A rare case of lichen planus pemphigoides with palmoplantar hyperkeratotic plaques

Patricia K. Mansfield, BS,[a] Rylee Moody, BS,[a] Yang Li, MD,[b] and Gillian Heinecke, MD[b,c]

Key words: lichen planus; lichen planus pemphigoides; palmoplantar hyperkeratotic plaques.

INTRODUCTION

Lichen planus pemphigoides (LPP) is an autoimmune blistering disorder, clinically characterized by the lichenoid features associated with lichen planus, along with the bullae associated with bullous pemphigoid.[1] LPP is an exceedingly rare diagnosis, with an incidence of around 1 per 1,000,000 patients per year.[1] Typically in this condition, bullae develop on both lichenoid and nonlichenoid skin, with a reported average lag time of 8.3 months between the onset of lichenoid dermatitis and the presentation of bullae.[2]

The bullae formation is due to antihemidesmosomal autoantibodies and therefore, the diagnosis requires direct immunofluorescence (DIF) of perilesional skin demonstrating basement membrane linear deposits of IgG or C3,[2] in conjunction with clinical and histopathologic findings. Additional DIF findings may include fibrinogen deposition along the dermoepidermal junction and Civatte body formation, associated with the lichenoid features that manifest clinically.

We present here a rare case of LPP, with a unique presentation of hyperkeratotic plaques on palmar and plantar surfaces.

Abbreviations used:

DIF: direct immunofluorescence
LPP: lichen planus pemphigoides
PPK: palmoplantar keratoderma

CASE REPORT

A 64-year-old Caucasian woman with no significant past medical history presented with a 2-month history of progressively worsening pruritic blisters on her hands, feet, and mouth. Before presentation with dermatology, triamcinolone 0.1% and clotrimazole cream had been prescribed but provided no improvement. Her medications at time of presentation included buspirone, celecoxib, citalopram, cyclobenzaprine, gabapentin, lisinopril-hydrochlorothiazide, and simvastatin; of note, she denied any changes to her medications within the last year.

Skin examination revealed erythematous papules on the lower portion of the legs and wrists coalescing into hyperkeratotic plaques on her palms and soles (Fig 1). There were also superficially eroded and lacey appearing white papules and plaques on the lower mucosal lip, buccal mucosa, and tongue (Fig 2). At the time of initial presentation, there were no active bullae; however, the patient did report a history of blisters on her palms and soles. Laboratory workup including complete blood count, comprehensive metabolic panel, hepatitis B, hepatitis C, and HIV revealed no abnormalities.

Punch biopsies from lesional and perilesional skin on the shin showed compact hyperorthokeratosis, hypergranulosis, saw-tooth epidermal hyperplasia, lichenoid infiltrate obscuring the dermoepidermal junction, and necrotic keratinocytes, consistent with the typical features of lichen planus (Fig 2). DIF demonstrated linear deposition of IgG and C3 along the basement membrane, focal shaggy basement

From the Saint Louis University School of Medicine, St. Louis, Missouri[a]; Department of Dermatology, Saint Louis University, St. Louis, Missouri[b]; and SLUCare, SSM Health, St. Louis, Missouri.[c]

Funding sources: None.

Patient consent: The authors obtained written consent from patients for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available. Patient consent forms were not provided to the journal but are retained by the authors.

IRB approval status: Not applicable.

Correspondence to: Patricia K. Mansfield, BS, Saint Louis University School of Medicine, St. Louis, MO. E-mail: katie.mansfield@health.slu.edu.

