Koilonychia secondary to Raynaud's phenomenon: A rare co-occurrence

Sepideh Ashrafzadeh, BS,[a] and Sotonye Imadojemu, MD, MBE[a,b]
Boston, Massachusetts

Key words: koilonychia; nail dystrophy; Raynaud's phenomenon; spoon nails; vasospasms.

INTRODUCTION

Koilonychia, also called spoon nails, is a nail disease characterized by concave nail plates.[1] It can be either hereditary or acquired, with the latter typically occurring in individuals with iron deficiency, trauma, occupational exposures, or diabetes mellitus.[1,2] Rarely, it has also been associated with thyroid disorders or autoimmune diseases such as alopecia areata, psoriasis, or lichen planus.[2] This report presents the unusual case of koilonychia occurring secondary to primary Raynaud's phenomenon and the successful reversal of koilonychia with topical nitroglycerin.

CASE REPORT

A 52-year-old woman with a thirty-year history of Raynaud's phenomenon presented with scooped nails on her bilateral hands. Over the past year, her nail plates had become increasingly brittle, ridged, and flat or scooped-shaped. She did not have ulcerations of her fingertips, but noted that she was developing frequent and severe Raynaud's phenomena with white and red color changes and pain in response to cold temperatures or stress on both hands and occasionally on her toes. The patient otherwise denied any hair loss, nail pitting, rashes, oral ulcers, joint pain, dysphagia, skin thickening, fever, fatigue, shortness of breath, chest pain, dry eyes or mouth, or cough. She had treated her Raynaud's phenomenon with behavioral modification. The patient also tried topical minoxidil on her dorsal hands and cannabidiol cream but saw little to no improvement in her nails and the frequency of her Raynaud's phenomenon with these interventions. She had no other medical conditions and only took biotin and multivitamin supplements. She did not work with petroleum-based products, and she had a predominantly vegetarian diet with occasional chicken and fish.

Her physical examination was notable for proximal leukonychia, erythronychia, longitudinal ridging, ragged cuticles, and concave-shaped nail plates on multiple nails on her left and right hands (Fig 1, *A* and Fig 2, *A*). Nailfold capillary microscopy revealed regular vessel architecture, density, and capillary size without signs of hemorrhage, avascular regions, or loss of capillary loops. She did not have conjunctival pallor.

Laboratory tests were negative for antinuclear antibodies, centromere antibody, anti-Smith antibody, anti-RNP antibody, anti-LA antibody, anti-RO antibody, and anti-SCL70 antibody. The patient's hemoglobin, ferritin, iron, total iron binding capacity, unsaturated iron binding capacity, fasting blood glucose, and thyroid-stimulating hormone levels were all within the normal limits. Based on her history, examination, and laboratory findings, a clinical diagnosis of koilonychia and nail dystrophy secondary to primary Raynaud's phenomenon was made.

The patient was started on 2% nitroglycerin ointment on the bilateral nail folds and web spaces daily. She was also advised to continue to keep her core warm to increase warming of her extremities. The patient reliably followed both of these recommendations, applying the nitroglycerin ointment 2% daily and keeping her core and hands warm. After five months of treatment, she had near resolution of her nail dystrophy and koilonychia (Fig 1, *B* and Fig 2, *B*).

From the Harvard Medical School,[a] and Department of Dermatology, Brigham and Women's Hospital, Boston.[b]
Funding sources: None.
Conflicts of interest: None disclosed.
IRB approval status: Not applicable.
Correspondence to: Sotonye Imadojemu, MD, MBE, Department of Dermatology, Brigham and Women's Hospital, 221 Longwood Ave, Boston, MA 02115. E-mail: simadojemu@bwh.harvard.edu.

JAAD Case Reports 2021;7:117-9.
2352-5126

https://doi.org/10.1016/j.jdcr.2020.11.021

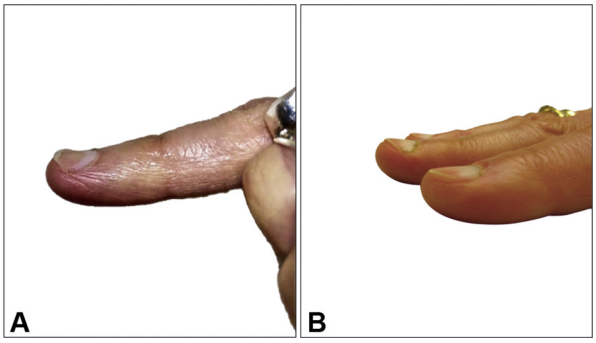

Fig 1. A, Lateral view of the patient's left index finger highlighting koilonychia secondary to primary Raynaud's phenomenon after six weeks of treatment with topical nitroglycerin. **B**, Resolution of koilonychia on the nail plates of the left hand after five months of treatment with topical nitroglycerin.

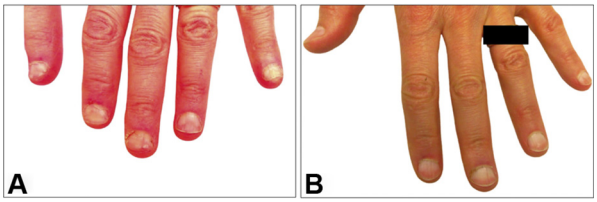

Fig 2. A, Proximal leukonychia, erythronychia, longitudinal ridging, ragged cuticles, and concave-shaped nail plates on multiple nails on the patient's left hand prior to initiation of nitroglycerin treatment. **B**, Interval improvement in nail dystrophy and cuticles after 5 months of treatment with topical nitroglycerin.

