Bullous pemphigoid superimposed on psoriasis

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History

- A 62-year-old white female presented with erythroderma and pruritus shortly after completing a course of steroids from prior admission for COPD exacerbation.
- PMH: psoriasis previously well controlled on topical steroids, COPD with frequent exacerbations, HFpEF, chronic idiopathic leukocytosis, osteoarthritis, and chronic tobacco use (40 pack-years)
- Inpatient biopsy consistent with psoriasiform dermatitis, favored eruptive erythrodermic psoriasis. Started on topical steroid wet wraps and was discharged on topical steroids.
- At outpatient follow-up, patient worsening with new painful blisters despite consistent topical steroid use.
  - Biopsy repeated for H&E and DIF. Type VII antibody ELISA and IIF salt split skin were ordered.

Examination

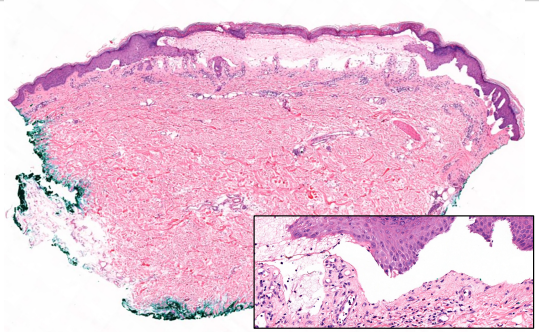
Initial exam:

- Large erythematous plaques and papules with scale scattered throughout scalp, ears, neck, trunk, extremities, palms, and soles, ~90% BSA **(Fig 1A, 1B)**.
- Tense bullae and erosions with serous and hemorrhagic crusting on extremities and lower abdomen **(Fig 1B, 1C)**.

Outpatient follow up:

- Erythematous macules coalescing into patches and mildly scaly plaques, sparing skin folds and face, ~80% BSA **(Fig 2A-E)**.
- Tense serous and hemorrhagic bullae both intact and ruptured on lower extremities, infrapannus, and wrists **(Fig 2B-2D)**.
- Erosions with hemorrhagic crust on dorsal hands and forearms **(Figure 2D)**. Palms and soles had hyperkeratotic scale **(Figure 2E)**.

Histopathology



H+E low power: On low power of this punch specimen, there are areas of detached epidermis with subepidermal clefting.
H+E high power: On high power, there is festooning of the dermal papillae with collections of eosinophils in the papillary dermis.

DIF (not shown here): Linear staining of IgG and C3 deposits along DEJ

Clinical Photography

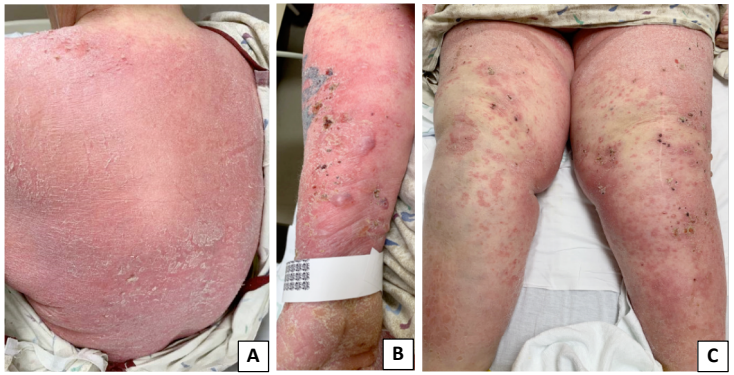


Figure 1A-C. Inpatient encounter: Diffuse erythematous plaques with scale on a background of erythema present on the trunk (**A**) and extremities **(B, C).** Tense bullae and crusted erosions are shown on the and arms **(B)**.

