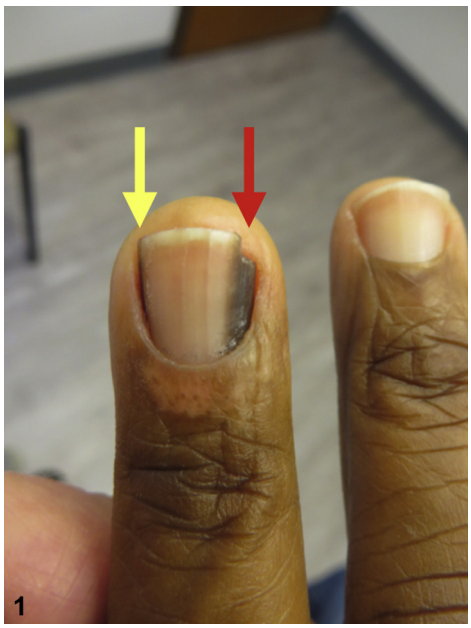
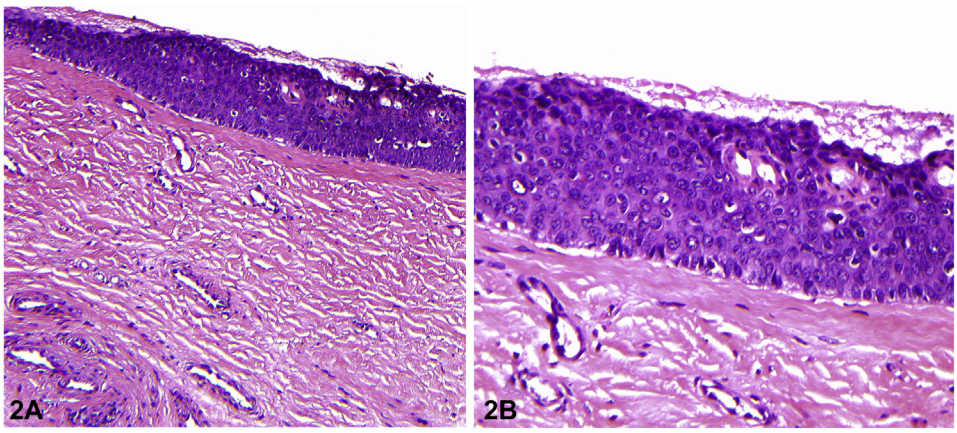
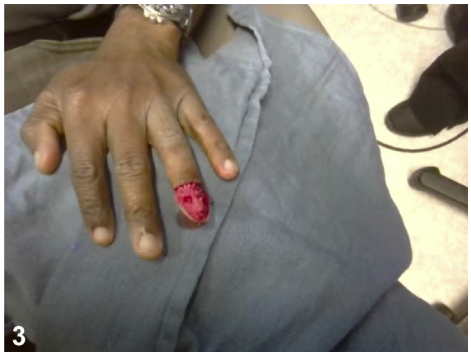
Two discrete bands of longitudinal melanonychia on one fingernail

Anne Y. Ning, BA,[a] Melissa A. Levoska, MD,[b] Kord Honda, MD,[b] Jeremy S. Bordeaux, MD, MPH,[b] and Christina Wong, MD[b]
Cleveland, Ohio







A 52-year-old African-American man with no history of immunosuppression or human papillomavirus infection presented with a 5-month history of a 3.5-mm brown longitudinal band on the medial left fourth digit nail and a 2-month history of a 2-mm brown band on the lateral aspect of the same nail (Fig 1). Hypopigmentation proximal to the lateral nail fold was noted with negative Hutchinson sign. He was treated with topical econazole for 1 month without improvement. A nail fungal culture was negative. He reported no local trauma and no personal or family history of skin cancer. Two tangential shave biopsies of the nail matrix were performed, one from each band (Fig 2).

Question 1: Given the clinical picture (Fig 1) and histology (Fig 2) shown, which of the following is the best diagnosis?

A. Junctional nevus

B. Squamous cell carcinoma in situ (SCCis)

C. Hemorrhage

D. Melanoma in situ

E. Lentigo

Answers:

A. Junctional nevus — Incorrect. Junctional nevi of the nail matrix can present as longitudinal melanonychia (LM) but usually develop during childhood.[1] Biopsy would show melanocyte nests in the basal layer of the epidermis at the dermoepidermal junction.

B. SCCis — Correct. LM is an uncommon presentation of nail unit SCCis. Risk factors for nail unit SCCis include radiation exposure, trauma, immunosuppression, and high-risk human papillomavirus infection. The disease typically progresses slowly and can be misdiagnosed as a benign nail injury, bacterial infection, or melanoma. Presence of nail plate splitting, single digit involvement, rapid growth, or heterogeneity can be concerning for malignancy. In our patient, biopsies were taken sampling both the nail matrix and the nail bed of both pigmented bands. Histopathology showed full-thickness keratinocyte atypia without invasion, diagnostic of SCCis (Fig 2). SOX-10 staining was negative for increased melanocytes.

