Case Letters

OPEN

Brunsting–Perry pemphigoid: case report and review of current management

Keywords: autoimmune blistering dermatosis, Brunsting–Perry pemphigoid, cicatricial alopecia

Introduction

Brunsting–Perry pemphigoid (BPP) is a rare autoimmune disease on the pemphigoid spectrum that most commonly affects older males[1] and presents with tense pruritic bullae on the head and neck regions followed by cicatricial alopecia with minimal mucosal involvement.[2] Etiology is unclear, but possible triggers include sun exposure, mechanical trauma, medications, and internal malignancies.[3] It has been considered a variant of mucous membrane pemphigoid (MMP) or epidermolysis bullosa aquisita (EBA). We present a case of BPP in a woman.

Case report

A 61-year-old woman with a history of hypothyroidism was referred for persistent localized erosions on the scalp for 6 months that did not improve with short courses of doxycycline and systemic steroids. On examination, there was a large erosion on the frontal scalp extending to the vertex with an erythematous rim, surrounded by scarring alopecia (Fig. 1). Patient did not complain of oral or ocular symptoms and no lesions were identified. Punch biopsy at the edge of the sclerotic plaque showed severe perifollicular inflammation and follicular scarring. Direct immunofluorescence of the periulcer skin showed linear deposition of IgG and C3 along the basement membrane zone, with reactants on the epidermal side of salt-split skin. She was treated with a prolonged prednisone taper starting at 40 mg daily, doxycycline 100 mg twice per day, and topical clobetasol 0.05% ointment, resulting in healing of the existing ulcer with no new lesion development. She tolerated reducing doxycycline to 100 mg once daily but noted new erosions when attempting to taper further.

Discussion

Diagnosis of BPP requires histology showing subepidermal bullae formation and direct immunofluorescence showing linear IgG and C3 deposits along the basement membrane zone. BPP demonstrates overlapping clinical, histological, and immunopathologic qualities with MMP and EBA, making its classification a topic of debate. Given the rarity of BPP, there are no studies comparing BPP-like EBA and BPP-like MMP. Further studies are needed to better characterize these conditions.

BPP has a chronic and recurrent course that proves challenging to treat. BPP following physical trauma may be managed with local protection of affected regions.[4] While there are no standardized guidelines for the treatment of BPP, topical[3,4] and/or intralesional[4] corticosteroids are often used. Various immunosuppressive agents have been tried, including prednisone, dapsone, colchicine, cyclosporine, methotrexate (MTX), azathioprine, mycophenolate, cyclophosphamide, rituximab, and intravenous immunoglobulin.[5] According to a recent systematic review, tetracyclines were the most commonly used steroid-sparing agent, followed by dapsone and MTX.[3] Combination therapy using a systemic steroid-sparing agent, topical corticosteroids, and/or antihistamines was most commonly pursued, with a 31% complete remission rate.[3] For mild forms of BPP, topical corticosteroid monotherapy can achieve complete remission.[3] New bullae development was more likely in patients treated with systemic corticosteroids alone.[3] Therefore, potent topical[3,4] and/or intralesional[4] corticosteroids should be considered for mild cases, with added tetracyclines and/or low-dose MTX for more severe cases.[4] In our patient, a regimen of prednisone taper, doxycycline, and topical clobetasol was effective without the need for MTX. Given the chronicity and recurrent course of BPP, management should be individualized with periodic evaluations for treatment efficacy.

Conflicts of interest

None.

Funding

None.

What is known about this subject in regard to women and their families?

- Brunsting–Perry pemphigoid (BPP) is a rare autoimmune condition presenting with tense pruritic bullae that may progress to cicatricial alopecia, which could have a significant impact on the psychosocial health and quality of life of those affected.
- Although the incidence of BPP in women is lower than that of older males, it should remain a differential diagnosis in women with vesiculobullous or cicatricial lesions on the scalp.

What is new from this article as messages for women and their families?

