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“Not Normal Nails”

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Disclaimer

- No conflict of Interest
- No Financial Associations
- Off-label medications may be discussed
- Honorarium: SDPA
Objectives

• Learn disease affecting the nail unit
• Learn nail unit diseases associated with systemic disease
• Learn therapeutic modalities for nail unit disease
• Be able to differentiate normal nails from “not normal nails”
What’s a normal Nail?
What’s an abnormal Nail?
Definitions of Abnormal Nails

- Anonychia - absence of nail
- Brachyonychia - short nails
- Chromonychia - color changes in the nail unit
- Clubbing - increase in the ungula-phalangeal angle >180
- Fragilitas unguium - brittle nails
- Hapalonychia - soft nails
- Koilonychia - spoon nails
- Leukonychia - whitening of nail plate
- Macronychia - large nail wider than normal
- Micronychia - small nail shorter and narrower
- Onychalgia - nail pain
- Onychauxis - hypertrophied nail plate
- Onychia - inflammation of nail unit
- Onychoclavus - subungal corn
- Onychodystrophy - abnormal nail
- Onychogryphosis - ram horn nails
- Onychoheterotopia - abnormally placed nail from displaced nail matrix
- Onycholysis - separation

- Onychomadesis - proximal separation of nail plate from nail bed
- Onychomalacia - soft nails
- Onychomycosis - nail fungus
- Onychophagia - nail biting
- Onychophosis - hyperkeratosis of nail folds
- Onychoptosis - loss of nail plate
- Onychorrhexis - longitudinal ridging/striations of nail plate
- Onychoschizia - splitting of nail plate into layers
- Onychotillomania - tearing, picking or destroying nails
- Pachyonychia - thickening of entire nail
- Paronychia - inflammation of nail folds
- Polynychia - more than one nail on a digit
- Pterygium - scarring of the PNF/matrix (See Tables at end)
- Pterygium inversus - scarring of distal nail plate to the hyponychium
- Trachyonychia - rough nails
- Unguis/Unguium - nail/nails
- Unguis incarnates (onychocryptosis) – ingrown nail
Nails and Skin Disease

- Psoriasis
- Lichen planus
- Alopecia areata
- Dermatitis

A hand surgeon's guide to common onychodystrophies.
Fowler JR¹, Stern E², English JC 3rd², Goitz RJ¹.
Nail Psoriasis

• Key Points:
  – 50-80% of cases have nail involvement
  – Severe nail involvement may not imply severe skin disease
  – Affects quality of life, interferes with work, painful, ugly
  – Nail Features:
    • Pitting
    • Dyschromia
    • Subungual hyperkeratosis
    • Dystrophy and splinter hemorrhages
  – Pustular Variant
  – Acro-osteolysis/Arthritis
  – Treatment:
    • Topical and oral retinoids, MTX, Cyclosporine, Biologics
Nail Lichen Planus

• Key Points:
  – 2-16% of LP patients; Only 25% of Nail LP have skin lesions
  – Permanent dystrophy in up to 4%
  – Nail features:
    • Pitting
    • Pterygium
    • Ridging, fissuring, fragility
    • Atropy
  – Treatment:
    • Corticosteroids, Tacrolimus, Tazarotene
    • Oral retinoids
Pterygium

Dorsal
Pterygium
Cicatricial pemohigoid
Diabetic vasculopathy
Onychotillomania
Raynaud’s disease
Radiodermatitis

Ventral
Pterygium
SLE
Systemic sclerosis
Allergic contact
Nail Alopecia Areata

Key Points

- Up to 25% of AA pts have Nail AA
- More common in more severe cases

Nail Features

- Pitting
- Trachyonychia
- Onychoschizia
- Red Lunula (Diff Dx: CTD, Endocrine, Renal)

Treatment

- Oral and IL corticosteroids, MTX, Cyclosporine, Cell Cept
Nail Dermatitis

• Key Points:
  – Nail matrix involvement with subsequent abnormal nail production
  – ACD/ICD, Chronic Vesicular Hand Dermatitis
  – Nail Features:
    • Beau’s lines
    • Onychomadesis
    • Periungual inflammation and scaling
  – Treatment:
    • Avoid allergen/irritant, Topical Corticosteroids, MTX
Nail Question 1

• Which of the following is the best diagnosis?

  Patient failed oral antibiotic treatment.