C. Hemorrhage — Incorrect. Trauma can lead to accumulation of blood under the nail plate, creating a subungual hematoma. This is generally associated with pain secondary to increased pressure under the nail plate. Histopathology shows erythrocyte extravasation without inflammation.

D. Melanoma in situ — Incorrect. Nail unit melanoma is a rare cause of LM and is associated with a poor prognosis because of delayed diagnosis. Concerning features include the ABCDEs for melanoma (asymmetry, irregular borders, heterogeneous color, diameter > 6 mm, and evolution) and a positive Hutchinson sign (periungual pigmentation). Histopathology of melanoma in situ shows melanocyte hyperplasia with nuclear atypia without invasion of the dermis on hematoxylin-eosin staining and an increased number of melanocytes with SOX-10 immunohistochemistry.

E. Lentigo — Incorrect. Lentigo has been reported to cause 9% of LM cases in adults.[1] On histopathology, a lentigo is identified by a mild increase in matrix melanocytes without cellular atypia or nests.

Question 2: Which of the following is the best next step in management for this patient?

A. Amputation

B. Surgical excision

C. Mohs micrographic surgery (MMS)

D. Cryotherapy

E. Topical 5-fluorouracil

Answers:

A. Amputation — Incorrect. Amputation should be considered only if there is bony involvement or recurrence after more conservative treatment such

From Case Western Reserve University School of Medicine[a] and the Department of Dermatology, University Hospitals Cleveland Medical Center, Case Western Reserve University.[b]

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Correspondence to: Christina Wong, MD, Department of Dermatology, University Hospitals Cleveland Medical Center, Case Western Reserve University, Lakeside 3500, 11100 Euclid Ave, Cleveland, OH 44106. E-mail: christina.wong2@uhhospitals.org.

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as MMS. In general, digit preservation is important to keep in mind when treating SCC of the nail.

B. Surgical excision — Incorrect. Surgical excision of subungual SCC has been reported to have a 4% to 56% rate of local recurrence for cases without evidence of bony invasion. It is a more aggressive approach to treatment and can be considered when MMS is not available.[2]

C. MMS — Correct. Performing MMS on the nail unit can be difficult and time consuming but has been associated with high cure rates and maximum tissue sparing.[3] Although there are currently no standardized guidelines for treatment of nail unit SCC, the literature suggests MMS should be the primary method of treatment.[2] Our patient did not have evidence of bony invasion or metastatic disease and was treated with MMS of the nail bed and nail matrix with healing by secondary intention (Fig 3). Three months after surgery, there was no evidence of recurrence or complications.

D. Cryotherapy — Incorrect. Because the nail plate overlies the nail matrix where SCC is found, cryotherapy is not an effective treatment for nail unit SCC, as it would be difficult to penetrate the nail plate.

E. Topical 5-fluorouracil — Incorrect. Topical 5-fluorouracil is a highly effective therapy for SCC in situ of the skin. However, topical therapy for nail unit SCC is not effective because it does not penetrate the nail matrix.

Question 3: What is the most likely mechanism underlying this patient's melanonychia?

A. Trauma

B. Melanocytic proliferation

C. Melanocytic activation

D. External pigment deposition

E. Inflammation

Answers:

A. Trauma — Incorrect. Although trauma can be a risk factor for development of nail unit SCC, it does not explain increased pigment causing melanonychia. Melanonychia secondary to trauma is generally associated with nail plate dystrophy, which was not seen here. Furthermore, the patient denied any history of trauma to the area.

B. Melanocytic proliferation — Incorrect. Melanocytic proliferation is seen in melanonychia caused by nevi or melanoma. Depending on the diagnosis, there are varying degrees of nuclear atypia and melanocytic extension into the dermis. Our patient's histopathology did not show evidence of melanocytic proliferation.

C. Melanocytic activation — Correct. Melanocytic activation, or hypermelanosis, is the most common cause of melanonychia and is consistent with a normal number and distribution of melanocytes seen on histopathology.[1] Hyperpigmentation may not be evident on hematoxylin-eosin stain and often requires special staining for identification (ie, Fontana-Masson, Melan-A, S-100).[4] Development of pigmented SCC is thought to be secondary to increased expression of stem cell factor, possibly secondary to production of promelanocytic factors by tumor cells.[5]

D. External pigment deposition — Incorrect. Nonmelanocytic causes of nail pigmentation include external nail pigmentation, which can occur as a result of tobacco, cosmetics, and topical medications including silver nitrate. However, pigmentation secondary to exogenous pigment is not known to result in LM.

E. Inflammation — Incorrect. Inflammation can cause transient melanocytic activation. Infections and dermatoses like psoriasis may result in melanonychia that typically disappears after the inflammation has resolved.[4] The lack of keratinocyte atypia on histopathology is more consistent with a diagnosis of SCC.

Abbreviations used:

LM: longitudinal melanonychia
MMS: Mohs micrographic surgery
SCCis: squamous cell carcinoma in situ

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