JAAD Case Reports 2024;49:14-6.
2352-5126

https://doi.org/10.1016/j.jdcr.2024.04.001

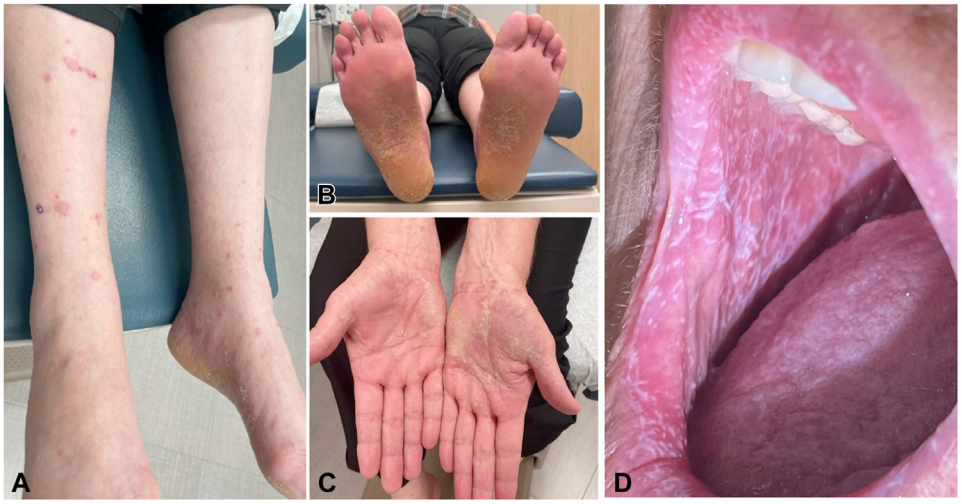

Fig 1. Physical examination demonstrates erythematous papules on **(C)** wrists and **(A)** lower portion of the legs coalescing into hyperkeratotic plaques on **(C)** palms and **(B)** soles of feet. **D**, Examination of her oral mucosa demonstrates reticulated white papules and plaques along with superficial erosions.

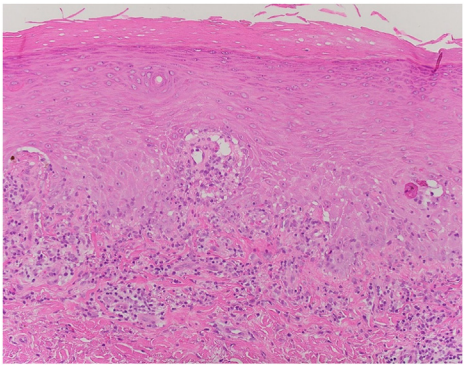

Fig 2. Histopathologic findings demonstrate compact hyperorthokeratosis, hypergranulosis, lichenoid infiltrate obscuring the dermoepidermal junction, saw-tooth epidermal hyperplasia, and occasional necrotic keratinocytes.

membrane zone staining with fibrinogen, and IgA and IgM colloid bodies. The clinicopathologic correlation was supportive of a diagnosis of LPP.

The patient was initially treated with topical triamcinolone ointment and a gradual taper of systemic corticosteroids. Mild improvement was made on this regimen after 2 months, but with persistent oral flares, methotrexate 15 mg weekly was started. With persistence of symptoms, new flares, and accompanying nausea associated with methotrexate use, methotrexate was switched to dapsone,[1] and patient now reports overall improvement of symptoms.

DISCUSSION

Differential diagnoses for clinical presentations of LPP include many of the various lichenoid diagnoses. Because the clinical manifestations of LPP (lichenoid dermatitis and bullae) are relatively nonspecific, diagnosing this condition requires clinical suspicion precipitating further workup, specifically, perilesional biopsy for DIF. DIF findings demonstrating linear deposits of IgG and C3 in the basement membrane zone, indicating an autoimmune antihemidesmosomal pathogenesis, allowed for diagnosis of LPP.[2,3]

Regarding treatment and management, most cases are successfully treated with systemic corticosteroids, although topical corticosteroids, dapsone,[1,2] methotrexate,[4] and dupilumab[5] have demonstrated improvement in specific cases. In our patient specifically, methotrexate showed little effect, whereas dapsone demonstrated symptom improvement. Further, in managing LPP, considerations should be made for previously reported associations and potential triggers of this autoimmune disease, including coexisting neoplasms and the use of angiotensin-converting enzyme inhibitors or simvastatin,[1] both of which this patient had been prescribed without changes to her medications in the last year.

