Diagnosis and Management of Common Nail Disorders

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Nail Anatomy: Overview

Nail Anatomy: Nail Plate Production

• Made “from the top down”

• Dorsal nail plate:
  - Produced first
  - Made by cells in the proximal nail matrix

• Ventral nail plate:
  - Produced last
  - Made by cells in the distal nail matrix

Nail Anatomy: Proximal Nail Fold

- Defined as proximal border of nail plate
- Extends from skin above proximal most aspect of nail matrix to cuticle

Nail Anatomy: Proximal Nail Matrix

• Extends distally from the blind pocket to the cuticle

• Produces dorsal nail plate
  - Proximal 50% of nail matrix produces >80% of the nail plate

Nail Anatomy: Distal Nail Matrix

- Extends from cuticle to proximal nail bed
- Represents lunula - Visible through nail plate
- Produces ventral aspect of nail plate

Nail Anatomy: Cuticle

• Also termed: eponychium

• Layer of epidermis that adheres to dorsal nail plate

• Extends distally from the distal aspect of the proximal nail fold

• Protects nail matrix from outside pathogens, allergens, irritants

Nail Anatomy: Nail Bed

- Extends from distal nail matrix (lunula) to hyponychium
- Represents majority of structures visible through the nail plate
- **Responsible for adhering nail plate to digit**
- Arranged in longitudinal ridges that parallel ridges on undersurface of nail plate
- Vessels of nail bed oriented similarly
  - Shearing results in splinter hemorrhages
- No role in production of nail plate

Nail Anatomy: Nail Bed

Nail Anatomy: Nail Plate

• Durable keratinized structure

• Produced throughout life
  - Fingers: 3 mm/month
  - Toes: 1 mm/month

• Nail plate growth influenced by multiple internal and external factors

Nail Anatomy: Hyponychium

- Represents distal border of nail apparatus

- Extends from nail bed to distal groove
  - Distal groove: cutaneous ridge that delineates nail apparatus from pulp of digit

- Composed of hyperkeratinized epithelium
  - Acts as an extension of the cuticle
  - Protects nail bed from exogenous irritants and pathogens

Nail Anatomy: Overview

Nail Signs: Overview

- Nail Unit
- Nail Plate Shape
- Nail Plate Surface
- Nail Bed
Common Nail Signs: Clubbing

- Increased transverse and longitudinal curvature of the nail plate
  - Defined as unguo-phalanged angle (Lovibond’s angle) of >180°

- Schamroth’s sign: absence of diamond shape window view between fingers

- Represents enlargement of the underlying soft tissue

- Results from repeated prolonged vasodilation of vessels in nail bed
Common Nail Signs: Pincer Nails

- Over curvature of transverse axis of nail plate
- Results in painful incarceration of underlying nail bed
- Termed trumpet nails when curvature of distal nail plate becomes completely circumferential
- Etiologies:
  - Idiopathic (often with age)
  - Medication (β blockers)
Nail Signs: Koilonychia

- Concavity of the nail plate
- Lateral edges of the nail plate are elevated above a depressed center
- Water drop test

Etiologies:
- Iron deficiency
- Idiopathic
Nail Signs: Onychogryphosis

- Acquired nail dystrophy marked by thickening, lateral curvature, and brown/yellow discoloration of the nail plate
- Resembles a ram’s horn
- Most common in the elderly
Nail Signs: True Leukonychia

• Opacity of the nail plate that obscures underlying nail bed

• Results from structural abnormalities in the nail plate that alter diffraction of light

• Many subtypes defined by degree of involvement of nail plate
  - Total
  - Subtotal
  - Transverse
  - Puntate
  - Variegata
  - Longitudinal
Nail Signs: Apparent Leukonychia

- White appearance of nail plate resulting from changes (edema) of the nail bed

- Terry’s nails
  - Three quarters nails
  - Associated with liver disease

- Muehrcke’s lines
  - Transverse lines
  - Associated with hypoalbuminemia

- Lindsay’s nails
  - Half and half nails
  - Associated with impaired renal function
Nail Signs: Erythronychia

- Red discoloration of the nail

- Corresponds with partial loss of function of nail matrix resulting in nail plate thinning
  - Underlying vessels more apparent

- Longitudinal erythronychia
  - Solitary: neoplastic process
  - Multiple: inflammatory disorder

