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Single Case

Bullous Lichen Planus of the Nails: A Case Report and Review of the Literature

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Keywords
Nail · Nail disease · Nail disorder · Nail inflammatory disease · Nail pathology

Abstract
Lichen planus is a chronic inflammatory disorder that may affect the skin, nails, and/or oral mucosa. Bullous lichen planus is a rare variant of lichen planus, which is even less common in the nails. We present a case of nail bullous lichen planus, in a 48-year-old male presenting with a 10-month history of onychodystrophy of all ten fingernails. A longitudinal excision of the left thumbnail was performed, with histopathology consistent with lichen planus with focal transition to bullous lichen planus. He was treated with intralesional triamcinolone injections to the fingernails monthly, with improvements noted after three treatments. Our patient's nail bullous lichen planus manifested with longitudinal ridging, white-yellow discoloration, onycholysis, subungual hyperkeratosis, and v-shaped nicking. Histopathological findings included classical lichen planus changes, as well as formation of subepidermal bullae, colloid bodies, and extensive inflammatory infiltrate. Increased awareness and high index of suspicion for this condition are necessary, given the often late diagnosis reported in previously published cases.

Introduction

Lichen planus (LP) is a chronic inflammatory disorder that can affect the skin, nails, and/or oral mucosa [1]. Bullous LP (BLP) is a rare variant of LP, which is even less common in the nails [2]. Herein, we present a case of nail BLP, associated clinical and histopathological findings, and subsequent treatment.

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Case Report

A 48-year-old male presented with a 10-month history of onychodystrophy of all ten fingernails. He denied any bleeding, pus, or prior nail trauma. Six months prior to the visit, he had a rash in the axilla that was confirmed by biopsy with histopathology as LP. His rash had resolved by the time he presented to us. He had no other prior medical history. On physical examination, his fingernails exhibited longitudinal ridging and white-yellow discoloration proximally (shown in Fig. 1a, b), as well as onycholysis, subungual hyperkeratosis, and v-shaped nicking of his right thumbnail (shown in Fig. 1c). There were no involvement of the toenails, no Wickham striae noted in his oral mucosa, and no patches of hair loss or scarring of the scalp. The differential diagnosis included nail psoriasis, nail LP, onychomycosis, brittle nail syndrome, and traumatic onycholysis. A longitudinal excision of the left thumbnail was performed, with histopathology showing a cell-poor subepidermal bulla (shown in Fig. 2) in association with a focal lymphocyte-mediated interface dermatitis and colloid body formation (shown in Fig. 3). C3d, C4d, and IgG4 immunohistochemical studies did not disclose any linear basement membrane zone deposits. These findings were consistent with nail LP with focal transition to nail BLP. Since more than three nails were affected, the patient was offered intramuscular triamcinolone injections. However, since he was concerned about potential systemic side effects, he opted for local treatment. He was treated with intralesional triamcinolone injections (2.5 mg/mL in 1% lidocaine using topical ethyl chloride spray) to the fingernails monthly, which was well tolerated. Improvements were noted after three treatments.

Discussion

Nail LP is a chronic inflammatory disorder that may present with longitudinal ridging, red lunular discoloration, nail plate thinning, koilonychia, trachyonychia, onycholysis, subungual hyperkeratosis, nail plate atrophy, as well as anonychia and dorsal pterygium in end stages [1]. It can occur independently or concurrently with involvement of the skin and/or oral mucosa. It is considered a true nail emergency, requiring prompt and adequate treatment to prevent permanent nail loss [3]. BLP of the nails is a rare variant of nail LP. It most often occurs sporadically, but familial cases have been reported [2, 4]. Aside from typical nail LP findings, it has also been associated with nail hemorrhage. Similar to the classic form, it can involve both the fingernails and toenails, and lead to nail plate atrophy and dorsal pterygium [5]. Concomitant bulla and ulcerations of the feet and toes, white striations or plaques of the oral mucosa with erythema or ulcerations, as well as cicatricial alopecia have also been described [6].

