A case of bullous pemphigoid following pemphigus foliaceus

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Disclosures

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Background

**Bullous Pemphigoid**
- Tense blisters
- Negative Nikolsky’s sign
- BP180 and BP230
- Linear IgG and C3 at dermal-epidermal junction

**Pemphigus Foliaceus**
- Small flaccid blisters and crusted erosions
- No mucosal involvement
- + Nikolsky’s sign
- DSG1
- Intercellular IgG +/- C3

Adapted from Kershenovich et al. Autoimmunity reviews 2014:13;477-481
Visit 1

- 71-year-old woman
- 12-year history of biopsy and DIF confirmed pemphigus foliaceus (PF)
  - Manifested as erythematous plaques with crusting and scale of the trunk and scalp
  - Managed on chronic low dose mycophenolate mofetil

Presented to outpatient dermatology clinic with two weeks of new lesions on thighs and forearms

- Suspected PF flare
  - Increased mycophenolate mofetil to 500 mg daily
  - Initiated clobetasol 0.05% ointment
Case Report:

Visit 2: 6 days later

- Intensely pruritic lesions with two morphologies:
  - 1-2 cm bright red annular and arcuate plaques with peripheral edema without overlying scale or crust on lower extremities
  - Mild erosions with significant overlying scale on back

- Loratidine initiated for idiopathic urticaria
Visit 3: 1 month later

Diffuse blistering of mixed morphology noted with multiple flaccid blisters, associated erosions and numerous intact, tense, Nikolsky negative blisters
Visit 3 (continued): Right thigh perilesional punch biopsy

- Subepidermal bullous dermatosis with numerous eosinophils
- Direct immunofluorescence: linear deposition of IgG(1+) and C3(2+) along the dermal epidermal junction
- ELISA: antibodies to DSG1 and BP180, but not to DSG3 or BP230

Dx: simultaneous pemphigus foliaceus and bullous pemphigoid
Case Report:

- **Therapy**
  - Mycophenolate mofetil 1500 mg twice daily
  - Prednisone 20mg daily
  - Clobetasol 0.05% ointment to new lesions under occlusion daily

- **18 months after bullous pemphigoid diagnosis**
  - Patient was doing well with no skin lesions
  - Mycophenolate mofetil taper of 500mg each week for three weeks, followed by mycophenolate mofetil discontinuation
Discussion:

Coexistence of two autoimmune blistering diseases

- First described by Chorzelski T, et al. in 1974
  - 3 patients with pemphigus vulgaris and bullous pemphigoid

- Subsequent reports:
  - Timespan for development of concomitant autoimmune blistering diseases has ranged from coexistent at time of presentation to years prior to development of second process
    - Pemphigus vulgaris/bullous pemphigoid
    - Pemphigus foliaceus/bullous pemphigoid
    - Both men and women

Discussion

Epitope spreading (ES):

- An epitope is an antigenic determinant to which a specific antibody binds.
- ES refers to the development of an immune response to epitopes distinct from and non-cross-reactive with the disease causing epitope.
- In autoimmunity ES refers to the development of immune responses against endogenous epitopes secondary to the release of self-Ag during a chronic auto immune response.
  - Implicated in type 1 diabetes, rheumatoid arthritis, multiple sclerosis, systemic lupus erythematosus, and others.
  - Coexistent bullous pemphigoid and pemphigus represents intermolecular progression of antibodies from hemidesmosome to desmoglein protein antigens.


