytic activation by nonmelanocytic benign and malignant tumors such as onychomatricoma, Bowen's disease, squamous cell carcinoma, basal cell carcinoma, subungual fibrous histiocytoma, verruca vulgaris and subungual keratosis.¹⁸

Melanonychia in systemic diseases tends to involve multiple fingers and toe nails, and generally presents as either diffuse melanonychia or as multiple pigmented bands.¹⁹ It is uncommon and is seen in endocrine disorders (Addison's disease, Cushing's syndrome, hyperthyroidism and acromegaly), alkaptonuria, hemosiderosis, hyperbilirubinemia, and porphyria. Repeated trauma as seen in onychotillomania, onychophagia, or friction may lead to melanonychia. Also repeated trauma from ill-fitting shoes or overriding toes is a likely cause.²⁰

Iatrogenic causes of melanocytic activation include medications (especially chemotherapeutic agents), phototherapy, X-ray exposure, and electron beam therapy. The pattern of nail involvement can be diffuse, transverse, or longitudinal. Fortunately, when melanonychia is drug-related, it typically fades slowly following drug withdrawal. Transverse melanonychia has occurred in conjunction with use of the following: electron beam therapy,²¹ conventional radio-graphic therapy to treat hand dermatitis (used in the 1950s and 1960s), psoralen with ultraviolet A (PUVA),²² infliximab, zidovudine, prolonged antimalarial therapy with amodiaquine, chloroquine, mepacrine, or quinacrine, and chemotherapy with agents such as doxorubicin, bleomycin, cyclophosphamide, daunorubicin, dacarbazine, 5-fluorouracil, methotrexate, and hydroxyurea.²³

Transverse melanonychia associated with electron beam therapy and PUVA is benign and typically resolves with the cessation of treatment. Syndrome-associated melanonychia, which occurs in conjunction with Laugier-Hunziker, Peutz-Jeghers, and Touraine syndromes, typically involves multiple digits, and are also characterized by mucosal pigmented macules involving the lips and oral cavity.³

Longitudinal melanonychia is seen in malnutrition especially protein energy malnutrition and vitamin D deficiency. Vitamin B12 deficiency produces reversible nail pigmentation that can be longitudinal, diffuse bluish or reticulate probably due to reduced glutathione levels resulting in disinhibition of tyrosinase enzyme involved in melanogenesis.²⁴

Brown to black pigmentation of nails can be seen due to exogenous agents like henna, dirt, tobacco, potassium permanganate, tar, and silver nitrate. It presents as a horizontal band with the distal border parallel to the proximal nail fold and tends to move out with the nail plate. Silver nitrate presents with a dark color band with irregular medial border and staining of adjacent skin of nail folds.²⁵ Hematoma is the most common cause of nail brown-black pigmentation. It can be either acute (following single heavy trauma) or chronic (repeated, micro trauma). While acute subungual hematoma has a deep red/purple band and does not reach the free margin of the nail, chronic subungual hematoma has a red brown, elliptical shape mimicking a longitudinal streak. A true longitudinal band is very rarely seen. Small, round blood globules are seen at the periphery of hematoma on dermoscopy.²⁵ Benign melanocytic hyperplasia can be either due to nevus or lentigo. Lentiginosities are often seen

more than nevi in adults while nevi are more common in children. Nail matrix nevus may be congenital or acquired, majority being junctional. Nevus represents 12% of LM in adults and 48% of LM in children. It generally presents as a light brown to black, (3-5 mm) broad longitudinal band on fingernails especially thumb.¹⁸ The pigmentation is generally homogenous in intensity and color. But dark bands may arise on light background, appreciated better on onychoscopy. Darker bands may be associated with periungual pigmentation (pseudo Hutchinson's sign) in one-third of the cases.³ Histologically, nevus is characterized by the formation of nest of melanocytes. Lentigo is characterized by melanocytic hyperplasia with absence of melanocyte nests. Increased melanocytes (5-31/mm) are present within nail matrix epithelium. Lentigo in the nail matrix is more common in adults and is observed in around 9% of all longitudinal melanonychia (LM) cases in adults.²⁶

