

WHAT'S YOUR DIAGNOSIS?

Fingernail Abnormalities After a Systemic Illness

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Examining a patient's nails and understanding nail growth mechanics can provide helpful clues to help treat past injuries or underlying diseases.

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A 45-year-old African American woman presented with painless fingernail detachment and cracks on her fingernails that had developed over the previous month. Her medical history was notable for an episode of Stevens-Johnson syndrome 2 months prior that required treatment with prednisone, IV immunoglobulin, etanercept, acetaminophen, and diphenhydramine.

A physical examination revealed multi-

ple fingernails on both hands that exhibited 4 mm of proximal painless nail detachment with cream-colored discoloration, friability, and horizontal splitting (Figure). New, healthy nail was visible beneath the affected areas. Toenails were not affected.

- What is your diagnosis?
- How would you treat this patient?

FIGURE Painless Detachment of the Proximal Nail Plate With White Discoloration, Sparing the Distal Nail



DIAGNOSIS

Based on the timing and characteristics of her nail detachment, the patient was diagnosed with onychomadesis, which is defined as painless detachment of the proximal nail plate from the nail matrix and nail bed after at least 40 days from an initial insult. Air beneath the detached nail plate causes a characteristic creamy-white discoloration. The severity of onychomadesis

ranges from transverse furrows that affect a single nail without shedding, known as Beau lines, to multiple nails that are completely shed.^{1,2} Nail plate shedding is typical because the nail matrix, the site of stem cells and the most proximal portion of the nail apparatus, is damaged and transiently arrested.

Various etiologies can halt nail plate production abruptly within the matrix. These

typically manifest ≥ 40 days after the initial insult (the length of time for a fingernail to emerge from the proximal nail fold).² The annual incidence of these etiologies ranges from approximately 1 per 1 million people for Stevens-Johnson syndrome, a rare cause of onychomadesis, to 1 per 10 people for onychomycosis, one of the more common causes of onychomadesis.³ The Table compares the characteristics of the diagnoses that are most commonly associated with nail detachment and discoloration.

When a single nail is affected, the etiology of onychomadesis usually is primary and local, including mechanical nail trauma and fungal nail infections (onychomycosis).^{1,2} *Candida* onychia is onychomycosis caused by *Candida* species typically *Candida albicans*, which result in localized nail darkening, chronic inflammation of the paronychia skin, and cuticle loss. The infection favors immunocompromised people; coinfections are common, and onychomadesis or onycholysis can occur. Unlike onychomadesis, onycholysis is defined by painless detachment of the distal nail plate from the nail bed, but nail shedding typically does not occur because the nail matrix is spared. The preferred treatment for *Candida* onychia is oral itraconazole, and guided screenings for immunodeficiencies and endocrinopathies, especially diabetes mellitus, should be completed.^{3,4}

Tinea unguium is another form of onychomycosis, but it is caused by dermatophytes, typically *Trichophyton rubrum* or *Trichophyton mentagrophytes*, which produce white and yellow nail discoloration followed by distal to proximal nail thickening and softening. Infection usually begins in toenails and demonstrates variable involvement in each nail as well as asymmetric distribution among digits.³ This condition also may eventuate in onychomadesis or onycholysis. Debridement followed by oral terbinafine is the treatment of choice.⁴

Two other causes of localized nail discoloration with or without nail detachment include melanonychia and nail bed infection by *Pseudomonas aeruginosa* (*P aeruginosa*). Melanonychia can be linear or diffuse brown discoloration of 1 or more nails caused by melanin deposition. Either pattern is a common finding in dark-skinned

