Scarring Head Wound

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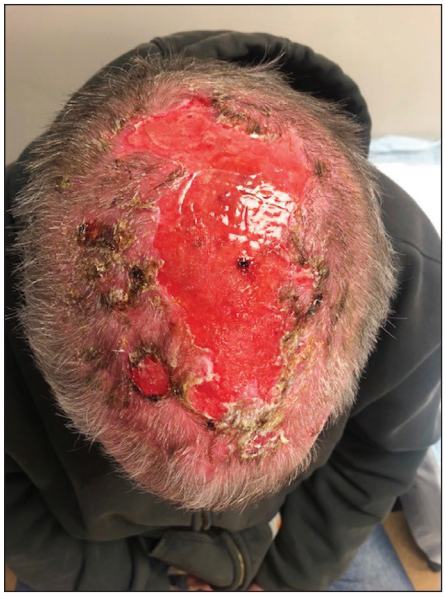

A 60-year-old man presented to a dermatology clinic with a wound on the scalp that had persisted for 11 months. The lesion started as a small erosion that eventually progressed to involve the entire parietal scalp. He had a history of type 2 diabetes mellitus, hypertension, and Graves disease. Physical examination demonstrated a large scar over the vertex scalp with central erosion, overlying crust, peripheral scalp atrophy, hypopigmentation at the periphery, and exaggerated superficial vasculature. Some oral erosions also were observed. A review of systems was negative for any constitutional symptoms. A month prior, the patient had been started on dapsone 50 mg with a prednisone taper by an outside dermatologist and noticed some improvement.

WHAT'S YOUR **DIAGNOSIS?**

a. anti–laminin 332 pemphigoid
b. anti-p200 pemphigoid
c. Brunsting-Perry cicatricial pemphigoid
d. bullous pemphigoid
e. epidermolysis bullosa aquisita

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The authors report no conflict of interest.
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THE **DIAGNOSIS**
Brunsting-Perry Cicatricial Pemphigoid

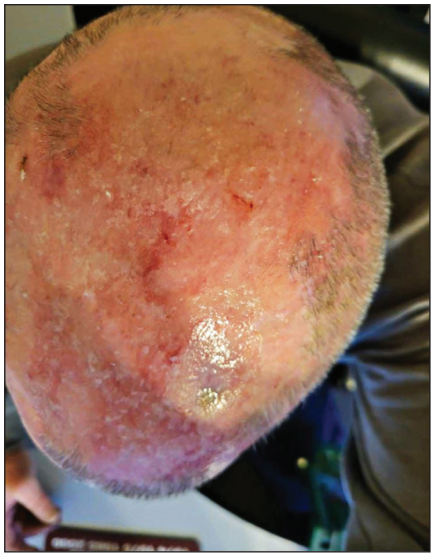
Physical examination and histopathology are paramount in diagnosing Brunsting-Perry cicatricial pemphigoid (BPCP). In our patient, histopathology showed subepidermal blistering with a mixed superficial dermal inflammatory cell infiltrate. Direct immunofluorescence was positive for linear IgG and C3 antibodies along the basement membrane. The scarring erosions on the scalp combined with the autoantibody findings on direct immunofluorescence were consistent with BPCP. He was started on dapsone 100 mg daily and demonstrated complete resolution of symptoms after 10 months, with the exception of persistent scarring hair loss (Figure).

Brunsting-Perry cicatricial pemphigoid is a rare dermatologic condition. It was first defined in 1957 when Brunsting and Perry[1] examined 7 patients with cicatricial pemphigoid that predominantly affected the head and neck region, with occasional mucous membrane involvement but no mucosal scarring. Characteristically, BPCP manifests as scarring herpetiform plaques with varied blisters, erosions, crusts, and scarring.[1] It primarily affects middle-aged men.[2]

Historically, BPCP has been considered a variant of cicatricial pemphigoid (now known as mucous membrane pemphigoid), bullous pemphigoid, or epidermolysis bullosa acquisita.[3] The antigen target has not been established clearly; however, autoantibodies against laminin 332, collagen VII, and BP180 and BP230 have been proposed.[2,4,5] Jacoby et al[6] described BPCP on a spectrum with bullous pemphigoid and cicatricial pemphigoid, with primarily circulating autoantibodies on one end and tissue-fixed autoantibodies on the other.

The differential for BPCP also includes anti-p200 pemphigoid and anti–laminin 332 pemphigoid. Anti-p200 pemphigoid also is known as bullous pemphigoid with antibodies against the 200-kDa protein.[7] It may clinically manifest similar to bullous pemphigoid and other subepidermal autoimmune blistering diseases; thus, immunopathologic differentiation can be helpful. Anti–laminin 332 pemphigoid (also known as anti–laminin gamma-1 pemphigoid) is characterized by autoantibodies targeting the laminin 332 protein in the basement membrane zone, resulting in blistering and erosions.[8] Similar to BPCP and epidermolysis bullosa aquisita, anti–laminin 332 pemphigoid may affect cephalic regions and mucous membrane surfaces, resulting in scarring and cicatricial changes. Anti–laminin 332 pemphigoid also has been associated with internal malignancy.[8] The use of the salt-split skin technique can be utilized to differentiate these entities based on their autoantibody-binding patterns in relation to the lamina densa.

Treatment options for mild BPCP include potent topical or intralesional steroids and dapsone, while more severe cases may require systemic therapy with rituximab, azathioprine, mycophenolate mofetil, or cyclophosphamide.[4]



The patient demonstrated complete resolution of Brunsting-Perry cicatricial pemphigoid symptoms on the scalp following treatment with dapsone; scarring hair loss persisted.

This case highlights the importance of histopathologic examination of skin lesions with an unusual history or clinical presentation. Dermatologists should consider BPCP when presented with erosions, ulcerations, or blisters of the head and neck in middle-aged male patients.

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