Unusual Presentation of Bullous Pemphigoid: A Clinical Challenge

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Background

Bullous Pemphigoid (BP) is the most common autoimmune subepidermal blistering disease (~80% of cases) that typically affects older adults (65-75 years). This condition is often characterized by autoantibodies to hemidesmosomal proteins (BP180, BP230), complement activation, and dermal-epidermal separation.

- Classic presentations include tense bullae, urticarial plaques, and severe pruritus, with atypical variants presenting as pruritus without bullae or with mucosal involvement (10–20%).
- Diagnosis requires correlation of clinical, histologic, and immunofluorescence findings.

This case is notable because BP with a “linear full-house” DIF pattern (IgA, IgG, IgM, C3) is highly unusual.

Case Presentation

Initial Presentation
- 66-year-old man with history of rheumatoid arthritis, lymphoma (remission), and cutaneous SCC
- Presented with pruritic, eczematous plaques on thigh → diagnosed as nummular dermatitis, treated with clobetasol

Clinical Course
- Six months later, developed painful, pruritic blisters at trauma sites
- Lesions followed relapsing-remitting course over 3 months
- No oral lesions on exam, but photos showed intermittent oral involvement triggered by coarse foods

Biopsy Findings
- Subepidermal bullous dermatosis with early dermal scarring
- Subtle vacuolar changes at dermoepidermal junction
- No fungal organisms or excess dermal mucin

Direct Immunofluorescence (DIF)
- Linear deposition of IgA, IgG, IgM, and C3 (“full-house” pattern)

Serology & IIF
- BMZ IgG positive on monkey esophagus substrate
- BP180 elevated; BP230, Type VII collagen, and DSG1/3 negative
- IgG localized to epidermal side of salt-split skin; IgA absent

Other Labs
- CBC, CMP, IgE, UA, hepatitis panel, QuantiFERON gold all unremarkable

Final Diagnosis
- Bullous pemphigoid with rare “full-house” DIF pattern

Clinical Images

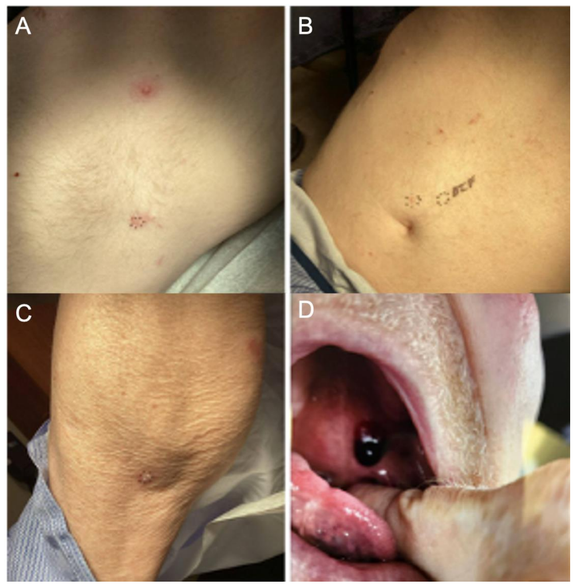

Fig. 1 Clinical findings (A) Left flank (B) Periumbilical Abdomen (C) Left calf (D) Oral lesion

Histologic Findings

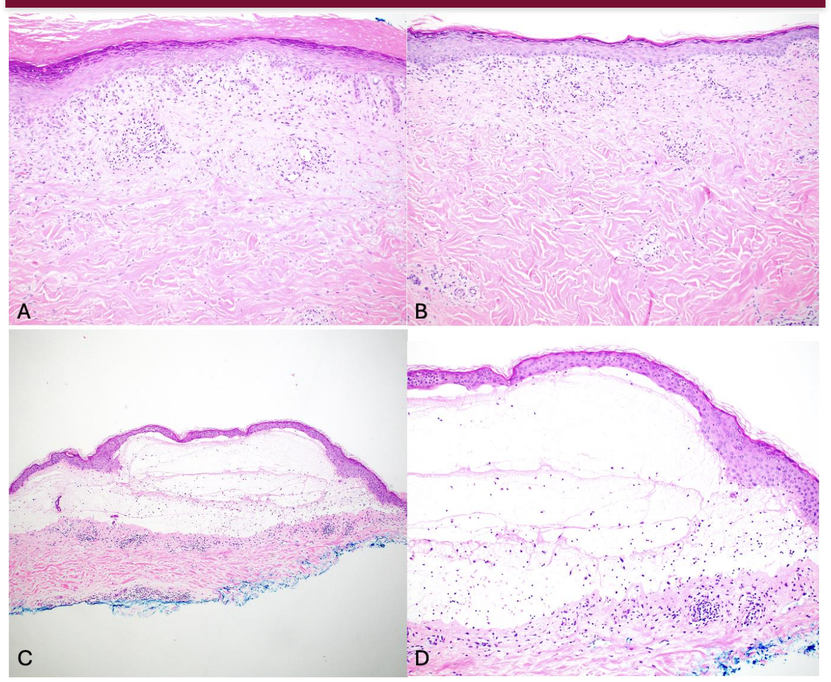

Fig. 2 H&E (A) Left Calf Shave Biopsy (10x) (B) Left Flank Shave Biopsy (10x) (C) Periumbilical Bulla (4x) (D) Periumbilical Bulla (10X)

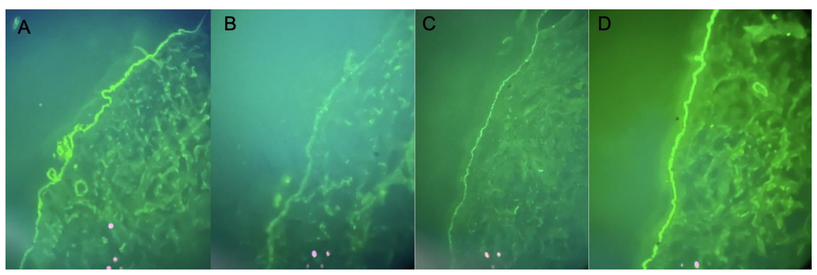

Fig. 3 Direct Immunofluorescence periumbilical skin: (A) IgG (B) IgM (C) IgA (D) C3

Discussion

Typical Presentation
- Bullous pemphigoid (BP) most often presents with tense bullae and intense generalized pruritus.
- Atypical cases may show only pruritus without bullae or oral lesions (10–20% of patients) .

Pathogenesis
- Autoantibodies against hemidesmosomal proteins (BP180, BP230) activate complement and damage the dermoepidermal junction.

Histology & DIF Patterns
- BP can mimic urticaria or epidermolysis bullosa acquisita (EBA).
- DIF usually shows linear C3 ± IgG at the basement membrane zone; IgA/IgM are less common.
- The “full-house” linear deposition of IgA, IgG, IgM, and C3 in this case is extremely rare.

Confirming the Diagnosis
- Despite unusual DIF, salt-split skin IIF showed IgG on the epidermal side, and serology confirmed BP180 antibody positivity.
- Type VII collagen antibody was negative, ruling out EBA.
- Literature supports that IgA/IgM positivity, while rare, can occur in BP.

Significance
- To our knowledge, this is the first reported case of BP with a linear full-house DIF pattern.
- Highlights the variability of BP immunopathology and the importance of correlating clinical, histologic, and serologic data.

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