A large scalp ulceration

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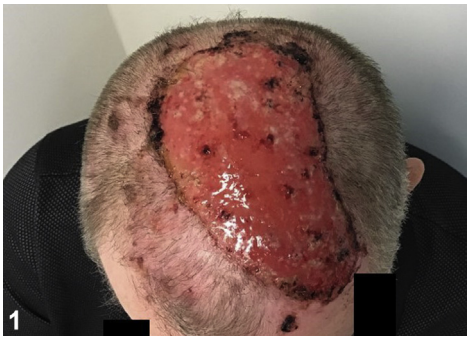
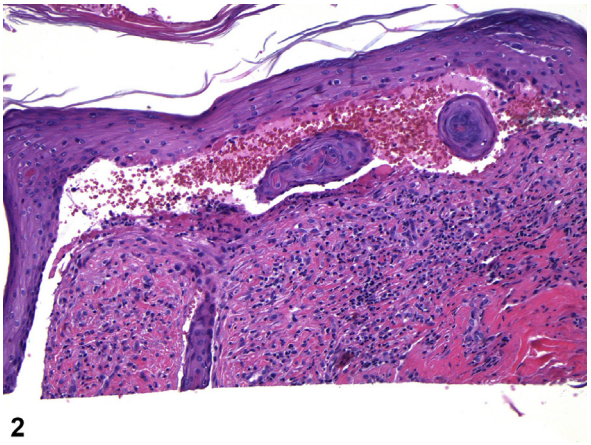
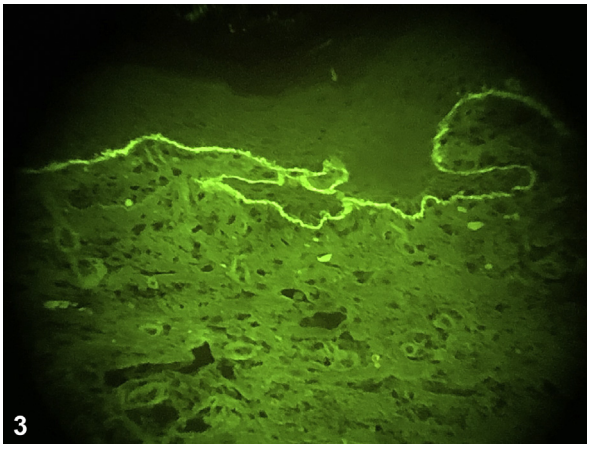
Key words: autoantigen; cicatricial pemphigoid; immunobullous disease; immunofluorescence; ulcer.







A 57-year-old Caucasian man presented with a 3-month history of an expanding irregularly shaped superficial ulcer on the vertex of the scalp with associated hair loss. The patient reported blisters and pustules prior to the onset of the ulcer and alopecia. He denied other skin or oral lesions and specifically denied picking at the scalp. At the time of presentation, physical examination revealed a superficial ulcer, measuring roughly 15 × 8 cm (Fig 1), on the vertex of the scalp. A shave biopsy (Fig 2) was initially performed, followed by 2 punch biopsies; one for hematoxylin and eosin staining, and one for direct immunofluorescence (Fig 3).

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Funding sources: This research was supported (in whole or in part) by HCA Health care and/or an HCA Health care affiliated entity. The views expressed in this publication represent those of the authors and do not necessarily represent the official views of HCA Health care or any of its affiliated entities.
IRB approval status: Not applicable.

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JAAD Case Reports 2021;14:72-4.
2352-5126

https://doi.org/10.1016/j.jdcr.2021.06.005

Question 1: What is the most likely diagnosis?

A. Bullous impetigo

B. Ulcerated basal cell carcinoma

C. Dermatitis artefacta

D. Pemphigus vulgaris

E. Brunsting-Perry cicatricial pemphigoid

Answers:

A. Bullous impetigo — Incorrect. Bullous impetigo would show a subcorneal neutrophil-rich bulla with associated Gram-positive cocci.[1]

B. Ulcerative basal cell carcinoma — Incorrect. Basal cell carcinoma would show a basaloid neoplasm extending off of the epidermis, with peripheral palisading of nuclei and clefting artifact surrounding tumor nodules.[1]