- Red lunulae represent vasodilation of the nail matrix
Nail Signs: Melanonychia

- Represents melanin in the nail plate

- Often presents as longitudinal band originating in the nail matrix

- Hutchinson’s sign is defined as extent of pigment onto the cuticle, proximal nail fold, or hyponychium
Nail Signs: Nail Pitting

• Shallow depressions in the dorsal nail plate

• Represents inflammation in the proximal nail matrix resulting in defective nail plate production

• Pattern and depth may suggest underlying pathology

• Alopecia areata:
  - Regular, shallow pits
  - “Checkerboard” pattern

• Psoriasis:
  - Deep, irregular pits
  - Scatter shot pattern
Nail Signs: Beau’s Lines

- Transverse depressions, grooves, or furrows involving part or all of the nail plate
  - Parallel profile of the lunula

- Represent temporary reduction in nail matrix activity

- Result from systemic or exogenous stress or trauma
  - Width corresponds with duration and severity of insult
  - Systemic causes typically result in involvement of multiple nails
Nail Signs: Onychomadesis

- Spontaneous detachment of the nail plate from the nail matrix
- Results in nail plate shedding at the proximal nail fold
- Represents severe insult to the nail matrix arresting all activity
Nail Signs: Onychoschizia

- Lamellar splitting and peeling of the dorsal surface of nail plate

- Associated with nail fragility and prolonged/frequent exposure to water or solvents

- More common in postmenopausal women

- Benign
Nail Signs: Onycholysis

• Separation of the nail plate from the underlying nail bed

• Most often results from disruption or compromise of hyponychium

• Many associated dermatologic and systemic conditions
  - Idiopathic – candidal colonization
  - Psoriasis
  - Onychomycosis
  - Medications
  - Thyroid disease
  - Trauma
Nail Signs: Onychorrhexis

- Confluent, longitudinal ridging and grooving of the nail plate

- Often associated with in thinning of the nail plate
  - Distal “v-shaped” nicking often present

- Occurs as a physiologic consequence of aging
  - Regular
  - No other nail changes present

- Prominent feature of lichen planus of the nails
  - Irregular
  - Associated with atrophy of the nail plate, onycholysis, onychoschizia
Common Nail Disorders
Case Presentations
Case 1
Case 1

• 54 year old man with a no significant past dermatologic history presented with new nail discoloration

• Slowly enlarging over the past year

• No history of trauma

• No personal, family history of melanoma

• No personal history of non-melanoma skin cancer
Case 1 – Clinical Examination

- Irregular 13 mm longitudinal pigmented band involving ~80% of the nail plate
- Wider at proximal aspect: “pyramid sign”
- +Hutchinson’s sign
Case 1 – Work up: Proximal nail matrix shave biopsy

- More precise
- Reduces risk of permanent nail dystrophy
  - Shallower
  - Scarring of the nail matrix less likely
- Less morbid post-operative course
  - Nail plate acts as biological dressing
Case 1 – Work up: Proximal nail matrix shave biopsy

- Equipment:
  - T-ring tourniquet
  - Nail elevator
  - English Anvil (nail splitter)
Case 1 – Work up: Proximal nail matrix shave biopsy

- Anesthesia with lidocaine without epinephrine for “wing block”
- Hemostasis with T-ring tourniquet
- Cuticle, proximal nail fold released with nail elevator
- Incisions made proximally at lateral nail folds
- Proximal nail fold reflected with skin hook or suture
- Nail plate split transversely with English anvil, avulsed to expose nail matrix
- Pigmented lesion scored with 15 blade scalpel, shave removal
- Nail plate, proximal nail fold reapproximated

Proximal nail fold sutured
Case 1 – Work up: Proximal nail matrix punch biopsy

- Indicated for smaller (≥3 mm) lesions

- Greater depth
  - Increases risk of permanent nail dystrophy

- In some cases reflection of the proximal nail fold not required

- Avulsion of the nail plate not necessary

Photo courtesy of Nathaniel Jellinek, MD
Subungual Melanoma
Subungual Melanoma - Epidemiology

• Relatively rare
  - Accounts for 0.7-3.5% of all melanomas

• Thumbs, index fingers, halluces most commonly affected
  - Often preceded by reported trauma
  - Possible etiologic component?