The pathogenesis of BLP involves an autoimmune cytotoxic T-cell-mediated response targeting the basilar and parabasilar layers of the skin [2]. This leads to a dermal-epidermal separation that can be apparent both clinically and with light microscopy. Cutaneous BLP may present as vesicles or tense bullae, developing near or on pre-existing LP papules. BLP must be distinguished from LP pemphigoides (LPP), in which the formation of bullae is due to circulating autoantibodies against basement membrane antigens [7]. Cases of BLP and LPP can often be difficult to differentiate clinically, but nail and mucosal involvement are more common with BLP [2, 7].

Nail BLP has been described in 7 other case reports, as well as 1 case series including 14 patients with nail involvement (shown in Table 1) [4–6, 8–10]. These cases have had variable involvement of the fingernails (50.0%) and/or toenails (70.0%), with an average of 5.5 years to diagnosis (range 2 months–13 years). Toenail involvement was more common than

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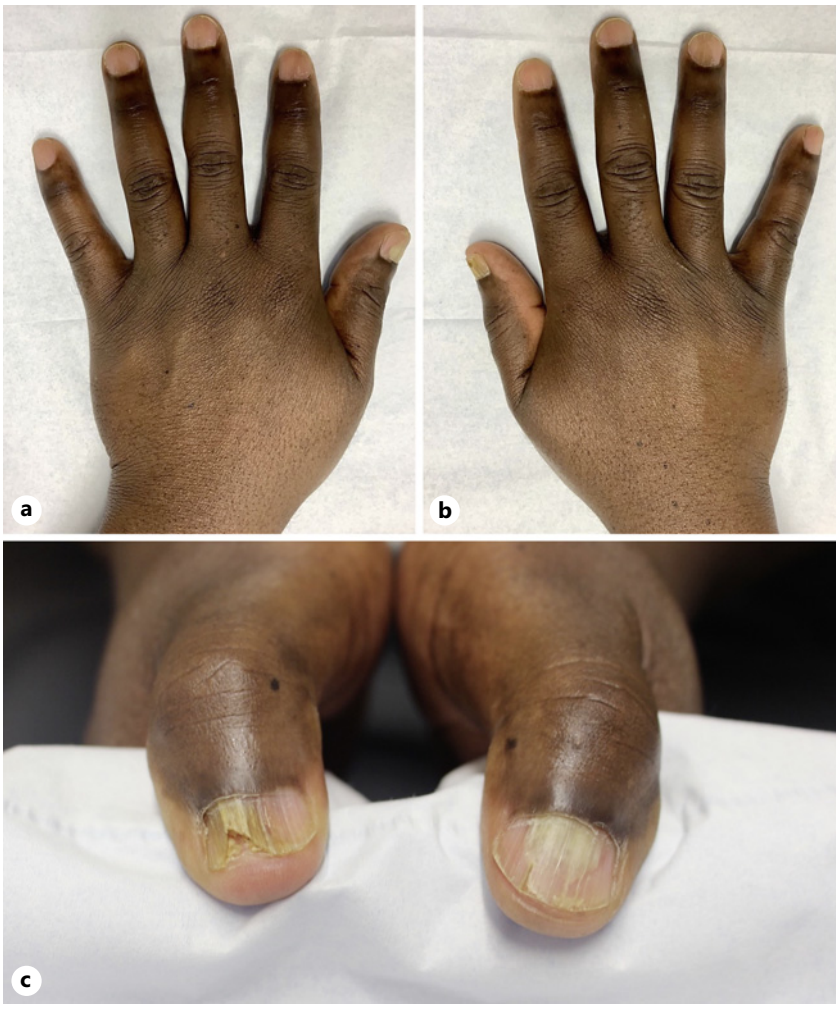

Fig. 1. Clinical image of left fingernails (**a**) and right fingernails (**b**) exhibiting proximal yellowing and longitudinal ridging, with right thumbnail (**c**) exhibiting proximal yellow discoloration, subungual hyperkeratosis, and v-shaped nicking.

