Isolated case of acquired onychodystrophy

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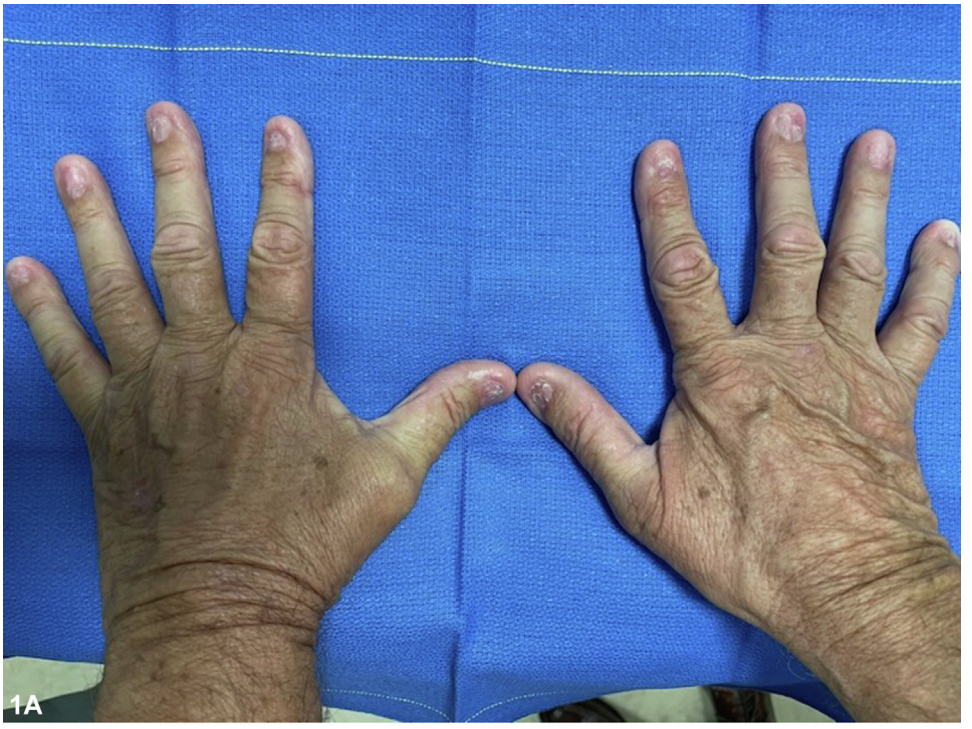
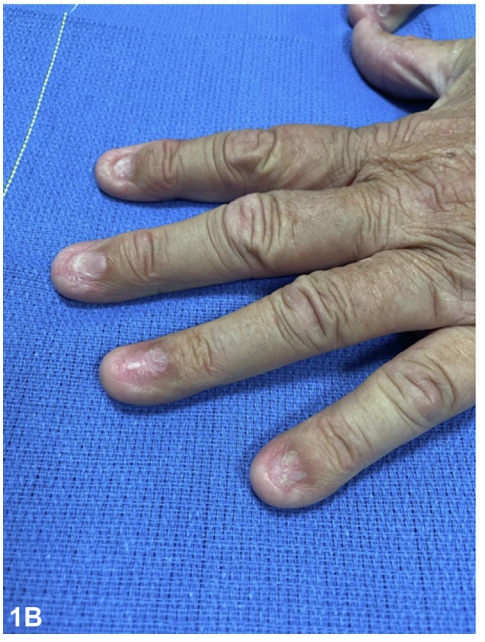
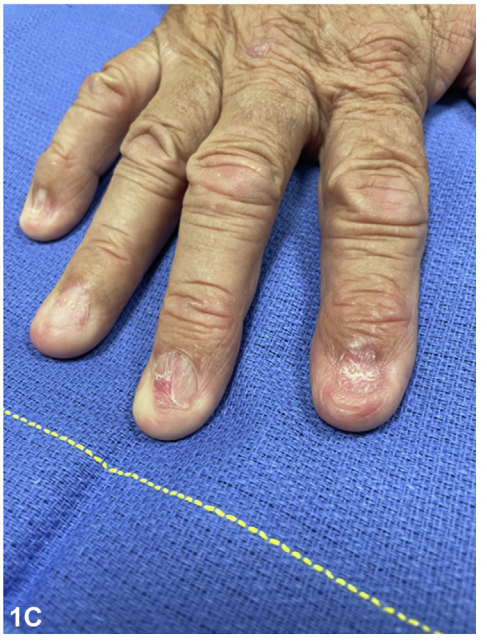
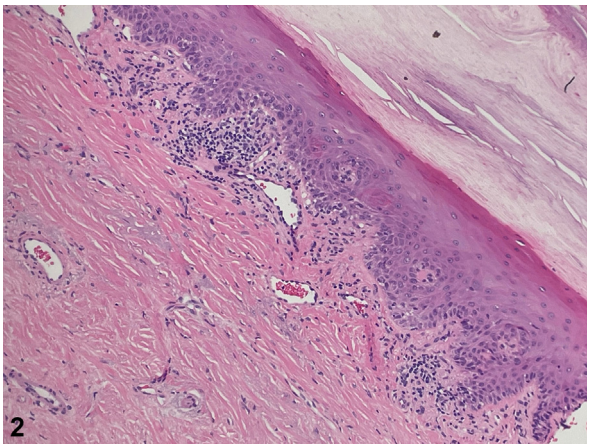
Key words: onychodystrophy; lichen planus; nail pterygium.









