Melanonychia in children and adults: Clinical evaluation including advanced imaging techniques

Barriers to arriving at a diagnosis
- The nail plate covers the source of the pigmentation
- Clinical cues can be misleading
  - Overlap of findings in benign and malignant lesions
- There may be a lack of familiarity with nail anatomy and biopsy techniques
- Suboptimal specimens may result
- Most pathologists lack familiarity with normal nail unit histology and pathology
- Diagnostic criteria are used inconsistently, in evolution

Objectives
- Discuss the clinical evaluation of pigmented nail bands
  - Clinical signs (ABCDE rule)
  - Dermatoscopy (“Onychoscopy”)
  - Intraoperative dermoscopy
- Explore the future prospects for clinical assessment
  - Reflectance confocal microscopy
  - Optical coherence tomography
Key clinical information in evaluating longitudinal pigmented bands

- Solitary or multiple bands
- Width of the band
- Color and homogeneity of the band
- Hutchinson's sign (periungual pigmentation)
- Duration, and any change over time
- Which digit affected
- Age of the patient
- Any extenuating clinical history (drugs, pregnancy, baseline pigmentation)

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Table 2: Causes of Nail Pigmentation

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Melanocytic activation/hypermelanosis</td>
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<tr>
<td>Trauma</td>
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<tr>
<td>Medications</td>
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<tr>
<td>Racial pigmentation</td>
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<td>Endocrinopathy</td>
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<tr>
<td>Pregnancy</td>
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<tr>
<td>Pseudo-Jaehara and Leser-Huneker syndromes</td>
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<tr>
<td>Disruptions to the nail unit, including inflammatory and neoplastic conditions</td>
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<tr>
<td>Melanocytic proliferations</td>
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<tr>
<td>Lentigo</td>
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<tr>
<td>Nevus</td>
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<tr>
<td>Melanoma</td>
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<tr>
<td>Other</td>
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<tr>
<td>Nail hemorhage</td>
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<tr>
<td>Pigmented squamous cell carcinoma</td>
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<tr>
<td>Pigmented atypical nevus</td>
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</tbody>
</table>

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Melanocytic activation/hypermelanosis

Lentigo

Melanocytic nevus
Nail apparatus melanoma (NAM)
- Most occur in thumb and great toe
- Higher mortality than conventional melanoma
- Mean tumor thickness 4.8 mm at dx.
- May arise in matrix or rarely nail bed (amelanotic)
- Irregular pigmented band, greater than 3 mm
- Hutchinson’s sign (periungual pigmentation)
- Exceedingly rare in children


ABCDEF rule for nail unit melanoma
- A: Age (50-70 peak); also ? race (African Americans, Asians, Native Americans)
- B: Band (black to brown); Breadth > 3 mm; Blurred borders
- C: Change (enlarging or darkening; lack of change/response to treatment for nail dystrophy)
- D: Digit (thumb>hallux>index finger; single versus multiple; dominant hand)
- E: Extension of pigment onto surrounding tissues
- F: Family or personal history of melanoma

Melanonychia in children

- Does not play by the rules!
- Nevus and lentigo are most common; MIS is rare
- Nevus may be broad and involve periungual skin, may have blurred borders
- Nevus may be unstable:
  - Darkening, fading, enlargement of the band and appearance of nail plate abnormalities
- Dermatoscopy may display abnormal findings
- Histology may also be disturbing

Subungual hemorrhage

Onychoscopy for melanonychia

- Useful for distinguishing hemorrhage from melanin
  - Caveat: Melanoma may produce hemorrhage
- Benign: brown background with regular parallel lines of identical color, spacing and thickness
  - Caveat: Rarely observed!
- Melanoma: a brown background with longitudinal lines, irregular in color, thickness, spacing, loss of parallelism is most reliable pattern
  - Caveat: Nevus in children and sometimes in adults can have irregular lines
  - An individual irregular line is very suspicious
  - Granular pigmentation is more common in melanoma
  - Helpful to see micro Hutchinson’s sign

Melanocytic activation (hypermelanosis)

Melanocytic nevus

Courtesy of Nilton Gioia DiChiacchio, MD

Melanoma in situ

Onychoscopy in children - NEVI!

- No contact of the device with the wound
  - Courtesy of Nilton Gioia DiChiacchio, MD

Patterns of nail matrix and bed of longitudinal melanonychia by intraoperative dermatoscopy

- Expose matrical area containing the lesion
- Use DermLite polarized light dermatoscope, no contact needed
- Biopsy and perform routine microscopy on all cases

4 dermatoscopic patterns were identified:

- Regular gray pattern (hypermelanosis)
- Regular brown pattern (benign melanocytic hyperplasia/lentigo)
- Regular brown pattern with globules or blotches (melanocytic nevus)
- Irregular pattern (melanoma)

Established and validated patterns for intraoperative dermatoscopy of the nail matrix and bed, in 100 consecutive cases

Patterns of nail matrix and bed of longitudinal melanonychia by intraoperative dermatoscopy

- Intraoperative dermatoscopy
  - Courtesy of Nilton DiChiacchio, MD, PhD

Intraoperative dermatoscopy

- Regular gray pattern: presence of fine, regular, grayish lines (HYPERMELANOSIS)

- No contact with the wound

- Courtesy of Nilton Gioia DiChiacchio, MD
Patterns of nail matrix and bed of longitudinal melanonychia by intraoperative dermatoscopy

Hirata SH, Yamada S, Enokihara MI, Di Chiacchio N, de Almeida FA, Enokihara MMSS, Michalany NS, Zaiac M, and Tosti A.