fingernail involvement, in contrast to fingernails being more often involved than toenails in classic LP (94% vs. 54%) [1]. In general, when BLP was diagnosed early (<2 years), patients presented with onychorrhexis, nail plate hyperkeratosis, and onycholysis, similar to that of classic nail LP. When diagnosed late (5–10 years), there was often hemorrhagic crusting and partial or complete nail plate loss. Prior history or simultaneous skin involvement was noted in almost all cases (95.0%), with some having oral mucosal involvement (40.0%) and/or cicatricial alopecia (15.0%). In one case, a 30-year-old male presented with yellow, markedly thickened fingernails and toenails with longitudinal ridging, resembling yellow nail syndrome, a rare disorder characterized by a triad of yellow nails, lymphedema, and respiratory tract involvement [9]. Another BLP case in a 10-year-old boy was associated with the recombinant hepatitis B vaccine, presenting 2 months after receiving the vaccine [10]. In the case series of cutaneous BLP patients, 36 patients with BLP family history were compared to 21 patients without [4]. Of the familial BLP patients, 13 had involvement of the nails (36.1%), compared to only 1 patient (4.8%) in the nonfamilial cohort. Nail changes in the familial cases were variable and included dorsal pterygium, anonychia, onychorrhexis, nail plate hyperkeratosis, and onycholysis. Nail changes in the nonfamilial case were not described. Familial

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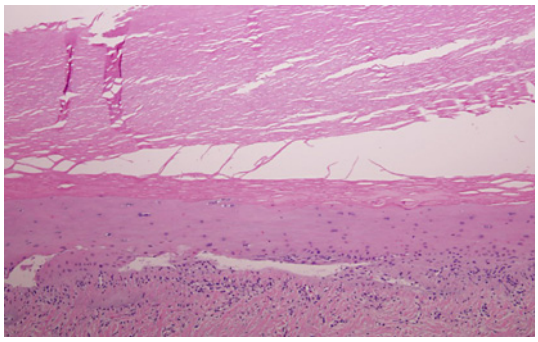

Fig. 2. Histopathology (H&E section, ×100) of nail biopsy specimen showing a pauci-inflammatory area of epidermal-dermal separation with overlying hyperkeratosis.

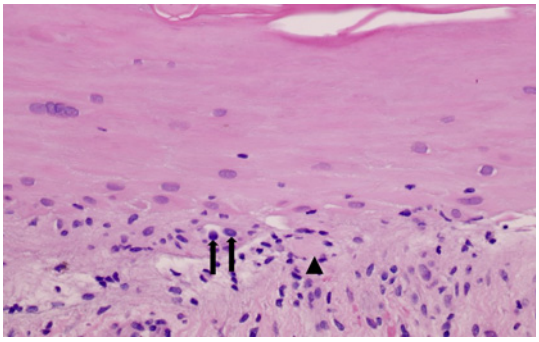

Fig. 3. Histopathology (H&E section, ×400) of nail biopsy specimen showing focal minimal lymphocyte-mediated interface dermatitis (arrows) with focal colloid body formation (arrowhead).

compared to nonfamilial cases had earlier age of BLP onset (average 9.6 vs. 21.1 years old, respectively), as well as a more prolonged course (average 30.1 vs. 11.3 years, respectively). Notably, our patient was diagnosed earlier (10 months) compared to other BLP patients (average 5.5 years), but similar to almost all previous nail BLP cases, he had prior cutaneous LP involvement. Our patient did not have involvement of the oral mucosa or scalp. Given the often late diagnosis of nail BLP and variable involvement of skin, scalp, and mucosa, increased awareness and a high index of suspicion for this condition are necessary. Specifically, diagnosis of the bullous variant of nail LP may frequently be missed, given the similar clinical presentation to that of classical nail LP, as illustrated with our patient. Overall, both nail LP and nail BLP may be more common than currently reported. Further workup and biopsy are warranted in any suspected cases.

