A Step-Wise Approach to Longitudinal Melanonychia

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Presenter Disclosures

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The following relationships with commercial interests related to this presentation existed during the past 12 months:

Senior Vice-President, Podiatric Medical Education
Sagis Diagnostics
Division of Podiatric Pathology
Planned talking points:

1) Define longitudinal melanonychia
2) Characteristics
3) Anatomy and physiology of nail growth and melanocyte deposition in nail units
4) Relevance
5) Epidemiology
6) Dermoscopy findings
7) Biopsy techniques
Longitudinal Melanonychia

• Deposition of melanin in the nail plate derived from many causes:
  • Moles
  • Melanotic macule – hyperplasia and activation
  • Trauma
  • Pregnancy
  • Drugs
  • Addison’s disease, Cushing’s
  • Many others……and

• Melanoma
Nail Unit Melanoma

- 30% of malignant melanoma of the nail unit have a preceding history of LM
- Many findings of LM and malignant melanoma are similar
- Criteria for biopsy are not well-defined.
- Color
- Width
- Rapidly expanding or sudden onset of LM
- Fuzzy borders
- Variegated colors
- Hutchinson's sign pathognomonic of malignant melanoma if other causes can be eliminated.
- Possibly over 50,000,000 Americans with LM
Nail Anatomy

Good relationship anatomy but fat layer exaggerated
Hallux nail with three longitudinal pigmented streaks, medial, central, and lateral of varied width and pigment intensity
Presentations of longitudinal melanonychia can occur in multiple nails in the same patient.
This is the ventral surface of a nail plate removed during a phenol procedure for a chronically painful nail. The arrow points to a medially based totally ventral longitudinal melanonychia, which was not visible through the dorsal plate. This caused me to search for a lesion in the distal matrix which was evident exactly where it would be expected.

Distal lunula melanocytic lesion corresponding to the ventral nail plate pigmentation.
Physiology*

- In the proximal nail matrix, melanocytes are mostly located in the lower second to fourth cell layers, as opposed to the distal matrix, where they are more superficial in the first and second cell layers.
Physiology*

- Many proximal matrix melanocytes do not produce melanin, but approximately 50% of distal matrix melanocytes do produce melanin.
Physiology*

• Melanocytes in the nail bed (distal to lunula) are least numerous and do not synthesize melanin, which explains why nail bed melanomas are often amelanotic and present a more difficult challenge for timely diagnosis.

*Ruben, B; Pigmented Lesions of the Nail Unit: Clinical and Histopathologic Features; Seminars in Cutaneous Medicine and Surgery; Elsevier, 2010, pp148-158
Nail Biopsies
The nail biopsy is not just one technique but a variety of techniques, each of which plays a specific role in diagnoses.

1. **Nail Plate**, Distal Clipping
2. **Longitudinal**
3. **Nail Bed**, Longitudinal Elliptical Excision
4. **Nail Bed**, Punch
5. **Nail Matrix**, Transverse Elliptical Excision, Distal Matrix
6. **Nail Matrix**, Punch, Distal Matrix
7. **Nail Matrix**, Transverse Elliptical Excision, Proximal Matrix
8. **Nail Fold**, Punch, Shave or Excision

StatPearls Publishing Illustration; Source: Adam Levin, MD.
BAD? or GOOD? news for the DPM

- **Acral** melanoma, defined as melanoma involving the palms, soles and nail units, has a worse long term prognosis than non acral melanoma*

Amelanotic melanoma

Hutchinson’s sign
Nail Growth Mechanics

Long Matrix = thicker nail plate
Relative melanocyte synthesis and location in nail unit
Epidemiology and Clinical Criteria

• Melanoma of the nail unit is relatively rare, with an incidence of about 1% to 3.5% of melanoma in white-skinned individuals.

• It occurs equally in dark skin and fair skin individuals, and it is the most common type of melanoma diagnosed in deeply pigmented individuals, mostly diagnosed between the age of 40 and 70.

