Stump Pemphigoid Demonstrating Circulating Anti–BP180 and BP230 Antibodies

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PRACTICE **POINTS**

- Bullous pemphigoid (BP) can mimic friction blisters and should be considered in amputees who present with vesicles and bullae on their amputation stump.
- Circulating anti–basement membrane antibodies BP230 and BP180 IgG may aid in diagnosis when skin biopsy results are equivocal and also may be helpful in gauging treatment response.

To the Editor:

Bullous pemphigoid (BP) is a rare complication of lower limb amputation. Termed *stump pemphigoid,* it previously was described as a late complication arising on the stumps of leg amputees and tends to remain localized. We describe a case of stump pemphigoid presenting with an urticarial prodromal phase without generalized progression, confirmed by serum assay for circulating anti–basement membrane antibodies.

A 62-year-old man with a history of a right above-knee amputation initially presented with erythema as well as coalescing erosions and ulcers with fluid-filled vesicles and bullae on the amputation stump (Figure 1). The amputation was performed 15 years prior after a motorcycle accident. A skin biopsy of a vesicle on the amputation stump revealed subepidermal and focal intraepidermal clefting with hemorrhage and rare inflammatory cells composed of neutrophils and eosinophils (Figure 2). A tissue direct immunofluorescence test demonstrated linear C3 and IgG deposition along the dermoepidermal junction. Serum enzyme-linked immunosorbent assay (ELISA) demonstrated an anti-BP180 IgG of 50.90 U/mL and anti-BP230 IgG of 129.40 U/mL (reference range, <9.00 U/mL [for both]).

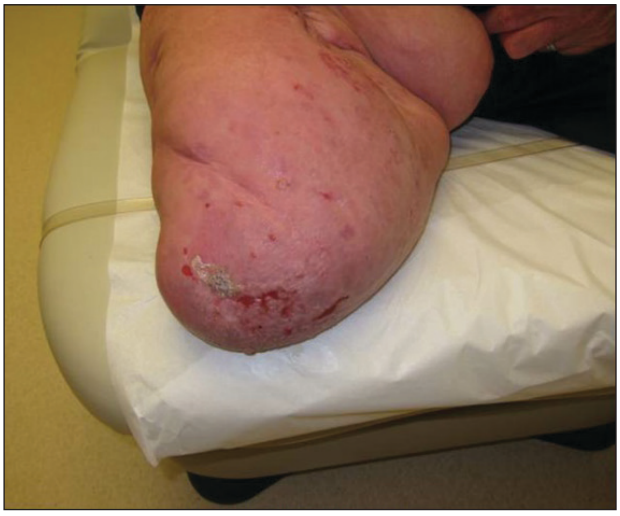

FIGURE 1. Stump pemphigoid. Erosions and bullae on an amputation stump.

Topical clobetasol led to only modest improvement of blistering on the stump. Minor frictional trauma related to his leg prosthesis continued to trigger new vesicles and bullae on the stump. Oral prednisone 0.5 mg/kg daily was administered and tapered slowly over the course of 6 months. He also received oral niacinamide and doxycycline. He was completely clear after 3 weeks of

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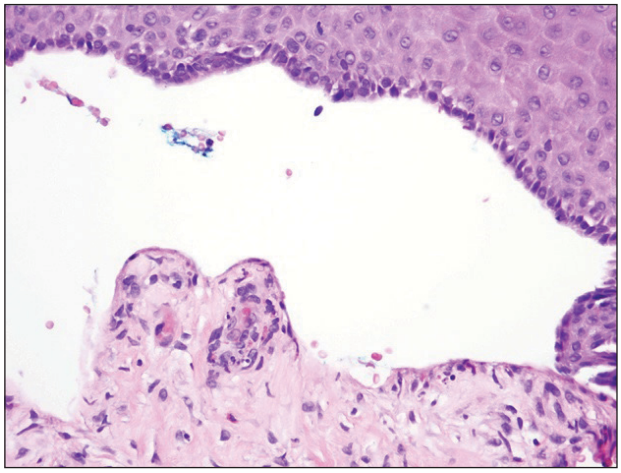

FIGURE 2. Subepidermal cleft with red blood cells and sparse lymphocytic and eosinophilic infiltrate (H&E, original magnification ×400).

initiating treatment and remained clear while prednisone was slowly tapered. One month after stopping prednisone he had recurrence of blisters on the stump only after he resumed wearing his prosthesis. Mycophenolate mofetil was started at a dosage of 1 g twice daily while he refrained from wearing the prosthesis. After 3 months he was able to wear the prosthesis without developing blisters. Two years after the initial presentation, repeat serum ELISA demonstrated normalization of the anti-BP180 IgG and anti-BP230 IgG titers. Thirty months after the initial presentation, mycophenolate mofetil was tapered and discontinued. The patient remained blister free and continued to wear his leg prosthesis without further blistering.

Amputees experience a high rate of skin complications on their stump,[1] including friction blisters, shear injury, contact dermatitis, infections, and autoimmune blistering disorders (ie, BP, epidermolysis bullosa acquisita). The etiology of stump pemphigoid is not entirely understood but could be related to exposure of structural components of the hemidesmosome (eg, BP230, BP180), leading to autoantibody production as a consequence of either the underlying limb injury or from recurrent trauma related to limb prosthetics.[2]

Two previously reported cases of stump pemphigoid demonstrated a positive direct immunofluorescence antibody test.[3,4] Another case demonstrated the presence of circulating IgG antibodies on indirect immunofluorescence to salt-split skin.[5] We report a case of stump pemphigoid confirmed by presence of circulating anti–basement membrane antibodies on ELISA, supporting its use in the diagnostic workup and monitoring treatment response.

REFERENCES

1. Colgecen E, Korkmaz M, Ozyurt K, et al. A clinical evaluation of skin disorders of lower limb amputation sites. *Int J Dermatol*. 2016;55:468-472.
2. Lo Schiavo A, Ruocco E, Brancaccio G, et al. Bullous pemphigoid: etiology, pathogenesis, and inducing factors: facts and controversies. *Clin Dermatol*. 2013;31:391-399.
3. Reilly GD, Boulton AJ, Harrington CI. Stump pemphigoid: a new complication of the amputee. *Br Med J (Clin Res Ed)*. 1983;287:875-876.
4. de Jong MC, Kardaun SH, Tupker RA, et al. Immunomapping in localized bullous pemphigoid. *Hautarzt*. 1989;40:226-230.
5. Brodell RT, Korman NJ. Stump pemphigoid. *Cutis*. 1996;57:245-246.