C. Dermatitis artefacta — Incorrect. Dermatitis artefacta typically presents with irregularly shaped erosions/ulcers with jagged borders. While the patient's clinical presentation revealed an asymmetric ulcer, dermatitis artefacta should test negative on direct immunofluorescence.[1]

D. Pemphigus vulgaris — Incorrect. Pemphigus vulgaris would demonstrate a suprabasilar intraepidermal acantholytic bulla with potential follicular involvement. Additionally, it is not typically a scarring process on biopsy.[1]

E. Brunsting-Perry cicatricial pemphigoid — Correct. This patient's presentation of a rapidly progressing ulcer on the head with loss of hair growth in addition to a subepidermal bulla with positive direct immunofluorescence findings suggests localized cicatricial pemphigoid, with the Brunsting-Perry variant being the most likely diagnosis.[1]

Question 2: What separates this entity from other variants of cicatricial pemphigoid?

A. Target antigen

B. Lack of significant mucous membrane involvement

C. Lack of response to immunosuppressive therapy

D. Most cases clinically display one large focal area of cutaneous involvement

E. It has an acute presentation with spontaneous resolution

Answers:

A. Target antigen — Incorrect. Autoantibodies to BP180, type VII collagen, and laminin-332 have been detected. However, the exact antibody for Brunsting-Perry cicatricial pemphigoid is unknown.[2]

B. Lack of mucous membrane involvement — Correct. Brunsting-Perry cicatricial pemphigoid typically does not affect the mucous membrane, though cases have been reported with mild mucous membrane involvement.[3]

C. Lack of response to immunosuppressive therapy — Incorrect. Brunsting-Perry cicatricial pemphigoid can be treated with prednisone and nonsteroidal immunosuppressants such as azathioprine or mycophenolate.[4]

D. Most cases clinically display one large focal area of cutaneous involvement — Incorrect. Most cases of Brunsting-Perry cicatricial pemphigoid present with multiple small areas occurring on the head and neck.[5]

E. It has an acute presentation with spontaneous resolution — Incorrect. Difficulty diagnosing Brunsting-Perry cicatricial pemphigoid often leads to a protracted clinical course and requires long-term treatment.[5]

Question 3: What is observed histologically and on direct immunofluorescence with this entity?

A. Subepidermal bulla with lymphocytes and eosinophils and focal dermal scar; linear basement membrane zone staining with anti-IgG and C3 antibodies

B. Subepidermal neutrophil-rich bulla; granular deposition of IgA in dermal papillae

C. Suprabasilar acantholytic bulla with possible follicular involvement; intercellular IgG or C3 within the epidermis

D. Subepidermal neutrophil-rich bulla; linear IgA staining along the basement membrane of the epidermis

E. Subepidermal bulla with mixed dermal infiltrate containing lymphocytes, eosinophils, and

neutrophils; linear deposition of IgA or IgG along the conjunctival epithelium basement membrane

Answers:

A. Subepidermal bulla with lymphocytes and eosinophils and focal dermal scar; linear basement membrane zone staining with anti-IgG and C3 antibodies — Correct. This histologic and direct immunofluorescent pattern describes Brunsting-Perry cicatricial pemphigoid.[1]

B. Subepidermal neutrophil-rich bulla; granular deposition of IgA in dermal papillae — Incorrect. This histologic and direct immunofluorescent pattern describes dermatitis herpetiformis.[1]

C. Suprabasilar acantholytic bulla with possible follicular involvement; intercellular IgG or C3 within the epidermis — Incorrect. This histologic and direct immunofluorescent pattern describes pemphigus vulgaris.[1]

D. Subepidermal neutrophil-rich bulla; linear IgA staining along the basement membrane of the epidermis — Incorrect. This histologic and direct immunofluorescent pattern describes Linear IgA bullous disease.[1]

E. Subepidermal bulla with mixed dermal infiltrate containing lymphocytes, eosinophils, and neutrophils; linear deposition of IgA or IgG along the conjunctival epithelium basement membrane — Incorrect. This histologic and direct immunofluorescent pattern describes mucous membrane cicatricial pemphigoid.[1]

Special thanks to Dr Maggie Noland, MD from the Department of Dermatology, University of Virginia for reviewing the histopathology and offering clinical correlation based on histology.

Conflicts of interest

None declared.

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