• Further complicating this issue is the fact that in darker skinned individuals, longitudinal melanonychia is commonly seen in one or multiple nails, often referred to as ethnic or racial pigmentation, and is mostly benign.

• The clinical presentation of pigmented streaks arising from benign versus malignant lesions is not as easily differentiated as for melanoma elsewhere on the skin, where the well-known and well taught ABCDE criteria for the most part is reliable.
When should we be suspicious? Clinical Hints:
There should be a heightened index of suspicion for melanoma when longitudinal melanonychia:**

- Begins in a single digit of a person during the sixth decade of life or later
- Develops suddenly in a previously normal nail plate
- Changes including darkening or widening
- Occurs in either the thumb, index finger, or hallux
- Occurs attendant to a history of digital trauma
There should be a heightened index of suspicion for melanoma when longitudinal melanonychia:**

- Occurs as a single band in the digit of a dark skinned individual, especially if in the thumb or hallux
- Appears with blurred as opposed to sharp lateral borders
- Occurs in a patient with a prior history of melanoma
- Occurs in a patient identified as having increased risk for developing melanoma, such as a patient with family history of melanoma or dysplastic nevus syndrome
- Is seen with nail dystrophy, destruction, or partial nail plate absence

When should we be suspicious? Clinical Hints:

• Levit, et al., after reviewing the world literature on subungual melanoma, developed a set of criteria for the clinical detection of subungual melanoma in an attempt to enable clinicians and the public to identify longitudinal melanonychia that should be considered for biopsy. The result was the ABCDEF of subungual melanoma as follows:

ABCDEF rule of subungual melanoma

- **A**: Age – range 20-90 years, peak 5th - 7th decade
  - African-American, Native American, Asian races
- **B**: Band: Pigment is Brown-Black. Breadth ≥ 3mm.
  - Border is irregular or Blurred
- **C**: Change: Rapid increase in size or growth rate of band.
  - Lack of Change: Failure of nail dystrophy to improve with treatment
ABCDEF rule of subungual melanoma

- **D**: Digit involved: Thumb most common-then **Hallux**-then Index finger. Single digit more worrisome than multiple digits. **Dominant** hand more common than non-dominant hand.

- **E**: Extension: Extension of pigment on to the proximal or lateral nail fold (Hutchinson’s sign) or free edge of nail plate.

- **F**: Family or personal history: Previous melanoma or dysplastic nevus syndrome.
Sawada Dermoscopy Study*
Identify suspicious bands at in-situ stage and determine follow up required

• Classified lesions into types I, II or III mostly based on depth of color of the band, background color, and presence of pigment on periungual skin.
• Type I lesions were clinically and dermoscopically benign and observed
• Type II intermediate and observed
• Type III lesions were considered suspicious and biopsied
Sawada Study Results – 5 years

• **Type III** lesions showed 100% in-situ melanoma
• 10% (5 lesions) of **Type II** lesions showed change. 2 out of the 5 were in-situ melanoma
• None of the **Type I** lesions showed any change

Conclusion: **dermoscopy** of nail lesions can be **very useful**, and three month follow up for Type II lesions to look for change is recommended.

Arch Dermatol 2010; 146:382-387

Conclusion: “overall accuracy of dermatologists in the diagnosis of nail matrix in-situ melanoma using dermoscopy and clinical criteria is low, confirming that biopsy is still the gold standard for diagnosis of these lesions.”
# The ABCDEFs of nail melanoma

<table>
<thead>
<tr>
<th>Letter</th>
<th>Meaning: Subungual</th>
<th>Meaning: Cutaneous</th>
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</table>
| A      | Age: Peak 5th-7th decades  
Race: African American, Native American, Asian | Asymmetry |
| B      | Band (nail band): Brown-black  
Breadth: ≥3 mm  
Border: Irregular/blurred | Border: Irregular/poorly defined |
| C      | Change: Rapid increase in size | Color: Varied |
| D      | Digit involved: Thumb > hallux  
Single digit > multiple digits | Diameter: >6 mm |
| E      | Extension of pigment to involve nail folds | Evolving (changing) |
| F      | Family or personal history: Previous melanoma or dysplastic nevus syndrome | Funny looking (ugly duckling) |


Source: Dr. Lipner
What is best way to biopsy LM?