J Am Acad Dermatol 2011;65:297-303

Regular brown pattern: regular brown lines. (BENIGN MELANOCYTIC HYPERPLASIA)

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Regular brown pattern with globules: regular longitudinal brown lines and presence of globules with regular size and distribution. (MELANOCYTIC NEVI)

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Hirata SH, Yamada S, Enokihara MI, Di Chiacchio N, de Almeida FA, Enokihara MMSS, Michalany NS, Zaiac M, and Tosti A.

J Am Acad Dermatol 2011;65:297-303

Irregular pattern: longitudinal lines of irregular color and thickness, presenting irregular globules and blotsches. (MELANOMA)

Patterns of nail matrix and bed of longitudinal melanonychia by intraoperative dermatoscopy

Hirata SH, Yamada S, Enokihara MI, Di Chiacchio N, de Almeida FA, Enokihara MMSS, Michalany NS, Zaiac M, and Tosti A.

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High degree of sensitivity and specificity for NMD when compared with NPD upon histopathologic examination

Table 4. Nail plate dermatoscopy and intraoperative nail matrix and bed dermatoscopy patterns according to histopathologic diagnosis

<table>
<thead>
<tr>
<th>Histopathology</th>
<th>Nail plate dermatoscopy</th>
<th>Intraoperative dermatoscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Melanocytic activation (N = 40)</td>
<td>3 12 0 0 0</td>
<td>3 12 0 0 0</td>
</tr>
<tr>
<td>Benign melanocytic hyperplasia (N = 15)</td>
<td>0 15 0 0 0</td>
<td>0 15 0 0 0</td>
</tr>
<tr>
<td>Melanocytic痣 (N = 25)</td>
<td>3 9 0 0 0</td>
<td>3 9 0 0 0</td>
</tr>
<tr>
<td>Melanoma (N = 15)</td>
<td>4 6 0 0 0</td>
<td>4 6 0 0 0</td>
</tr>
</tbody>
</table>

Melanocytic activation (hypermelanosis)

After removal

Dermoscopy of the specimen on the filter paper

Intraoperative dermoscopy to confirm complete removal

High degree of sensitivity and specificity for NMD when compared with NPD upon histopathologic examination
Can newer imaging techniques be useful in nails?

- Reflectance confocal dermoscopy

- Confocal microscopy of normal nail

- Confocal microscopy of onychomycosis

- Confocal microscopy of nail unit melanoma
Confocal microscopy for melanonychia

- Need to expose matrix (invasive)
- Dynamic exam at many levels
- One study showed some ability to differentiate benign lesions from melanoma (only 9 cases; MIS and thin MM versus lentigo, one equivocal)
  - Ex vivo exam was better; ? Histologic dx of MIS in some cases
- Intraoperative diagnosis to allow for one definitive procedure
- Helpful to differentiate other causes of melanonychia (fungal, SCCIS)
- ?Helpful in low cellularity early or hyperpigmented lesions


Confocal microscopy for melanonychia

- Optical coherence tomography
  - A laser-based technology, first used in skin in 1997
  - Resolution greater than 1 mm depth
  - Dynamic or speckle variance OCT has better resolution; microvasculature patterns used for diagnosis
  - Captures multiple images, stacks them, 10-20 sec to scan, processing software creates a coherent image in different planes


Lentigo (ex vivo)

- Main use is for non-MM skin CA
- Some inflammatory disease: psoriasis (inc. nail), scleroderma
- Possible use for melanocytic lesions, perhaps studying microvasculature (not ready for prime time)


Melanoma in situ

- Main use is for non-MM skin CA
- Some inflammatory disease: psoriasis (inc. nail), scleroderma
- Possible use for melanocytic lesions, perhaps studying microvasculature (not ready for prime time)


Optical coherence tomography

Clinical evaluation methods of melanonychia

- ABCDEF rule
- Dermatoscopy (onychoscopy)
- Intraoperative dermatoscopy
- Confocal microscopy
- Optical coherence tomography

Limitations

- Direct methods still invasive as nail matrix is relatively hidden
- Overlapping features of benign and malignant, especially in early lesions and in children
- High tech modalities need more validation, are costly
Microscopic diagnosis.....

Thank you!