• Amelanotic in ~25% of cases

• Poorer prognosis than cutaneous melanoma
  - 5 year survival of ~15%

Subungual Melanoma – Clinical Presentation

• **Longitudinal melanonychia**
  - Most common presentation
  - Dark band with blurred margins
  - Proximal aspect often wider than distal aspect

• **Hutchinson’s sign**
  - Clinical manifestation of radial growth phase
  - Defined as extent of pigment onto:
    - Cuticle
    - Proximal nail fold
    - Hyponychium

• **Nail plate abnormalities**
  - Indicative of matrical invasion of tumor
  - Thinning, fissuring, onycholysis, longitudinal furrows


Amelanotic Subungual Melanoma – Clinical Presentation

- Friable subungual or periungual nodule

- Often mimics pyogenic granuloma or onychocryptosis

- Onycholysis often first presenting sign

- Nail bed or nail fold ulceration appears later

Subungual Melanoma - Treatment

• Amputation
  - Traditional therapy
  - No definitive survival advantage
  - Significant associated functional, psychosocial morbidity

• Wide local excision
  - Recommended for cases of *in situ* or minimally invasive disease (>0.5 mm) (Sinno, et al. 2015)
  - Significant reduction in functional morbidity
Subungual Melanoma – Treatment

Photo courtesy of S. Tyler Hollmig, MD
Subungual Melanoma – Treatment

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Subungual Melanoma – Treatment

Photo courtesy of S. Tyler Hollmig, MD
Case 2
Case 2

- 75 year old Mexican American woman

- 6 month history of increased transverse curvature of the nails (fingers > toes), now complicated by onychocryptosis on multiple digits

- Significant associated pain, tenderness, swelling

- Treating with aggressive nail debridement, pedicures
Case 2 – Clinical Examination

- 10/10 fingernails with increased transverse curvature of the nail plate
- Lateral nail folds with erythema, edema
- Early incarceration of the lateral aspects nail plate
Case 2 – Work up

- Clipping for pathology, fungal culture
  - Negative x2

- Review of past medical history, current medications
  - Recently started on atenolol for hypertension
Medication-Induced Pincer Nail Deformity
Pincer Nail Deformity – Clinical Presentation

• Pincer nails represent an uncommon adverse effect of multiple medications
  - β-blockers (most common)
  - Ca channel blockers
  - Polychlorinated biphenyl exposure

• Often involve multiple nails, acute onset

• Resolve with discontinuation of offending agent
Pincer Nail Deformity – Differential Diagnosis

- Subungual exostosis
  - XR
- Age related
- Hemodialysis
- Onychomycosis
- Connective tissue disease (Lupus)
- Psoriasis
- Trauma
Case 3
Case 3 – History

- 65 year old Caucasian woman with a recent history of nail “discoloration” affecting 20/20 nails

- Appeared slowly over time

- Multiple medical problems
  - Type II DM
  - COPD
  - Hypertension
  - Hyperlipidemia
  - History of polysubstance abuse

- No treatments to date
Case 3 – Clinical Examination

- Leukonychia of the proximal 2/3 of the nail plate
  - Inversion of lunula/nail bed ratio

- Lunula obscured

- Color discrepancy less evident with pressure

- No abnormalities of the nail plate
Terry’s Nails
Case 3 – Work Up

• Labs:
  - CBC
  - LFTs
  - Creatinine
  - Albumin
  - +/- hepatitis panel
  - Consider hepatic US

• Examination
  - Pedal edema
  - Stigmata of end stage liver disease
Terry’s Nails

- **Apparent leukonychia**
  - Proximal 2/3 appear white, lunula obscured
  - Distal 1/3 appear pink (normal)
- Represents nail bed edema
- No true discoloration of the nail plate
- Associated with liver disease
Case 4
Case 4 – History

- 66 year old Caucasian man with progressive thinning, brittleness of nails
- Recent onset
- No relevant past medical, dermatologic history
- No new medications
Case 4 – Clinical Examination

- Onychorrhexis
- Onychoschizia
- Onychoatrophy
- Hapalonychia
Case 4 – Work Up

- Detailed Social History
  - Occupation
  - Exposures
  - Manicures

- Total body skin examination for any other cutaneous involvement
  - Including oral/genital examination

- Biopsy of lunula (distal nail matrix) for pathology
Amyloidosis of the Nails
Amyloidosis of the Nail – Background

- Relatively common among patients with systemic disease
- Rare etiology of thin/weak/brittle nails
- May represent earliest manifestation of systemic amyloidosis
- Nails appear uniformly thin, ridged (onychorrhexis), brittle, distally split
Amyloidosis of the Nails – Differential Diagnosis