What is agreed on?

1) Every presentation of LM should initially raise concern, certainly in Caucasians. However in dark skinned races, acral location of melanoma (not incidence) is more common and therefore especially important for podiatrists

2) LM in children representing melanoma is very rare

3) Adequate tissue sampling and procedure choice depends on location and size of the pigmented band

4) Consideration of discomfort, cost, recovery time and resulting nail cosmesis also play a role

5) Proximal nail matrix more sensitive to cosmetic disruption than distal matrix

6) 3mm punch biopsy of lesions < 3mm in width shown to be adequate for sampling and cosmesis. 3mm punch for wider lesions risks sampling error and leaves lesion behind.

MATRIX SHAVE MAY BE PROCEDURE OF CHOICE FOR ALL BUT DISTAL MATRIX LESIONS SMALLER THAN 3MM
Study validated that tangential shave provided enough tissue for accurate diagnosis
Immediate Post-Op Matrix Shave Technique
11 Days Post-Op
19 Days Post-Op
29 Days Post-Op
78 Days Post-Op
Streak resolved
One year post-op
In Situ Melanoma

10 Year hx of slowly enlarging LM
Previously Amended
Reason For Amendment #1: Removal of extraneous information from comment field

SPECIMEN SOURCE:
A. RIGHT HALLUX LATERAL NAIL PLATE AND MATRIX

DIAGNOSIS:

Segment of nail plate and bed, and matrix with melanocytic hyperplasia.
PAS stain is negative for organisms. Masson Fontana is strongly positive. Anti-Melan-A and SOX-10 show a considerable hyperplasia of melanocytes. The number is greater than 30 per mm linear distance. This is an atypical feature. An additional consultation is pending.

Robert G. Phelps, MD
Attending Pathologist, Dermatopathology

Addendum 1:
The slides reviewed again at consensus and given the considerable hyperplasia of melanocytes, re-excision to ensure complete removal with adequate margins of the lesion is recommended. An additional outside consultation is also pending.
SURGICAL PATHOLOGY  MSHTAM
COLLECTION DATE: 09/06/2019
Dermatopathology Report
SPECIMEN SOURCE:
A. LATERAL NAIL FOLD RIGHT HALLUX
DIAGNOSIS: **Residual compound nevus and dermal scar**, recent.
Immunohistochemical studies reveal positive staining with Mel-A confirming the diagnosis.
Note: The case was reviewed in the dermatopathology daily intradepartmental conference with Dr. Phelps and the members present concur with above diagnosis.
Rajendra S. Singh, MD
Attending Pathologist, Dermatopathology
PUNCH BIOPSY THROUGH INTACT NAIL
Biopsy of LM and Growth Disturbance ?
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Volar skin dermoscopy patterns:

- Parallel furrow:
- Fibrillar:
- Lattice:
- Parallel ridge:

MALIGNANT
Nail unit dermoscopy patterns

Lentigo: Increased pigment, melanocytes do not form nests. Melanocyte number may be increased.

Nevus: Melanocytes forming nests, may or may not be pigmented.
Onychomycosis (A) Overview. (B) Dermoscopy. The whitish brightening with parallel striation, proximal jags, and nail discolorations with yellow colors are dermatoscopically detectable. (Copyright: ©2014 Haenssle et al.)
Subungual hematoma after trauma.

(A) Overview. (B–C) Dermatoscopy. The characteristic red-blue to blue-black homogeneous color, numerous satellite droplets towards the distal nail edge and the jagged margins are depicted. (Copyright: ©2014 Haenssle et al.)
Ethnic lentigo of the right great toenail.