- We report a rare case of a woman with BPP and provide a review of current management approaches.

International Journal of Women's Dermatology (2025) 11:e193

Received: 2 December 2023; Accepted 26 October 2024

Published online 14 January 2025

DOI: 10.1097/JW9.0000000000000193

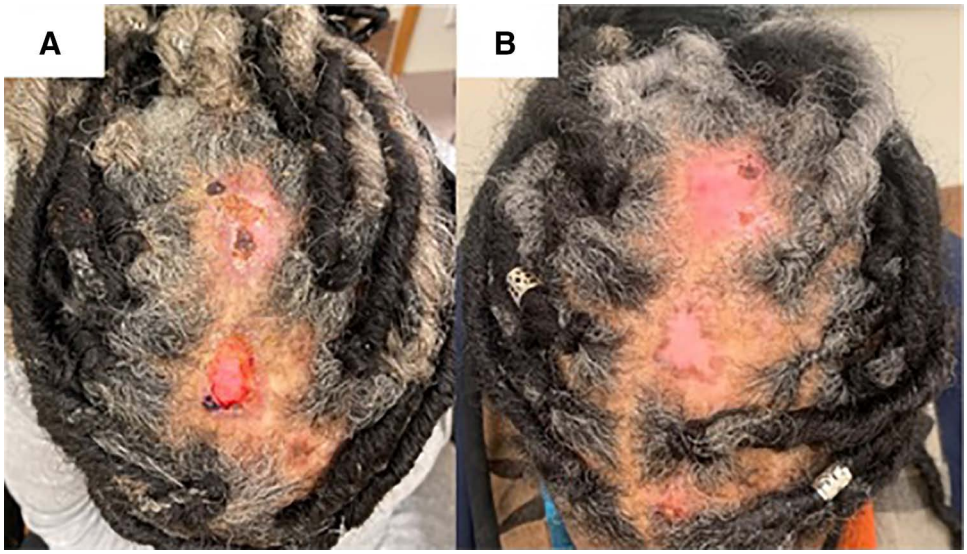

Fig. 1. Erosions on the frontal scalp extending to the vertex, accompanied by erythema and scarring alopecia before (A) and approximately 1 month after (B) treatment with prednisone, doxycycline, and clobetasol ointment.

Study approval

N/A

Author contributions

All authors participated in the ideation and editing of the manuscript and addressing reviewer comments.

Patient consent

Informed, written consent was received from all patients for whom photographs are present in the manuscript.

References

1. DeCastro P, Jorizzo JL, Rajaraman S, Solomon ARJ, Briggaman RA, Raimer SS. Localized vulvar pemphigoid in a child. Pediatr Dermatol 1985;2:302–7.
2. Asfour L, Chong H, Mee J, Groves R, Singh M. Epidermolysis bullosa acquisita (Brunsting-Perry pemphigoid variant) localized to the face and diagnosed with antigen identification using skin deficient in type VII collagen. Am J Dermatopathol 2017;39:e90–6.
3. Aromolo IF, Maronese CA, Moltrasio C, Genovese G, Marzano AV. Brunsting-Perry pemphigoid: a systematic review. Int J Dermatol 2022; 61:1353–8.
4. Chandan N, Juhl ME, Tsoukas MM. Brunsting-Perry pemphigoid: a case with antigen identification. Int J Dermatol 2018;57:e41–3.
5. Kirtschig G, Murrell D, Wojnarowska F, Khumalo N. Interventions for mucous membrane pemphigoid/cicatricial pemphigoid and epidermolysis bullosa acquisita: a systematic literature review. Arch Dermatol 2002;138:380–4.

Maggie Chen, BS[a]
Shealinna Ge, MD[a,*]
Marcia Driscoll, MD, PharmD[a]
[a] *Department of Dermatology, University of Maryland School of Medicine, Baltimore, Maryland*

* Corresponding author.
E-mail address: SGe@som.umaryland.edu (S. Ge).