- Brittle nail/Hapalonychia differential diagnosis:
  - Idiopathic
  - Irritant
  - Malnutrition/Vitamin deficiency
  - Chemotherapy/Radiation
  - Hypothyroidism
  - Peripheral vascular disease
  - Graft versus host disease
  - Raynaud’s
  - Lichen planus of the nails
Brittle Nails/Hapalonychia – Treatment

- Irritant avoidance
- Biotin 5 mg/day
- Consider oral contraceptives vs hormone replacement therapy in women if symptomatic and no contraindications
- Avoid manicures, nail cosmetics
- Clear matte cosmetic lacquer
  - Avoid nail hardeners
- NuVail
- Genadur
Case 4
Case 4

- 55 year old Caucasian woman with 3 year history of lifting of the nail plate affecting multiple fingernails
- Treated with triamcinolone 0.1% ointment in the past with no improvement
- No relevant past medical, dermatologic history
- No medications
Case 4 – Clinical Examination

- Distal onycholysis of variable degrees affecting most fingernails
- No erythema of the nail bed, proximal nail folds
- No pitting, “oil spots,” subungual debris
- No other stigmata of hand dermatitis or xerosis
Case 4 – Work up

- Detailed social history
  - Occupation
  - Housework
  - Manicures
  - Nail cosmetics

- Past medical history
  - Thyroid disease
  - Vascular disease

- Medications
  - Tetracyclines
  - Other photosensitizing agents
Simple Onycholysis
Onycholysis – Pathophysiology

• Most often secondary to candidal colonization of subungual space
  - Act as space occupying lesion
  - Prevents reattachment of nail plate to nail bed

• Multiple contributing factors:
  - Manicures/Nail cosmetics
  - Frequent wet/dry cycles
  - Manipulation of subungual space

• Also associated in some cases with hand dermatitis, xerosis
  - Hyponychium desiccated, unable to adhere to nail plate
Onycholysis – Differential Diagnosis

- Photo-onycholysis
- Psoriasis
- Onychomycosis
- Systemic etiology
  - Thyroid disease
  - Vascular insufficiency
Onycholysis – Therapy

• Irritant avoidance
  - Cotton lined vinyl gloves

• Avoid manicures, manipulation of the subungual space
  - Clean with white bar soap

• Trim nail plates short

• Topical antifungals
  - Ciclopirox 0.77% cream

• Oral antifungals
  - Fluconazole 150 mg/week
A Word About Manicures…

• Source of many skin, nail infections
  - Onychomycosis
  - Paronychia
  - Hepatitis C

• Potential UV exposure (*Shellac* and other gel manicures)

• Limited regulatory oversight

• Human rights/trafficking concerns
A Word About Manicures… Recommendations

- Purchase tools/manicure set
- Avoid footbaths with blowers
- Avoid debridement of calluses
- Avoid pushing, trimming, or nipping of cuticles, lateral nail folds
- Avoid manipulation of the subungual space
  - Clean with white bar soap
- Single-use tools should be only used once
  - Emery boards
  - Toe separators
  - Buffing blocks
  - Cuticle brush

Photo courtesy of Dana Stern, MD
Case 5
Case 5 – History

- 75 year old man with a longstanding history of thickened, yellow nails, onycolysis and subungual hyperkeratosis
- Past dermatologic history significant for tinea pedis, tinea cruris
- Past medical history significant for type II diabetes, diet controlled
- Treated with multiple topical agents in the past with no improvement
- No reported medications
Case 5 – Clinical Examination

- Distal onycholysis
- Xanthonychia
  - Spikes represent dermatophytoma
- Onychauxis
- Subungual debris
Case 5 – Work Up

• Clippings
  - Pathology (PAS stain) – distal
  - Fungal Culture – proximal

• Screening for other sites of dermatophytosis

• Discontinue all topical antifungals if in use
Onychomycosis
Onychomycosis – Management

• Hold treatment until proven by PAS and causitive organism identified
  - Dermatophytes
  - Non-Dermatophyte mold

• Review relevant medical history
  - Liver disease
  - Congestive heart failure
  - Family planning
  - Other medications

• Screening labs if considering oral therapy
  - CBC
  - CMP
  - Comprehensive hepatitis panel

• Treat other sites of dermatophytosis
  - Butenafine 1% cream
  - Ciclopirox 0.77% cream
Onychomycosis – FDA Approved Treatments