DISCUSSION

This case highlights the rare occurrence of koilonychia in a patient with primary Raynaud's phenomenon who was able to successfully reverse her koilonychia by treating her Raynaud's phenomenon with topical nitroglycerin. Raynaud's phenomenon has been associated with several nail findings, including parrot beak nails, brittle nails, and longitudinal ridging, but reports about koilonychia are sparse in the literature.[3,4] While the pathogenesis of koilonychia is unclear, the nail disease is attributed to diminished digital blood flow disrupting the growth of subungual connective tissue, hyperkeratosis of the nail bed (as in psoriasis), and capillary shunting, all of which may cause a depression in the distal matrix and lead to concave nail plates.[2,5,6] In this patient's case, we hypothesize that the frequent vasoconstriction of peripheral vessels in the fingers from Raynaud's led to poor blood flow and hypoxia of the nail matrix, leading to altered formation of the distal nail matrix.

Koilonychia can be accompanied by thin and brittle nails.[2] It is classically associated with iron deficiency, which may be due to nutritional deficiencies, gastrointestinal bleeding, or Plummer-Vinson syndrome.[2] Interestingly, koilonychia has also been reported in up to 49% of patients with hemochromatosis, although phlebotomy does not appear to reverse the condition.[7] Koilonychia is also associated with endocrinopathies such as thyroid disorders and diabetes; nutritional deficiencies in amino acids, vitamin C, zinc, copper, and selenium; occupational exposures to petroleum-based products, ammonium thioglycolate, and mineral oils; and living at a high altitude.[2] Early childhood koilonychias are often secondary to tight-fitting shoes or finger sucking, both of which resolve with behavior modification.[2,8] Hereditary cases of the disease also occur in an autosomal dominant pattern. Rarely, koilonychia has been observed in patients with systemic lupus erythematous and Raynaud's phenomenon, typically involving abnormal proximal nail fold capillary loops, splinter hemorrhages, and red lunulae—signs that were not present in our case.[2,9]

The diagnosis of koilonychia is often clinical, requiring a complete history, review of systems, and physical examination.[2] While some causes of koilonychia are idiopathic, the workup of the disease should include iron concentration studies, complete blood counts, and when an autoimmune disease is suspected, inflammatory markers and autoimmune disease antibodies in order to identify and manage underlying causes of koilonychia. Onychomycosis and inflammatory skin diseases (eg, psoriasis, alopecia areata, lichen planus) should also be considered.[2] Because the concavity of nails may be difficult to detect by visual inspection, the water-drop test, whereby a few drops of water are placed on the patient's nail, may facilitate the diagnosis of koilonychia.[1] If the water droplets pool on the nail plate without sliding off, the concave shape of the nails is verified.[1]

As Raynaud's phenomenon affects an estimated 11% of middle-aged women and 8% of middle-aged men,[10] clinicians should be aware that koilonychia may be a potentially reversible manifestation of Raynaud's phenomenon. This case serves as an important reminder that nail changes are often manifestations of systemic disease. Further research is needed to elucidate the pathophysiology of koilonychia, but this case suggests that diminished digital blood flow to the nail matrix may be contributory. With proper detection and treatment, koilonychia secondary to Raynaud's phenomenon can be managed successfully.

REFERENCES

1. Chelidze K, Lipner SR. The water-drop test for the diagnosis of koilonychia. *J Am Acad Dermatol*. 2017;77(6):e157-e158.

2. Walker J, Baran R, Vélez N, Jellinek N. Koilonychia: an update on pathophysiology, differential diagnosis and clinical relevance. *J Eur Acad Dermatol Venereol*. 2016;30(11):1985-1991.
3. Solomons HD. Raynaud's phenomenon. *Cardiovasc J Afr*. 2011;22(5):233.
4. Chen SX, Cohen PR. Parrot beak nails revisited: case series and comprehensive review. *Dermatol Ther (Heidelb)*. 2018;8(1):147-155.
5. Stone OJ. Clubbing and Koilonychia. *Dermatol Clin*. 1985;3(3):485-490.
6. Rathod DG, Sonthalia S. Spoon nails (Koilonychia). In: *StatPearls*. StatPearls Publishing; 2020. Accessed October 8, 2020. Available at: http://www.ncbi.nlm.nih.gov/books/NBK559311/
7. Chevrant-Breton J, Simon M, Bourel M, Ferrand B. Cutaneous manifestations of idiopathic hemochromatosis. Study of 100 cases. *Arch Dermatol*. 1977;113(2):161-165.
8. Starace M, Alessandrini A, Piraccini BM. Nail disorders in children. *Skin Appendage Disord*. 2018;4(4):217-229.
9. García-Patos V, Bartralot R, Ordi J, Baselga E, de Moragas JM, Castells A. Systemic lupus erythematosus presenting with red lunulae. *J Am Acad Dermatol*. 1997;36(5):834-836.
10. Suter LG, Murabito JM, Felson DT, Fraenkel L. The incidence and natural history of Raynaud's phenomenon in the community. *Arthritis Rheum*. 2005;52(4):1259-1263.