Accurate diagnosis of nail LP, including nail BLP, requires a biopsy with histopathological correlation 11. A longitudinal excision is suggested to ensure sufficient sampling of nail matrix and nail bed. Characteristic histopathologic findings of nail LP include acquisition of a granular cell layer with hypergranulosis of the nail matrix and bed epithelium. There is also a band-like lymphocytic infiltrate within the superficial corium, with lymphocyte apposition to the basal layer keratinocytes and ensuing epithelial destruction [2]. BLP histopathological findings include these classical LP changes, as well as formation of subepidermal bullae with prominent collections of colloid bodies [7]. Extensive T-cell-mediated injury to the basal layer results in bullae formation referred to as subepidermal Max-Joseph spaces. Colloid or civatte bodies are cytoplasmic remnants of apoptotic keratinocytes that can be identified in the superficial dermis [2]. In differentiating BLP from LPP, colloid bodies are often more prominent in BLP. With LPP, C3d, C4d, and IgG4 studies conducted on paraffin-embedded tissue, as well as IgG and C3 direct immunofluorescence, will show a linear homogeneous deposition pattern within the epidermal basement membrane zone [7].

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Table 1. Summary of demographics, clinical findings, histopathology, and treatment of cases of BLP involving the nails in the literature

Reference	Age/sex	Nails	Nail findings	Cutaneous/oral findings	Histopathology	Nail treatment
Gardner et al. [8], 1955	80/F	Fingernails, toenails	Violaceous papules of nail beds Atrophic fingernails and toenails with pterygium Duration of 5 years	Discrete and confluent violaceous papules on heels and instep with deep-seated vesicles White plaques on dorsal tongue and buccal mucosa	Left medial foot: moderate hyperkeratosis, prominent stratum granulosum, moderate acanthosis, edema and dissociation of basilar layer, migration of lymphocytes into basilar layer, sharply marginated band of infiltration in upper corium consisting of small round cells, and an occasional histiocyte	No treatment noted
Cram et al. [6], 1966	44/F	Toenails	Atrophy or absence of toenails Duration of 12 years	Intermittent erythema and spontaneous bullae of the feet Papules of LP in extremities Cicatricial alopecia	Biopsy not performed	No treatment noted
	55/F	Toenails	Absence of all toenails Duration of 13 years	Bullae and chronic ulceration of the feet Cicatricial alopecia No oral mucosal involvement	Biopsy not performed	No treatment noted
	54/F	Toenails	Absence of all toenails Duration of 10 years	Erythema and bullae of the feet LP on volar wrists Ulcerations on toes and plantar feet Cicatricial alopecia Oral mucosal involvement	Biopsy not performed	No treatment noted

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Table 1 (continued)

Reference	Age/sex	Nails	Nail findings	Cutaneous/oral findings	Histopathology	Nail treatment
Haneke et al. [9], 1982	30/M	Fingernails, toenails	Yellow and markedly thickened fingernails and toenails with longitudinal ridging Duration of 2 years	No cutaneous/oral findings noted	Nail: marked hypergranulosis, thick compact orthokeratotic horny layer, few exocytotic mononuclear cells in lower spinous layer, marked liquefaction degeneration of basal cell layer, dense band-like cellular infiltrate of lymphocytes and histiocytes with weakly PAS-positive cytoid bodies in subepithelial layer, fibrin deposits along BM, upper corium, and in cytoid bodies	Chloroquine diphosphate *x* 11 weeks → no improvement
Miteva et al. [10], 2005	10/M	R3/R4 fingernails	Onychorrhexis of R3 fingernail Onychodystrophy of R4 fingernail Associated with recombinant HBV vaccine 1 and 2 months prior to onset of symptoms	Numerous violaceous polygonal flat-topped papules and plaques on trunk, arms, and legs Clear bullae observed on lesional skin No oral mucosal involvement	Left leg: hyperkeratosis, focal hypergranulosis, basal cell degranulation, band-like lymphohistiocytic infiltrate in upper dermis along DEJ, few eosinophils Skin bullous lesion: subepidermal blister with destruction of basal cells and dense lymphomononuclear infiltrate in upper dermis, DIF: IgM deposits in DEJ and cytoid bodies in dermis, IF: no circulating antibodies	No treatment noted
Huang et al. [4], 2005	14 patients	Fingernails (n = 5), toenails (n = 8), unspecified (n = 1)	Dorsal pterygium, anonychia, onychorrhexis, nail plate hyperkeratosis, onycholysis 13 familial BLP patients, 1 nonfamilial	Cutaneous involvement noted in 13/13 familial BLP patients Oral mucosal involvement noted in 4/13 familial BLP patients	Histopathological findings not presented	No treatment noted