TABLE Common Diagnoses Associated With Nail Plate Detachment and Discoloration

Diagnosis	Characteristics
Onychomadesis	Painless proximal detachment of ≥ 1 nail(s) from the nail matrix and bed due to local trauma or systemic causes, respectively, resulting in nail shedding
Onycholysis	Painless distal detachment of ≥ 1 nail(s) from the nail bed often due to mechanical trauma, psoriasis, or infections and not associated with nail shedding
<i>Candida</i> onychia	Asymmetric distribution of dark nail discoloration with proximal nail fold swelling and erythema plus cuticle loss from <i>Candida</i> onychomycosis
Tinea unguium	Asymmetric distribution of soft white hyperkeratosis and nail yellowing from dermatophytic onychomycosis
Melanonychia	Dark brown nails often with longitudinal bands and commonly seen as a normal variant in dark-skinned individuals
<i>Pseudomonas</i> infection	Opportunistic nail infection that causes gray-green staining associated with chronic paronychia from frequent water submersion

people, especially by age 50 years, but melanocyte hyperplasia should be excluded in all individuals along with drug adverse effects, exogenous pigments, infections, and systemic diseases.^{3,5} *P aeruginosa* produces pyocyanin, the green pigment responsible for the discoloration seen in this opportunistic infection often localized to a single nail. Prior maceration of the nail apparatus by repeated water submersion is common among affected individuals. Avoidance of submerging fingernails in liquids followed by nail debridement and oral antipseudomonal antibiotics is the preferred treatment course.³

The etiology is usually secondary and systemic when multiple nails demonstrate onychomadesis, but the exact pathophysiology is poorly understood. One of the most studied infectious etiologies of onychomadesis is hand-foot-and-mouth disease (HFMD), which typically affects children aged < 10 years. Parents often will recall their child being ill 1 to 2 months prior to the nail findings. Scarlet fever and varicella also can result in onychomadesis. Although not common systemic causes, Stevens-Johnson syndrome and toxic epidermal necrolysis can trigger onychomadesis of multiple nails that usually resolves in several months, but other nail

deformities often persist.^{2,6} Onycholysis also can accompany this finding.⁷ Autoimmune etiologies of onychomadesis include alopecia areata and pemphigus vulgaris. Inciting medications that are toxic to the nail matrix include chemotherapy agents, valproic acid, carbamazepine, lithium, and azithromycin. Rare congenital disorders and birth trauma also can present with onychomadesis of multiple nails during infancy.²

Systemic etiologies typically affect fingernails more than toenails because of the faster growth rate of fingernails. Once the source of onychomadesis is controlled or eradicated, complete regrowth of fingernails can take from 4 to 6 months. Toenails can take twice as long and older age increases all regrowth periods.⁵

Our patient was treated with analgesics until her mucosal surfaces fully healed, and topical emollients and keratolytics were used to soften eschars from previous blisters and prevent further scar formation. Her affected fingernails shed and regrew after 6 months without additional interventions.

CONCLUSION

Although Stevens-Johnson syndrome is a rare cause of onychomadesis, and the pathophysiology of this sequela is poorly understood, this case illustrates a common nail abnormality with multiple potential etiologies that are discerned by an accurate history and thorough exam. In the absence of decorative nail polish, nails can be easily examined to provide helpful clues for past injuries or underlying diseases. An understanding of nail growth mechanics and associated terminology reveals the diagnostic and therapeutic implications of proximal vs distal nail detachment, the hue of nail discoloration, as well as single vs multiple affected nails.

Onychomadesis in single nails should prompt questions about nail trauma or risk factors for fungal infections. Depending on the etiology, manual activities need to be adjusted, or antifungals need to be initiated while investigating for an immunocompro-

mised state. Onychomadesis in multiple nails in children should raise suspicion for HFMD or even birth trauma and congenital disorders. Multiple affected nails in adults should prompt guided questions for autoimmune diseases and inciting medications. For onycholysis, trauma, psoriasis, or certain infections should be the target. Green nails are easily recognized and treated with a defined regiment, whereas dark nails should be examined closely to differentiate *Candida* onychia from melanonychia. Whether from a rare cause in an adult to a common illness in a child, primary care providers have sufficient expertise to diagnose and treat various nail disorders and reassure worried patients and parents with an understanding of nail regrowth.

Author disclosures

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