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Bullous Pemphigoid with Excoriation Disorder in a 59 Year Old Woman

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Background

- Bullous pemphigoid is the most common autoimmune blistering disease.
- Classic symptoms include blisters overlying urticarial plaques on the torso and extremities.
- Etiology includes an abnormal T-cell response that triggers production of IgG and IgE autoantibodies which attack the hemidesmosomes of the basement membrane (1).
- The condition can result in intense pruritus that begins during the prodromal period (2).
- Excoriation disorder is related to obsessive-compulsive disorder (OCD) and is characterized by recurrent skin picking that results in lesions, repeated attempts to stop or decrease the picking, and resultant mental distress or impairment in functioning (3,4).

Patient Presentation

- 59-year-old Caucasian female presented with diffuse rash of all four extremities, trunk, abdomen, and back but sparing the upper feet and face. The current rash had been present for the past four months but the patient had a recurring rash “on and off for the past year”.
- Unable to associate symptoms with change in medication or illness.
- Rash associated with diffuse itching and burning pain.
- Uncontrollable urge to pick at skin in response to feelings of anxiety.
- Denied fever, chills, weakness, or recent insect bites.
- Medical History: type 2 diabetes, polycystic ovarian syndrome, eczema, and bipolar disorder type II, anxiety, depression
- Family History: no history of autoimmune disease
- Medications: duloxetine 60 mg daily (started 1 month ago), triamcinolone 0.5% cream BID as needed for eczema, atorvastatin 40 mg, metformin 500 mg, hydroxyzine HCl 25 mg BID, metoprolol succinate 25 mg, oxybutynin 15 mg, aspirin 81 mg

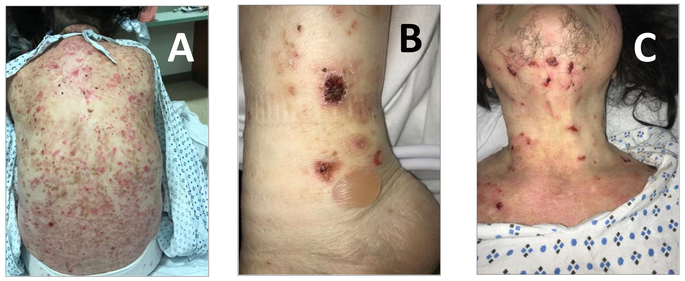

Figure A: Diffuse rash of posterior torso

Figure B: A 2 cm closed bullous lesion containing clear serosanguinous fluid seen on the right medial lower extremity

Figure C: Diffuse scabs on the anterior neck region

- Physical Exam:
  - Severe rash of the lower face, neck, chest, trunk, and extremities with an erythematous base accompanied by moderate diffuse excoriation and severe dryness (Figures A-C).
  - No lesions of mucosa of the oropharynx or nares.
  - Clear bullae filled with serosanguinous fluid and scabs in stages of healing (Figure B).
  - Bullae did not slough off when pressure was applied.

Investigation

- Two punch biopsies were taken, one from each thigh.
- Histology showed subepidermal bullous dermatosis with mixed inflammatory infiltrate of predominantly eosinophils (Figures D and E).
- Direct immunofluorescence of samples demonstrated linear deposition of IgG and C3 along the basement membrane (Figure F).
- Leukemia panel of the peripheral blood indicated no diagnostic immunophenotypic abnormalities detected by flow cytometry.
- Findings consistent with bullous pemphigoid.

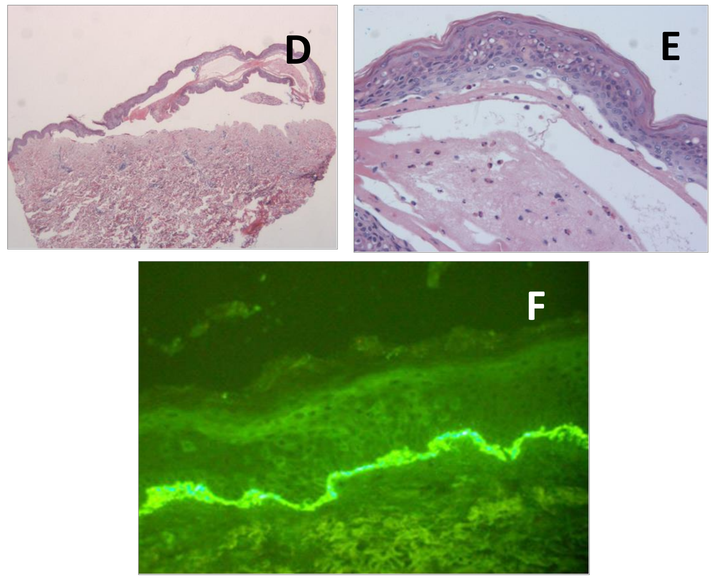

Figure D: Subepidermal bullous dermatosis with mixed inflammatory infiltrate in the blister cavity (10x magnification).

Figure E: The mixed inflammatory infiltrate in the subepidermal blister cavity consists predominantly of eosinophils (40x magnification).

Figure F: Direct immunofluorescence shows linear deposition of IgG (primarily IgG4) and C3 along the basement membrane.

Treatment and Outcome

- Single dose of methylprednisolone 125 mg IV given in emergency room
- Four day course of IV vancomycin 15 mg/kg Q12 hours
- Cross tapered from duloxetine to fluoxetine 40 mg daily to treat excoriation disorder
- Discharged on oral prednisone taper starting at 60 mg and decreasing by 10 mg every five days until complete
- Referred for outpatient psychotherapy
- Significant improvement in skin symptoms on prednisone taper before lost to follow up 3 months later

Discussion

- Treatment for bullous pemphigoid with Level 1A evidence typically includes systemic steroids for severe cases and topical clobetasol for mild to moderate cases (1).
- Treatment for excoriation disorder involves psychotherapy, such as cognitive behavior therapy or habit reversal therapy. Pharmacotherapy options include SSRIs and lamotrigine (5).
- Excoriation disorder is related to OCD, for which fluoxetine is known to be especially efficacious (6).
- There is emerging yet still controversial evidence suggesting links between bullous pemphigoid and neuropsychiatric conditions. Some studies have found increased incidence of bullous pemphigoid in patients with psychiatric histories including those with major depressive disorder and bipolar depression (7,8).
- Diagnosis was made with direct immunofluorescence that showed linear deposition of IgG and C3 along the basement membrane (Figure F).
- Alternative diagnostic methods include:
  - Serological detection of autoantibodies by indirect immunofluorescence on human salt-split skin (9)
  - ELISA using recombinant fragments of BP180 and BP230 (9)
- This particular case is notable for its concurrent severe dermatological and psychiatric components.

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