Nail unit melanoma (NUM) is rare. It comprises about 0.7% - 3.5% of all cases of cutaneous melanoma in the western world. Thumb, great toe, and middle finger are common sites. LM is the first manifestation of NUM in 38% - 76% of the cases.²⁷ Variegation in color, irregular/blurred borders of the band, crisscrossing, periungual pigmentation (Hutchinson sign), nail dystrophy, and ulceration or blood spots indicate malignant change and a biopsy should be considered. In rapidly growing melanomas, the proximal end of the band may be wider than the distal portion known as pyramidal sign. NUM carries a poor prognosis as compared to cutaneous melanomas probably due to delay in diagnosis, the average delay being 2 years.²⁸ The five-year survival rate is 51%; 88% for a Breslow thickness of 2.5mm

or less and 44% for a thickness greater than 2.5mm.³⁰

Examination of all finger and toe nails and a complete muco-cutaneous examination is important for evaluation of melanonychia. We should note: a) Number and location of involved nails-single/multiple; b) Morphology of LM-whether similar or different, if multiple nails involved; c) Site or location of melanonychia on the nail plate-above, within, or beneath. Examination of the distal free edge of the nail plate or hyponychium might indicate the origin of pigment. Ventral nail plate pigmentation originates from the distal matrix, while the dorsal nail plate from proximal matrix. The localization of pigment helps in selecting the anatomic site for exploration and biopsy, and can prevent a visible definitive nail dystrophy, if the surgery is confined to the distal matrix. Pattern of melanonychia-complete; longitudinal; or transverse.

Complete melanonychia-The extent and configuration (proximal and distal end including shape of proximal end of pigmentation with respect to proximal nail fold); Longitudinal melanonychia (LM)-Color, homogeneity, regularity, width, whether wider proximally or distally, shape, margins and lateral borders of the band. Pyramid-shaped melanonychia with base toward proximal nail fold is suggestive of NUM. Other nail signs-Nail dystrophy, nail plate changes (abrasion, splitting or fissuring) and periungual pigmentation, bleeding are pointers to NUM. In addition, nail dystrophy, nail plate changes and pigmentation may be seen in fungal melanonychia, other mucocutaneous inflammatory disorders, syndromic associations.¹⁸

Onychoscopy has become an indispensable tool in the evaluation of pigmented nail lesions as it

helps in validating the clinical findings to differentiate the melanin and nonmelanin pigmentations.³⁰ It is important to identify subungual hematoma by the history of trauma and characteristic onychoscopic appearance. A subungual hematoma is identified by globules of various sizes and color-from bright red, brown to black depending upon the depth and duration of hemorrhage. However, it should be remembered that subungual hemorrhage does not rule out melanoma. Determine the cause of pigmentation—melanocytic activation or proliferation. Distinguish benign and malignant proliferations.³¹

Whenever there is a doubt of NUM on clinical features and onychoscopy, histopathological diagnosis is the gold standard for confirmation of NUM. The preoperative diagnostic accuracy of melanoma ranges from 46% to 55%.³² The type and site of biopsy depends upon the morphological characteristics of LM-its width and location of pigmentation in the matrix. The biopsy sample should be of good quality, representative, adequate and from right location for proper diagnosis. An excisional biopsy, whenever possible is recommended and matricial origin of pigmentation should be removed completely. Full-thickness matrix biopsy avoids any misdiagnosis and aids in determining the prognosis by assessing Breslow's depth. Nail matrix biopsy can cause nail scarring and deformity. To minimize these, distal nail matrix biopsy is preferred over proximal, and the resultant defect of >3 mm should be sutured.³³

Treatment of melanonychia depends on the underlying cause. The treatment of associated systemic or locoregional disease, withdrawal of offending drug, avoidance of trauma, treatment of infections or correction of nutritional deficiency

cies may cause regression of pigmentation. Benign cases do not necessitate treatment and can be kept in follow up. Depending on thickness and histopathological characteristics, subungual melanoma may be managed by functional surgical treatment (wide local excision) or digit amputation, with or without sentinel lymph node mapping/biopsy.³⁴

Prognosis of melanonychia depends on its etiology and its benign or malignant nature. Benign lesions can be followed up, while NUM may carry a poor prognosis. There is still no consensus for follow-up of melanonychia which requires periodic medical examinations and photographic and dermoscopic documentation. Onychoscopy can be used for follow up visits, but there are no precise criteria for its frequency in patients with LM. Good-quality clinical as well as onychoscopic photographs highlighting the involved nail and the pigment morphology, color, and extent is necessary for follow ups. Patients should be counselled for self-examination and to report whenever any morphological change in pigmentation is noticed.³⁴

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