Pincer nails in a patient with systemic lupus erythematosus and lupus nephritis: A case report



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INTRODUCTION

Pincer nail is the transverse overcurvature of the nail plate which can lead to painful pinching of underlying tissues, loss of soft tissue of the involved fingers, and rarely bone resorption of the terminal phalynx.¹ Pincer nails may be congenital or acquired. Acquired pincer nail deformity has multiple associations including ill-fitting shoes, medications such as β -blockers, psoriasis, and various systemic diseases, including gastrointestinal malignancy and renal failure.^{2,3} A case of pincer nails in association with systemic lupus erythematosus (SLE) was described in 2005, but, to our knowledge, no further cases have been reported.³ We present a case of pincer nail deformity associated with SLE and lupus nephritis.

CASE REPORT

A 25-year-old man with SLE, lupus nephritis, and refractory myasthenia gravis was admitted with a 2week history of weakness secondary to hyperkalemia. The dermatology department was consulted for painful, bleeding toes.

The patient was diagnosed with SLE at age 9 when he presented with fever, joint pain, and lethargy and was found to have serologies consistent with SLE (+antinuclear, +anti-double-stranded DNA, +anti-smooth muscle, +anti-ribonucleoprotein, +anti-cardiolipin, +anti-Ro, + anti-La antibodies). Later that year, proteinuria developed, and a renal biopsy confirmed lupus nephritis class V. Treatment

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Abbreviation used: SLE: systemic lupus erythematosus

had included systemic steroids, cyclophosphamide, mycophenolate mofetil, azathioprine, plaquenil, and, most recently, a combination of prednisone and cyclosporine. Hemodialysis was initiated at age 23. That same year he was diagnosed with myasthenia gravis, leading to respiratory failure requiring a tracheostomy. Plasma exchange, rituximab, and intravenous immunoglobulin had minimal benefit.

Prior to his admission, the patient had been hospitalized elsewhere for ventilator-associated pneumonia, preventing him from undergoing scheduled plasmapheresis. He subsequently presented to the emergency department with muscle weakness and was found to have a potassium level of 7.8 mEq/L, blood urea nitrogen of 86 mg/dL, creatinine of 4.04 mg/dL, and electrocardiogram changes consistent with hyperkalemia. Medications on admission included prednisone, cyclosporine, carvedilol (initiated at age 23), budesonide, ipratropium, alprazolam, esomeprazole, sulfamethoxazole-trimethoprim, and vancomycin.

The dermatology department was consulted for a 1-month history of painful bleeding toes. There was no history of similar pain or trauma, and he reported seeing a podiatrist for routine care. There was no family history of nail deformities.

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Fig 1. Pincer nail of great toe sparing smaller digits.



Fig 2. Pincer nail, view of fingertip.

Examination revealed pincer deformities of all fingernails and both halluces, sparing the smaller toes (Figs 1 and 2), which, according to the patient, had been present for 2 years. There was serosanguinous drainage from both sides of the left great toenail and granulation tissue on the lateral aspect of the right great toenail. Additional findings included dolichonychia, arachnodactyly, half-and-half leukonychia, and transverse ridging of the nails.

Potassium hydroxide preparation from the scale at the nail folds, fungal culture, and Periodic acid-Schiff staining from a nail clipping were negative for fungus. Wound cultures from the periungual drainage were also negative for bacteria or fungus.

Initially, the periungual granulation tissue was treated with topical silver nitrate. However, the pain and granulation tissue persisted, so, after local anesthesia, the granulation tissue was curetted and silver nitrate reapplied. Daily vinegar soaks resulted in symptomatic improvement.

DISCUSSION

Acquired pincer nail deformity has been reported in association with numerous systemic diseases, although its pathogenesis is not well defined. It has been hypothesized that pincer nails result from enlargement of the base of the distal phalanx; because the nail matrix is firmly attached, the increase in tissue results in a reduced curvature

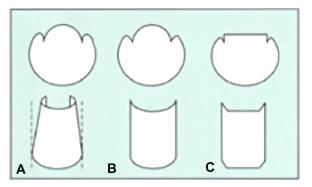


Fig 3. Varieties of pincer nail. **A**, pincer or trumpet type; **B**, tile shaped; and **C**, plicatured shaped. *Adapted from* Baran, R, Dawber, RPR, Haneke, E, et al. (2005) *A text atlas of nail disorders*, 3rd ed. London: Martin Dunitz.

proximally and greater curvature distally.⁴ There are 3 subtypes of nails with increased transverse curvature (Fig 3): (1) pincer or trumpet type, (2) tile shaped, and (3) plicatured shaped.⁴ The reported patient had the tile-shaped variety.

Congenital pincer nails tend to be symmetric in distribution and are associated with systemic findings, as in yellow nail syndrome and hidrotic ectodermal dysplasia.⁴

Ill-fitting shoes are one of the most common causes of acquired pincer nails. Other causes include onychomycosis, repeated trauma, nail avulsions, tumors of the distal phalynx such as mucus cysts, subungual exostoses or periungual pyogenic granuloma, psoriasis, osteoarthritis of the distal interphalangeal finger joints, and medications such as β -blockers. Of note, although our patient does take a β -blocker, this medication was initiated after the development of his nail changes. Numerous systemic diseases have been associated with pincer nail deformities including gastrointestinal malignancy, renal failure, Kawasaki disease,⁵ amyotrophic lateral sclerosis,⁶ and SLE.³

Nail changes are common in association with SLE, affecting up to 31% of patients.⁷ The most prevalent findings are prominent nail fold capillaries, vascular infarction, and red lunulae.³ Pincer nails have only been described once in association with SLE; Majeski et al.³ reported pincer nails in a patient with a 4-month history of SLE, who also had other cutaneous manifestations of his disease including oral ulcers and photosensitivity. In our case, the deformity developed 12 years after the diagnosis of SLE, and there were no other cutaneous manifestations of SLE.

Another possible explanation for pincer nails in this case lies with the development of renal impairment. More than 70% of uremic patients have some degree of nail abnormality,⁸ and widespread pincer

nail changes in patients with long-term renal failure have been described.² Kirkland and Sheth² reported the development of pincer nails in a patient with worsening renal function. One hypothesis states that the nail changes seen in renal failure could be the result of secondary hyperparathyroidism. Effective hemodialysis has not been found to reverse nail findings in patients with renal failure, leading some to suggest that long-term uremia may be causative.⁹ In our case, the deformity developed 12 years after the development of lupus nephritis but within months of his renal function deteriorating to the point of requiring hemodialysis.

Pincer nails have been directly attributed to hemodialysis in some patients. Although half-andhalf nails and absent lunulae are the most commonly reported nail changes in patients undergoing hemodialysis,¹⁰ pincer nails have been reported in 2% of patients and tend to affect the arm with the arteriovenous fistula.^{11,12} On average, the nail deformity presents 2 years after fistula creation and resolves with fistula reversal, suggesting that local alterations in microcirculation and resultant ischemia or venous hypertension led to the pincer deformity in this setting. In contrast, our patient had pincer deformity of all fingernails and several toenails.

It is difficult to determine with certainty whether the pincer nail deformity in our patient is caused by SLE or renal failure. It is possible that his nail deformity is multifactorial, and further investigation into the pathogenesis of pincer nail deformities is needed to better elucidate its relationship to systemic disease.

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