Bullous Lichen Planus of the Nails

Nail involvement may occur in 1% to 10% cases of lichen planus (LP) and mostly in the setting of widespread cutaneous disease. Longitudinal ridging, thinning, and distal splitting of the nail plate are the most common nail changes in LP.

Report of a Case | A man in his 60s presented with painful swelling of the proximal nail folds, drooping of blood from his fingernails and toenails for the last year, and occasional pus discharge. There was no history of preceding trauma or drug intake.

On physical examination, the proximal nail folds were found to be swollen and showed violaceous discoloration of 7 fingernails and 3 toenails. The index fingernails also demonstrated longitudinal ridging, thinning, and focal fragmentation of the nail plate, while the remaining fingernails and involved toenails showed partial to complete loss of nail plate with oozing of blood resulting in hemorrhagic crusting of the nail beds and folds (Figure 1A). Oral mucosa showed lichenoid plaques on the right angle of mouth, bilateral buccal mucosa, and erosions covered with hemorrhagic crusts on the upper and lower lips. A provisional diagnosis of bullous lichen planus of the nails was made. Laboratory investigations revealed raised liver enzyme levels and positive anti-hepatitis C virus antibody status.

The patient was treated with oral antibiotics for 1 week and a topical steroid-antibiotic combination for 6 weeks, and the hemorrhagic crusting and nail fold swelling subsided completely, resulting in anonychia of the affected nails (Figure 1B). A skin biopsy was performed from the proximal nail fold of the right index finger 1 month after the initial presentation to avoid secondary changes due to infection that was present at the time of the initial presentation. Histopathologic examination demonstrated hyperkeratosis, hypergranulosis, acanthosis, basal cell degeneration, numerous apoptotic keratinocytes in the epidermis with dense bandlike lymphohistiocytic infiltrate in the papillary dermis consistent with the diagnosis of LP (Figure 2).

Discussion | Nail LP (NLP) usually presents in association with cutaneous, mucosal, or scalp lesions but may be the sole manifestation of the disease. It may involve the nail matrix, proximal and lateral nail folds, nail bed, and/or the hyponychium. Proximal matrix involvement resulting in longitudinal ridging is the most common and the earliest manifestation of NLP. Trachyonychia usually occurs in children as an isolated finding in the absence of LP at other sites and of other typical signs of NLP. Pterygium, irregular nail pitting, onychorrhexis, crumbling, and fragmentation of the nail plate are other clinical manifestations of nail matrix involvement. Nail bed involvement may take the form of small red papules visible as violaceous lines or papules through the nail plate, subungual hyperkeratosis, and onycholysis. Complete destruction of the nail matrix and nail bed results in total nail atrophy. Idiopathic atrophy of the nails is a distinct manifestation of NLP that results in rapid and diffuse scarring of nails in children. Less frequent signs of NLP described by Tosti et al. include erythematous patches of the lunula, melanonychia, splinter hemorrhages, koilonychia, and yellow nail syndrome-like changes.

Our case exemplifies the unusual presentation of bullous LP of the nails in the form of hemorrhagic lesions resulting in complete shedding of the nail plate and nail atrophy. Bullous LP of the nails is an uncommon and extreme variant of LP that may be associated with bullous and ulcerative lesions on the feet and toes, with or without cicatricial alopecia and oral involvement. Diagnosis is often challenging, and the presence of characteristic lesions at other sites in conjunction with histopathologic examination aid in confirmation of the diagnosis.

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Atrophoderma Vermiculatum: A Cutaneous Feature of Loeys-Dietz Syndrome

Atrophoderma vermiculatum (AV) is a rare skin disorder, typically presenting in childhood with a reticular pattern of skin atrophy on the cheeks, preauricular area, and forehead that seems to result from inflammation around follicular plugs. The causative mechanism leading to AV is unclear. It can present as an isolated skin manifestation, or AV can be part of other conditions including genetic disorders. Herein we report the association of AV with TGFB2-related Loeys-Dietz syndrome (LDS).

Report of Cases | Patient 1. A 12-year-old boy with cardiac (widened aortic root and pulmonary artery, patent ductus arteriosus) and skeletal abnormalities (sagittal craniosynostosis, thoracic scoliosis, lordosis, pectus excavatum, long extremities, vertical talus, varus deformity, hyperlaxity) was diagnosed with LDS. Furthermore, dysmorphic facial features, amelogenesis imperfecta, and a high arched palate with lobulated uvula were also noted. A de novo pathogenic mutation in TGFB2 (c.1639G>C p.Asp547His) was identified. At age 4 years, the patient had developed atrophic skin lesions on both cheeks, which stabilized after a few months of progression. At age 10 years, both cheeks showed reticulate atrophic lesions with a few follicular papules, which was diagnosed as AV (Figure, A).

Figure 1. Bullous Lichen Planus of the Nails

A. Before treatment, swelling and violaceous discoloration are apparent on the proximal fingernail folds along with oozing of blood, hemorrhagic crusting of the fingernails, and pus discharging from the lateral nail folds. B. After treatment, the hemorrhagic crusting and paronychia have completely resolved, with resultant atrophy of the fingernails.
Patient 2. A 12-year-old boy (Figure, B) had been born with a cleft palate, associated Pierre Robin sequence, and talipes equinovarus. At age 6 years, he was noted to have mild reticulate scarring fitting the diagnosis of AV on both cheeks as well as milia in the infra-orbital region and the auricles. He also had cardiac (enlarged aortic annulus and aortic sinuses, aortic root dilatation, and bilateral carotid artery tortuosity) and skeletal abnormalities (postural scoliosis, mild pectus excavatum, metatarsus varus, joint hypermobility, bilateral fifth finger clinodactyly, and clinodactyly of left proximal interphalangeal joints II and III) and was diagnosed with LDS. In addition, a left inguinal hernia and a bifid uvula were documented. A de novo heterozygous mutation in \textit{TGFBR2} (c.1546_1557del12bp; p.Val516_Asp519del) was identified.

Discussion | Both of the described patients had AV and LDS due to heterozygous \textit{TGFBR2} mutations; LDS is characterized by aggressive arterial aneurysms and widespread systemic involvement. To our knowledge, only 2 patients with AV and an aortic aneurysm syndrome have been reported in the literature. The first, a 14-year-old boy, was evaluated for Marfan syndrome, but genetic analysis of the \textit{FBNI} gene was not completed at the time of publication.\textsuperscript{2} The second, 24-year-old patient, was included in a study of 25 patients with LDS.\textsuperscript{3} She had multiple facial milia around the eyes and malar AV. A mutation in \textit{TGFBR2} was identified. Of additional interest, 3 other patients in that same study also had facial milia and a \textit{TGFBR2} mutation.\textsuperscript{3} Therefore, milia may be a feature of \textit{TGFBR2}-related LDS.

Both AV and LDS are quite rare, so the association of these 2 conditions in several cases does not seem simple coincidence. It is difficult to explain the occurrence of AV in patients with LDS. The pathogenesis of LDS is aberrant TGF-\(\beta\) signaling due to mutations in \textit{TGFBR1/2}, but many questions still remain. Similarly, the pathogenesis of AV is largely unknown.

It has been hypothesized that the keratinocytes in patients with AV mediate release of inflammatory cytokines in response