  A. Lichen planus
  B. Pityriasis rubra pilaris
  C. Pustular psoriasis
  D. Bacterial paronychia
  E. None of the above
Nails and Systemic Disease

- Lines and Nails
- Koilonychia
- Clubbing
- Onycholysis
- Periungual Telangiectasia
- Nail Degloving
Lines and Nails

• “…. Lines” are a plate phenomenon
  – Beau’s lines
    • Nail plate transverse groove
    • Growth arrest due to systemic illness/drug
  – Mees’ Lines
    • Nail plate transverse leukonychia
    • Arsenic, infections, systemic illness
  – Bissells’ Lines
    • Longitudinal melanonychia (striata)
    • Addison’s disease

• “…. Nails” are a nail bed phenomenon
  – Muehrcke’s Nail
    • Nail bed edema from hypoalbuminemia giving appearance of two transverse red lines
  – Terry’s Nail
    • Crescent nail in liver disease, CHF, DM, normal aging
  – Lindsay’s Nail
    • Half and half nail in renal disease, normal aging
Koilonychia

• Key Points:
  – Spoon Nails
  – Associations:
    • Idiopathic
    • Familial
    • Iron deficiency Anemia
    • HIV
    • Hemochromatosis
    • Plummer-Vinson Syndrome
Hypertrophic Osteoarthropathy

• Key Points
  – Primary HOA
    • Clubbing, Pachyderma, Periostosis
  – Incomplete HOA
    • Isolated (“Familial Clubbing”), Cutis verticus Gyatus
  – Secondary HOA
    • Intrathoracic neoplasm
  – Incomplete Secondary HOA
    • Acquired clubbing – multiple causes
Clubbing

• Unilateral
  – Hemiplegia
  – Vascular lesions:
    • Dialysis fistula
    • Takayasu’s arteritis
    • Ulnar artery aneurysm

• Bilateral
  – Primary
    • Familial
    • Idiopathic
  – Secondary
    • Neoplastic
    • Pulmonary
    • Cardiac
    • Gastrointestinal
    • Infectious
    • Endocrine
    • Vascular
    • Psychiatric

### Clubbing:

<table>
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<tr>
<th>Sign</th>
<th>Degree</th>
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<tr>
<td>Profile sign (Red)</td>
<td>&gt;180</td>
</tr>
<tr>
<td>Modified Profile sign (Yellow)</td>
<td>&lt;180</td>
</tr>
<tr>
<td>Hyponychial angle (Green)</td>
<td>&gt;180</td>
</tr>
<tr>
<td>DIP/IP depth (Black)</td>
<td>&gt;1</td>
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</table>
**Key Points:**
Determine diagnostic accuracy compared to Phalangeal depth ratio.

1. 141 patients in cross sectional study

3. Results:

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<tr>
<th>Schamroth Sign:</th>
<th>Observer 1</th>
<th>Observer 2</th>
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<tbody>
<tr>
<td>Sensitivity</td>
<td>87%</td>
<td>77%</td>
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<tr>
<td>Specificity</td>
<td>90%</td>
<td>90%</td>
</tr>
<tr>
<td>PPV</td>
<td>84%</td>
<td>82%</td>
</tr>
<tr>
<td>NPV</td>
<td>92%</td>
<td>87%</td>
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Concordance

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<th>SS k=0.64</th>
<th>PDR k=0.98</th>
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3. Conclusion:
SS reasonable test to identify clubbing
Onycholysis

• Key Point
  – Separation of nail from nail plate
  – Etiology:
    • Trauma/Irritants/UV
    • Psoriasis
    • Infections
    • Drugs – tetracyclines, fluoroquinolones
    • Tumors
    • Thyroid Disease
      – Hyperthyroidism
      – Other nail:
        » Onychoschizia
        » Onychomalacia (Hapalonychia)
Periungual Telangiectasias

• Key Points:
  – Sign of Connective Tissue Disease
    • Lupus – Hairpin like vessels
    • Dermatomyositis – Globular vessels, skip spaces
    • Sclerodema – Globular vessels, skip spaces
Nail Degloving

• Key Points

  - Distal tip nail unit shedding
  
  - Etiology:
    - Trauma
    - TEN
    - Digital ischemia
    - Infection – gangrene
    - Lichen planus

Nail degloving, a polyetiologic condition with 3 main patterns: A new syndrome.
Baran R¹, Perrin C.
Nail Question 2

• Patient with s/p hospitalization for pneumonia 3 months ago. Which of the following is the best diagnosis?
  A. Bissells’ lines
  B. Mees’ lines
  C. Terry’s nails
  D. Beau’s lines
  E. None of the above
Nails and Systemic Medications

• Retinoids
  – Isotretinoin
  – Acitretin

• EGFR Inhibitors
  – Erlotinib
  – Gefitinib

• Tetracyclines
  – Doxycycline

• Chemotherapy
  – Taxol
Nails and Syndromes

• Yellow Nail syndrome
• Nail-Patella Syndrome
• Brachyonychia
• Darier’s Disease
• Pachyonychia congenita
Yellow Nail Syndrome

• **Key Points:**
  – Arrested nail growth
  – Yellow discoloration (pathognomonic)
  – Absence of cuticle
  – Overcurvature and Onychauxis
    • M=F, older, lower life expectancy
    • Familial
  – Association:
    • Lymphedema
    • Pulmonary disease – bronchitis, bronchiectasis, sinusitis, pleural effusions, malignancy
Nail Patella Syndrome