CASE

A 77-year-old otherwise healthy male presented with progressive nail dystrophy involving his fingernails that onset 10 years prior to presentation. He denied oral and other cutaneous lesions. He was otherwise feeling well without systemic symptoms. Review of systems was noncontributory and family history was unremarkable. Physical exam demonstrated dorsal pterygium with evidence of anonychia affecting multiple fingernails (Fig 1, *A* and *B*). A 3-mm punch biopsy of the right third digit nail matrix demonstrated a superficial perivascular and interface lichenoid dermatitis, predominantly lymphocytes, with jagged epidermal hyperplasia, and absence of eosinophils (Fig 2). Periodic-acid Schiff was negative for fungal elements.

Question 1: What is the most likely diagnosis?

A. Systemic amyloidosis

B. Lichen striatus

C. Dyskeratosis congenita

D. Isolated nail lichen planus (NLP)

E. Graft-versus-host disease

Answers:

A. Systemic amyloidosis — Incorrect. Nail findings seen with systemic amyloidosis include thinning and fissuring, often associated with splinter hemorrhages.[1] Histologic examination would also reveal amyloid deposits within the nail matrix and nail bed dermis.[1]

B. Lichen striatus — Incorrect. Lichen striatus often presents with lichenoid nail changes that are limited to 1 or 2 digits, not multiple.[1]

C. Dyskeratosis congenita — Incorrect. Dyskeratosis congenita is an inherited bone marrow failure syndrome with characteristic symptoms including lichenoid nail changes, premalignant oral leukoplakia, and reticulated hyperpigmentation. This diagnosis is less likely in a patient presenting without the mucocutaneous triad or a family history of dystrophic nails.[1] Furthermore, this genodermatosis usually presents in the first decade of life.

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Funding sources: This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the authors(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

IRB approval status: Not applicable.

Patient consent: Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

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JAAD Case Reports 2023;32:59-62.

2352-5126

https://doi.org/10.1016/j.jdcr.2022.11.031

D. Isolated NLP — Correct. NLP is most frequently seen without involvement of the skin, scalp, or mucosa and clinical manifestations of NLP often include thinning of the nail plate, longitudinal ridging, fissuring, and nail atrophy.[1] Other clinical findings include melanonychia and onychorrhexis.[2] The clinicopathologic correlation supported a diagnosis of NLP.

E. Graft-versus-host disease — Incorrect. Although graft-versus-host disease may present with similar nail changes, additional cutaneous manifestations are often present.[1] Additionally, this patient does not have a history of allogeneic hematopoietic stem cell transplantation to support this diagnosis.[1]

Question 2: Which of the following nail findings are indicative of severe disease presentation?

A. Mottled erythema of the lunula

B. Pterygium

C. Onycholysis

D. Distal nail splitting of <3 mm in length

E. Subungual hyperkeratosis

Answers:

A. Mottled erythema of the lunula — Incorrect. This finding is consistent with moderate disease presentation of NLP. Additional moderate NLP features include partial fissuring, longitudinal grooves, distal splitting of 3 to 5 mm in length, onycholysis between 25% and 50%, and subungual hyperkeratosis.[2-4]

B. Pterygium — Correct. Development of pterygium is consistent with severe disease presentation of NLP. Additional findings include complete fissuring, deep grooves, splitting of >5 mm in length, onycholysis >50%, diffuse erythema of the lunula, and anonychia.[2-4]

C. Onycholysis — Incorrect. This finding is consistent with mild disease presentation of NLP. Additional mild NLP features include nail thinning, longitudinal ridging, distal splitting of <3 mm in length, onycholysis <25%, and no nail bed hyperkeratosis.[2-4]

D. Distal nail splitting of <3 mm in length — Incorrect. This finding is consistent with moderate disease presentation of NLP.[2-4]

E. Subungual hyperkeratosis — Incorrect. This finding is consistent with moderate disease presentation of NLP.[2-4]

Question 3: What is the most appropriate first-line treatment for this condition?

A. Intralesional triamcinolone acetonide

B. Acitretin

C. Oral terbinafine

D. Methotrexate

E. Azathioprine

Answers:

A. Intralesional triamcinolone acetonide — Correct. The first-line treatment for NLP is intralesional triamcinolone acetonide into the nail matrix or nail bed; intramuscular triamcinolone may be utilized as an adjunctive therapy for severe presentations.[2] Prompt recognition of NLP and intervention with aggressive treatment is warranted to prevent permanent nail loss.[5]

B. Acitretin — Incorrect. Acitretin (0.2 to 0.3 mg/kg/d) and alitretinoin (30 mg/d) have demonstrated effectiveness for mucocutaneous lichen planus, but their role in NLP is less defined. Oral retinoids may be prescribed for the treatment of NLP but are considered as second-line therapy, if a patient has failed intralesional corticosteroid injections or if a contraindication is present.[3]

C. Oral terbinafine — Incorrect. Terbinafine, a squalene epoxidase inhibitor, is Food and Drug Administration-approved for the treatment of nail onychomycosis. While onychomycosis may be considered in the initial clinical differential of dystrophic nails, this oral therapy would be ineffective in the treatment of NLP.[1]

D. Methotrexate — Incorrect. While methotrexate is administered off-label for the treatment of cutaneous lichen planus, its role in the management of NLP is not well supported as compared with other therapies.[3,5]

E. Azathioprine — Incorrect. Azathioprine may be considered as monotherapy for the treatment of NLP, but it is a deemed third-line therapy, as are cyclosporine and mycophenolate mofetil, as its clinical efficacy is not well elucidated in the literature.[3]

Abbreviation used:

NLP: nail lichen planus

Conflicts of interest

None disclosed.

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