**Topical Agents**
- **Ciclopirox 8% lacquer**
  - Daily x 48 weeks
- **Efinaconazole 10% solution**
  - Daily x 48 weeks
- **Tavaborole 5% solution**
  - Daily x 48 weeks

**Oral Agents**
- **Terbinafine 250 mg daily**
  - Fingernails: x 6 weeks
  - Toenails: daily x 12 weeks
- **Griseofulvin 250 mg three times daily**
  - Fingernails: >4 months
  - Toenails: >6 months
- **Itraconazole 200 mg**
  - Fingernails: twice daily for one week/month x 2 months
  - Toenails: daily x 12 weeks

**Not FDA Approved Indication**
- **Fluconazole 150 – 300 mg weekly**
  - Fingernails 3-6 months
  - Toenails 9-12 months
Onychomycosis – New Therapies: Efinaconazole

- Triazole antifungal

- Improved vehicle allows for better penetration of the nail plate
  - Wait 10 minutes after showering
  - Apply on, around, under nail

- Limited side effects
  - Onychocryptosis
  - Allergic/Irritant contact dermatitis
  - Flammability

- Cost
  - 4 ml: $558.58
  - 8 ml: $1,108.81
Onychomycosis – New Therapies: Tavaborole

- Developed at Anacor Pharmaceuticals

- Novel mechanism of action
  - First in oxaborole class
  - Inhibits leucyl-tRNA synthase

- Limited side effects
  - Allergic contact dermatitis
  - Irritant contact dermatitis
  - Onychocryptosis

- Cost:
  - 4 ml: $547.03
  - 10 ml: $1383.93
Onychomycosis – Therapy

Dermatophyte
- Oral: Terbinafine
- Topical: Efinaconazole
- Other: aggressive debridement
  - Podiatry
  - Urea 40% cream

Candida
- Oral: Fluconazole > Itraconazole
- Topical: Efinaconazole
- Other: Irritant avoidance
  - Gloves
  - Stop manicures

Non Dermatophyte Molds
- **Aspergillus**
  - Oral: Terbinafine
  - Topical: Tavaborole

- **Scopulariopsis/Fusarium**
  - Oral: Itraconazole > Fluconazole
  - Topical: Tavaborole
  - Other: Expectation management
Case 6
Case 6 – History

- 50 year old British woman of Indian descent presenting with 6 month history of nail changes affecting 18/20 nails

- History of manicures – attributes onset to aggressive cuticle episode of cuticle pushing/trimming

- Treated in the past with 3 month course of oral terbinafine with no improvement

- Works in packing/shipping – frequent trauma to hands, nails

- No relevant past medical or dermatologic history
Case 2 – Physical Examination

- Diffuse regular hyperpigmentation nail plate with distal atrophy
- Erythema, edema of the proximal, lateral nail folds
- Distal onycholysis
- Nail bed hyperkeratosis, scale
- Complete absence of all cuticles
Case 2 – Physical Examination

- Onychorrhexis
- Diffuse hyperpigmentation of the nail plate with distal atrophy
- Nail bed hyperkeratosis, scale
Case 2 – Work up

• Punch biopsy of the distal nail matrix
• Total body skin examination
• Detailed medication history
• Labs
• Differential diagnosis
  - Psoriasis
  - Lichen planus
  - Pityriasis rubra pilaris
  - Onychomycosis
  - Acrokeratosis paraneoplastica
Nail Lichen Planus
Nail Lichen Planus - Epidemiology

- Nail involvement present in ~10% of all cases of lichen planus
- May occur in the absence of cutaneous or mucosal involvement
- More common in adults than children
- Rarely affects a solitary digit – multi-nail involvement far more common
Nail Lichen Planus – Clinical Presentation

- Onychorrhexis
- Onychoschizia
- Nail plate thinning, atrophy
  - Cicatricial process
  - Nail matrix may be destroyed by inflammation
  - May result in anonychia
- Dorsal pterygium
  - Adhesion of the proximal nail fold to the nail bed
  - Occurs when nail plate absent due to destruction of matrix
Nail Lichen Planus - Treatment

• Treatment primarily directed at preventing permanent scarring
  - Topical corticosteroids
  - Intralesional corticosteroids
  - Systemic corticosteroids (IM triamcinolone 0.5 mg/kg/month)
  - Irritant avoidance
  - Prevention of secondary infections
Questions?