• Key Points:
  – Triangular lunula
  – Absent patella, radial head dysplasia, iliac horns
  – Nephropathy 40% with CRF in 8%
  – AD, LMX1B gene mutation (collagen synthesis)
Brachyonychia

Key points

- Brachonychia (Racquet nails)
- Familial AD
- Associated with Syndromes
  - Larsen Syndrome
  - Brook—Spiegel Syndrome
  - Rubinstein-Taybi Syndrome
- Acquired:
  - Acroosteolysis
  - Psoriatic arthritis
  - Hyperparathyroidism
Darier’s Disease

• Key Points:
  – Longitudinal erythronychia with V notching
  – AD, Ch 12q23-24.1, ATP2A2
  – Keratosis follicularis
  – Punctate keratoses of palm
  – Exacerbated by UV
Pachyonychia congenita

• Key Points:
  – Pachyonychia
  – Palmar/plantar hyperkeratosis and hyperhidrosis, blistering under callosities
  – Benign leukokeratosis of the mucous membranes
  – AD, Keratin 16 (Type I) and Keratin 17 (Type II)
  – Types:
    • Type I – above
    • Type II – above + natal teeth, steatocystoma
    • Type III – above + leukokeratosis of corneas
    • Type IV – above + hyperpigmentation (Delayed onset)
Nails and Infections

• Bacterial
• Miscellaneous
  – Viral
  – Ectoparasite
• Fungal
  – Dermatophyte
  – Candida
Bacterial Infections of Nails

- Staph/Strep
- Pseudomonas

Complications:
  - Osteomyelitis
    - Adjacent soft tissue infection

Viral Infections of the Nail

• Herpes Simplex
• Human Papilloma virus
Onychomycosis:

• Dermatophytes
  • Superficial White
    – Trichophyton mentagrophytes
  • Proximal Subungual
    – T. rubrum
  • Distal Subungual
    – T. rubrum
    – T. mentagrophyte
  • Treatment: Terbinafine

• Non-Dermatophytes
  • Scytalidium, Scopulariopsis, Fusarium, Acremonium spp.
    – Treatment: Surgical/Chemical avulsion, Itraconazole
  • Candida spp
    – Treatment: Diflucan
Nail Question 3

• Which of the following infections is responsible for this nail finding?
  A. T. rubrum
  B. Fusarium
  C. Staphylococcus
  D. Enterococcus
  E. Pseudomonas
Nails and Trauma

- Hemorrhage
- Onychotillomania
- Onychophagia
Nails and Tumors

- Benign
- Malignant
Digital Mucous Cysts

• Key Points:
  – Often associated with interphalangeal joint osteoarthritis
  – MRI imaging reveals cysts have pedicles that extend into joint
Erythronychia

- Key Points:
  - Longitudinal erythronychia
    - Benign
      - Onychopapilloma (Most common)
      - Verruca
      - Glomus tumor
    - Malignant
      - Squamous cell carcinoma (in situ)
      - Amelanotic melanoma (in situ)
Onychomatricoma

• Key Points
  – Nail matric fibroepithelial tumor
  – Uncommon
  – Fingers>>>toes
  – Presentations:
    • Longitudinal band of yellow thickened nail
    • Splinter hemorrhages
    • Swirling of Keratin (Honeycomb cavities)
Glomus Tumor

• Key Points
  – Arise from glomus body
    • modified Smooth muscle cell
  – 75% occur on the fingertips, 66% subungual area
  – Intense, pulsating pain
  – Characteristic MRI imaging
Fibrous tumors

• Key Points
  – Fibrokeratomas:
    • Proximal nail fold
      – Hereditary (Tuberous Sclerosis)
      – Acquired (Garlic clove fibroma)
    • Dermal connective tissue
      – Post-traumatic/spontaneous
Osteocartilaginous tumors

• Key Points:
  – Subungual exostosis (not a true tumor)
  – Subungual osteochondroma
  – Chondroma
  – Enchondroma
  – Osteoid osteoma
  – Chondrosarcoma
Subungual Squamous Cell Carcinoma

• Key Points:
  – Linked to HPV 16, 34,35
  – Surgical management:
    • Mohs micrographic surgery
    • SLNB not indicated
Subungual Melanoma

- Key Points:
  - 0.1 cases per 100,000
  - Subungual variant represents 1% of all cutaneous melanomas
  - Treatment
    - WLE
    - Mohs
    - Interphalangeal amputation
    - SLNB

Nail Question 4

• Which of the following caused the U shaped defect?
  A. Melanoma
  B. Glomus tumor
  C. Squamous Cell Carcinoma
  D. Fibrokeratoma
  E. Onychotillomania
Just Crazy Nails