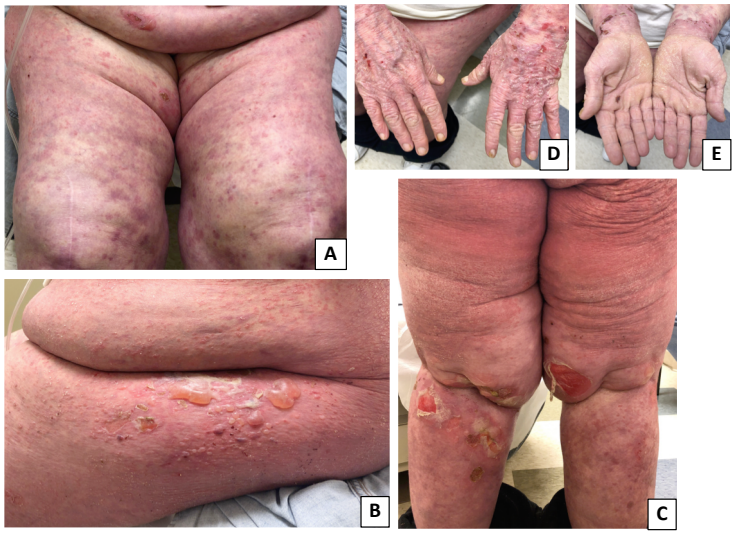


Figure 2A-E. At outpatient follow up: Blotchy erythematous patches admixed with thin, mildly scaly plaques on 80% BSA, sparing skin folds and face. Tense and ruptured bullae along with crusted erosions were visible on the extremities and infrapannus **(B-D).** Palms had hyperkeratotic scale **(E).**

Course and Therapy

- The patient was started on oral methotrexate 25mg weekly and triamcinolone 0.1% ointment BID
- Three weeks later, course was complicated by cellulitis of weeping bullae on the bilateral lower extremities and was admitted to the hospital for IV antibiotics
- Dermatology was consulted and patient was started on prednisone 40mg daily with 10mg weekly taper.
- Discharged with oral clindamycin, topical triamcinolone 0.1% twice daily, and methotrexate 50mg weekly

Discussion

- Psoriasis has been associated with multiple autoimmune bullous diseases, the most common being bullous pemphigoid.[6]
- Bullous pemphigoid (BP) is a sub-epidermal blistering disease caused by autoantibodies against hemidesmosome proteins BP 180 and BP 230. It most commonly occurs in those older than 70 years of age.[1]
- A prodromal phase may precede the development of tense bullae in BP by weeks to months. This can present as pruritus alone or urticarial-like or scaly eczematous papules and plaques.[1]
- Patients with BP and comorbid psoriasis are younger, have a more erosive phenotype, and lower levels of pathogenic autoantibodies.[2, 3]
- Studies suggest a 1.5x to 3.6x higher risk of developing BP in psoriasis patients.[4, 5]
- Over 1/3 of BP cases were diagnosed in the first year after psoriasis diagnosis in one study.[5] In others, the association was bidirectional and BP was diagnosed after psoriasis on average 3 to 14 years.[2, 6]
- BP and psoriasis was found to be more prevalent in males.[4]
- Patients with both BP and psoriasis have a higher prevalence of smoking and hypertension compared to patients with BP alone. They were also treated more frequently with prolonged systemic and topical corticosteroids.[2]
- The mechanism for this association is not well understood. There are several theories:
  - Chronic inflammation in the DEJ can trigger exposure to antigens to autoreactive T cells.[7]
  - BP antigens are unmasked in psoriasis through enzymatic degradation of the BMZ, making them more accessible by circulating antibodies.[2, 4]
  - Neutrophil chemo-attractants from keratinocytes in both psoriasis and BP lead to metalloprotease degradation of DEJ matrix proteins, leading to exposure of antigenic epitopes.[7]
  - UVB phototherapy and PUVA has also been implicated in altering the antigenicity of the basement membrane, which may trigger a bullous-like response in psoriasis patients on these therapies.[6]
- Psoriasis-associated BP can be treated with immunosuppressives, such as methotrexate, cyclosporine, or mycophenolate mofetil. [8-10]

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Disclosures

Ms. Liu and Drs. Ko, Ceresnie, and McHargue have no financial relationships to disclose.