

Appending the appendages: New perspectives on Netherton syndrome and green nail syndrome



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Dermatologists treat diseases of the skin, hair, and nails. In this issue of the *Journal of the American Academy of Dermatology*, there are 2 articles that append our knowledge of skin appendages.

Netherton syndrome (NS) (OMIM 256500) is a rare autosomal recessive disorder due to germline mutation of *SPINK5*, affecting 1 in 100,000 to 200,000 live births. Congenital ichthyosiform erythroderma and/or ichthyosis linearis circumflexa, hair shaft abnormalities (notably trichorrhexis invaginata [TI]), and an atopic diathesis (elevated serum immunoglobulin E) characterize the syndrome. Neonatal cases may be complicated by hypernatremic dehydration and failure to thrive. NS is often misdiagnosed as atopic dermatitis because of the presence of eczematous skin lesions and other allergies.¹

Affected hair in NS appears short, dry, dull, and brittle. TI, or bamboo hair, is the invagination of affected hairs caused by softness of the cortex in the keratogenous zone. TI may not be obvious in infants with NS, making the diagnosis of NS difficult. Dermoscopy and reflectance confocal microscopy may be the best way to observe TI.² The absence of TI does not exclude a diagnosis of NS.¹ TI often improves with age and may resolve completely. Utsumi et al³ described a band-like pattern in hairs with polarized light in 8 patients with NS, which were observed more often than TI. These findings were not present in healthy control individuals or patients with atopic dermatitis. A limitation of the study was that the procedure was not compared to other disorders of keratinization—further research will determine if this has the potential to be used as a diagnostic marker for NS.

Abbreviations used:

GNS: green nail syndrome
NS: Netherton syndrome
TI: trichorrhexis invaginata

Green nail syndrome (GNS), usually due to *Pseudomonas aeruginosa*, is a triad of variably green discoloration of the nail plate, proximal paronychia, and distal onycholysis, usually involving only 1 or 2 nails. An instantaneous diagnosis of GNS may obviate scrutiny for an associated nail disorder(s). GNS may accompany other diseases (onychomycosis, psoriasis, or paronychia), especially in the context of a moist environment.⁴

Surprisingly, there have been remarkably few studies defining the relationship of GNS with onychomycosis. Ohn et al⁵ performed a retrospective study of 23 cases of GNS in which PAS or GMS stains of clippings were performed. Five (21.7%) patients had immunosuppressive conditions, including internal malignancies, autoimmune disorders, and diabetes mellitus. A previous history of nail diseases was reported in 13 (56.5%) patients, including 12 patients with a history of onychomycosis and 1 patient with nail psoriasis. Eighteen (78.3%) affected nails were toenails, and 5 (21.7%) were fingernails. Involvement of the great toenail or thumbnail was found in 22 (95.7%) patients. Fungal coinfection was noted in 15 (65.2%) patients. Ohn et al concluded that onychomycosis was frequently associated with GNS and might be a predisposing factor of GNS. They recommended that the nails in patients with GNS be carefully examined to detect possible fungal infections.

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I started to turn green after thinking about how I may not have been managing GNS optimally—there is much more to GNS than eradicating its color.

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