(A) Overview. (B) Dermatoscopy. Dermatoscopically there is a homogeneous gray-ish-pale longitudinal melanonychia affecting the whole nail with a slightly diffuse demarcation of the lateral margin.

Benign nevus over the course of time (interval of approximately six months). (A) Overview. (B) Dermatoscopy. Due to an irregular, intensive brownish striation a digital dermatoscopic follow-up examination was performed. The follow-up image reveals a decrease in pigmentation that may vary with the level of sun-exposure and a constant width of the longitudinal melanonychia. (Copyright: ©2014 Haenssle et al.; first published in Der Hautarzt, 2014)
Subungual melanoma in situ. (A) Overview. (B) Dermatoscopy. The whole nail unit is affected by a continuous pigmentation composed of parallel lines showing an inhomogeneous color (brown to blue-gray) and pigment intensity (unpigmented followed by heavily pigmented streaks). A Hutchinson sign, being indicative of an invasion of melanoma cells into the periungual skin, is better visualized by dermatoscopy (arrows at proximal nail fold). (Copyright: ©2014 Haenssle et al.)
Early invasive subungual melanoma, thickness 0.2 mm. (A) Overview. (B) Dermatoscopy. Dermatoscopically, a homogeneous gray-brown band is visible, measuring approximately 6 mm across and continuously affecting the whole nail (longitudinal melanonychia). Sharply demarcated globular structures correspond to drop-like subungual hemorrhage. The macro-Hutchinson sign, being indicative of an invasion of melanoma cells into the periungual skin, is (still) negative (proximal nail fold). (Copyright: ©2014 Haenssle et al.)
Advanced subungual melanoma in a 30-year old male with skin type V (Indian skin type). (A–B) Overview. (C–D) Dermatoscopy. The complete nail unit and also the adjacent skin are heavily pigmented with black-brown colors. The nail plate shows a severe onychorrhexis with multiple longitudinal fissures and ridges. The macro-Hutchinson sign is positive at the proximal nail fold as well as at the distal tip of the toe. (D) Dermatoscopy of the distal tip of the toe reveals the typical parallel ridge pattern of acral melanoma. (Copyright: ©2014 Haensssle et al.)
Far advanced subungual melanoma of the right great toe. (A) Overview. (B) Dermatoscopy. The complete nail unit is destroyed by a black to pink colored, ulcerated melanoma. Dermatoscopy reveals black-blue to red colors with a cloud-like texture. (Copyright: ©2014 Haenssle et al)
Nail apparatus acral lentiginous melanoma, Clark level III (1.7-mm Breslow thickness) in a 57-year-old woman. Dermoscopy disclosed a brown coloration of the background (long arrow) and darker longitudinal lines with irregular thickness, coloration, and spacing (double arrows). Disruptions of parallelism were also observed (short arrows).
Nail apparatus acral lentiginous melanoma, Clark level II (0.9-mm Breslow thickness), in a 71-year-old man. Dermoscopy shows a brown background and longitudinal darker lines of irregular thickness, spacing, and coloration (arrows).
Nail apparatus acral lentiginous melanoma in situ, Clark level I (0.2-mm Breslow thickness), in a 72-year-old woman. A, The clinical view shows no evidence of pigment in the cuticle area; B, the dermoscopic image shows irregular lines overlying a brown background and a micro-Hutchinson sign (arrow).
Hydroxyurea-induced longitudinal melanonychia in an 81-year-old man treated for chronic lymphocytic leukemia. Dermoscopy shows a grayish coloration of the background (long arrows) and thin regular longitudinal gray lines (short arrows).
Nail apparatus lentigo in a 37-year-old woman with Laugier-Hunziker disease. Dermoscopy reveals a brown coloration of the background (long arrow) and mostly regular, thin, longitudinal gray lines (short arrows).
Ethnic-type nail pigmentation in a 42-year-old man of Gypsy lineage. Dermoscopy reveals a grayish coloration of the background and longitudinal microscopic grooves either associated (short arrows) or not (long arrow) with the pigmentation.