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Table 1 (continued)

Reference	Age/sex	Nails	Nail findings	Cutaneous/oral findings	Histopathology	Nail treatment
			Earlier age of onset and higher nail involvement in familial patients	Findings not specified in 1 nonfamilial BLP patient		
Khullar et al. [5], 2015	60s/M	Fingernails, toenails	Swelling and violaceous discoloration of proximal nail folds Longitudinal ridging, thinning, focal fragmentation of R2/L2 fingernails Partial to complete loss of nail plate with hemorrhagic crusting of nail beds and nail folds Duration of 1 year	Erosions with hemorrhagic crusting of lips Erosions with hemorrhagic crusting of lips	R2 fingernail: hyperkeratosis, hypergranulosis, acanthosis, basal cell degeneration, numerous apoptotic keratinocytes in epidermis with dense band-like lymphohistiocytic infiltration in papillary dermis	Topical steroid-antibiotic combination *x* 6 weeks → resolution of hemorrhagic crusting and nail fold swelling
Our patient	48/M	Fingernails	Longitudinal riding and proximal white-yellow discoloration of fingernails Onycholysis, subungual hyperkeratosis, and v-shaped nicking of R thumbnail Duration of 10 months	Prior cutaneous involvement of axilla No oral mucosal involvement No scalp involvement	Cell-poor subepidermal bulla in association with a focal lymphocyte-mediated interface dermatitis and colloid body formation No linear basement membrane zone deposits seen with immunohistochemical studies	Intralesional Kenalog injections every 4–6 weeks × 3 rounds → improvement of onychodystrophy and onycholysis noted at 3-month follow-up visit

Since nail BLP is extremely rare, there are no guidelines or consensus on management, with only two cases in our review reporting treatment. A 60-year-old male with nail BLP was treated with an unspecified topical steroid-antibiotic combination for 6 weeks, resulting in resolution of hemorrhagic crusting and nail fold swelling [5]. A 30-year-old male with nail BLP was treated with chloroquine diphosphate for 11 weeks, but without improvement [9]. Our patient was treated with intralesional triamcinolone injections, commonly used in nail LP, but not previously reported for bullous forms [1]. Topical treatments are rarely effective in nail LP due to limited absorption through the nail plate. Intralesional/intramuscular triamcinolone injections have thus been recommended as first-line therapies, followed by oral retinoids or immunosuppressive agents [12]. We demonstrate efficacy of intralesional triamcinolone injections for the bullous variant, in addition to classical nail LP.

In summary, BLP is an LP variant that can rarely involve the nail. Our case corroborates prior literature on nail BLP and emphasizes the need for a high index of suspicion for the condition, given the frequent delay in diagnosis. We also demonstrate intralesional triamcinolone injections to the nails as a successful management option for this case of nail BLP. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000533386).

Statement of Ethics

Ethical approval is not required for this study in accordance with national guidelines. Written informed consent was obtained from patient for publication of details of their medical case and any accompanying images.

Conflict of Interest Statement

JKH and CCM have no conflicts of interest to declare. SRL has served as a consultant for Hoth Therapeutics, Ortho-Dermatologics, Belle Torus Corporation and Moberg Pharmaceuticals.

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Author Contributions

J.K.H., C.C.M., and S.R.L. contributed to writing and editing of this manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material. Further inquiries can be directed to the corresponding author.

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