Our patient's presentation with prominent hyperkeratotic plaques on palmar and plantar surfaces, is a rarer clinical presentation of this entity. To our knowledge, 4 cases have been published in the medical literature detailing cases of LPP presenting with palmoplantar keratoderma (PPK).[6-10] One of

these patients had a history of hereditary Unna-Thost PPK since childhood,[7] however, the 3 other cases presented with hyperkeratotic plaques on palmar and plantar surfaces in temporal association with the onset of LPP symptoms,[6,8,9] which was the case in our patient's clinical presentation. Further, biopsy of plantar lesions in 1 case confirmed typical lichen planus features on histology.[8] Previous case reports have hypothesized that autoantibody production and epitope spreading may be implicated in the pathogenesis of bullae and PPK lesions.[6,11] Our case of LPP presenting with hyperkeratotic plaques on palmar and plantar surfaces contributes to these previously reported temporal associations of LPP and PPK, allowing for stronger evidence for the consideration of an immunologic mechanism linking these clinical presentations.

Our case of LPP is both rare, with a reported incidence of 1 per 1,000,000 patients per year,[1] and unique, with hyperkeratotic plaques presenting on the patient's palmoplantar surfaces. The growing number of cases of temporal associations between presentations of LPP and PPK demonstrate a potential association and warrants further investigation.

Conflicts of interest

None disclosed.

REFERENCES

1. Hübner F, Langan EA, Recke A. Lichen planus pemphigoides: from lichenoid inflammation to autoantibody-mediated blistering. *Front Immunol*. 2019;10:1389. https://doi.org/10.3389/fimmu.2019.01389
2. Zaraa I, Mahfoudh A, Sellami MK, et al. Lichen planus pemphigoides: four new cases and a review of the literature. *Int J Dermatol*. 2013;52(4):406-412. https://doi.org/10.1111/j.1365-4632.2012.05693.x
3. Papara C, Danescu S, Sitaru C, Baican A. Challenges and pitfalls between lichen planus pemphigoides and bullous lichen planus. *Australas J Dermatol*. 2022;63(2):165-171. https://doi.org/10.1111/ajd.13808
4. Mueller KA, Cordisco MR, Scott GA, Plovanich ME. A case of severe nivolumab-induced lichen planus pemphigoides in a child with metastatic spitzoid melanoma. *Pediatr Dermatol*. 2023;40(1):154-156. https://doi.org/10.1111/pde.15097
5. Ch'en PY, Song EJ. Lichen planus pemphigoides successfully treated with dupilumab. *JAAD Case Rep*. 2023;31:56-58. https://doi.org/10.1016/j.jdcr.2022.11.011
6. Mizawa M, Makino T, Hayashi M, Furukawa F, Shimizu T. A case of lichen planus pemphigoides with palmoplantar keratoderma. *Eur J Dermatol*. 2018;28(1):100-101. https://doi.org/10.1684/ejd.2017.3160
7. Mokos M, Ljubojević Hadžavdić S. Coexistence of lichen planus pemphigoides, palmoplantar keratoderma of Unna-Thost, and atopic dermatitis. *Acta Dermatovenerol Croat*. 2022;30(2):110-112.
8. Yoon KH, Kim SC, Kang DS, Lee IJ. Lichen planus pemphigoides with circulating autoantibodies against 200 and 180 kDa epidermal antigens. *Eur J Dermatol*. 2000;10(3):212-214.
9. Oomen C, Temmerman L, Kint A. Lichen planus pemphigoides. *Clin Exp Dermatol*. 1986;11(1):92-96. https://doi.org/10.1111/j.1365-2230.1986.tb00431.x
10. Loh T-H, Yosipovitch G, Tay Y-K. Palmar-plantar keratoderma of Unna Thost associated with atopic dermatitis: an under-recognized entity? *Pediatr Dermatol*. 2003;20(3):195-198. https://doi.org/10.1046/j.1525-1470.2003.20302.x
11. Mignogna MD, Fortuna G, Leuci S, Stasio L, Mezza E, Ruoppo E. Lichen planus pemphigoides, a possible example of epitope spreading. *Oral Surg Oral Med Oral Pathol Oral Rad Endod*. 2010;109(6):837-843. https://doi.org/10.1016/j.tripleo.2009.12.044