DEPARTMENT OF
PATHOLOGY AND
LABORATORY MEDICINE
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Infantile Bullous Pemphigoid in a 3-Month-Old Female: A Rare and Important Diagnostic Consideration

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INTRODUCTION

Bullous pemphigoid (BP) is a rare autoimmune blistering disorder, infrequently seen in children and exceptionally rare in infants. Due to significant clinical overlap with infectious, genetic, and other autoimmune conditions, definitive diagnosis in infants relies heavily on histopathologic and immunopathologic evaluation.We present a case of infantile BP to highlight its distinctive clinicopathologic features and diagnostic challenges.

CASE PRESENTATION

Patient: 3-month-old female.
History: 5-day history of erythematous plaques on hands/feet → progression to tense bullae → spread to forearms and trunk.
Systemic status: Afebrile, otherwise well.
Investigations:

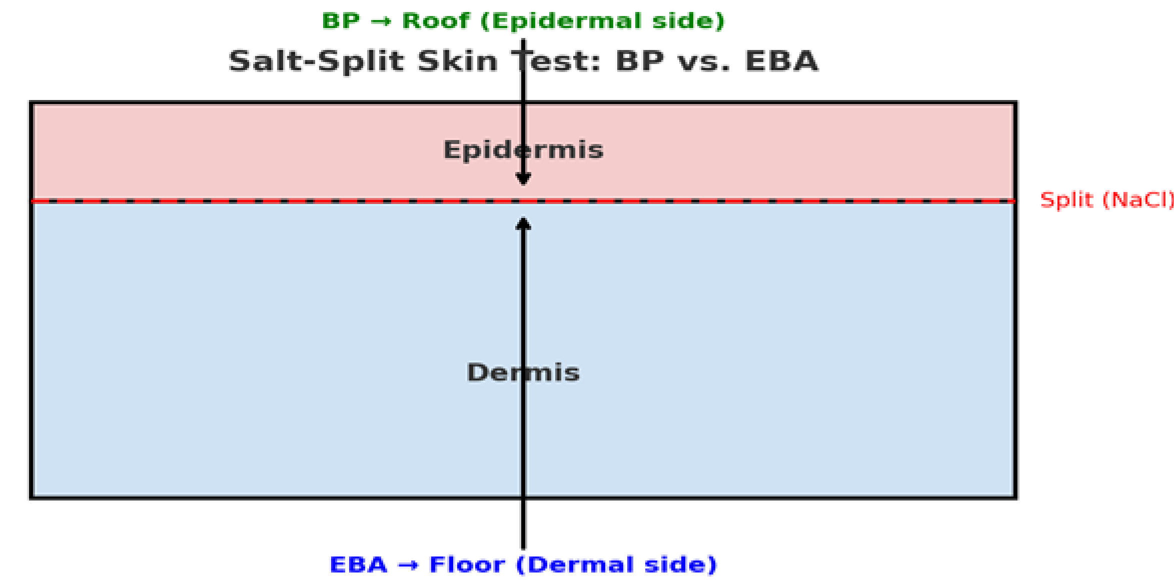
- Bacterial cultures and viral PCR: negative.
- **Histopathology (punch biopsy):** spongiotic dermatitis with eosinophilic exocytosis, papillary dermal edema, and perivascular lymphohistiocytic infiltrate with eosinophils.
- **Direct immunofluorescence:** linear IgG and C3 along basement membrane zone → diagnostic of bullous pemphigoid.

Family/Maternal history: no autoimmune or blistering disorders; maternal history unremarkable.

Differential Diagnosis

Clinical differential:
- Bullous pemphigoid (BP)
- Atypical coxsackievirus infection
- Epidermolysis bullosa (EB)
- Herpes simplex virus (HSV)
- Varicella zoster virus (VZV)
- Bullous impetigo

Histopathologic differential:
- **Epidermolysis bullosa acquisita (EBA)** may mimic BP.
- **Direct immunofluorescence (DIF):**
  - BP → linear IgG and **strong C3** at basement membrane; n-serrated pattern.
  - EBA → linear IgG, **u-serrated pattern**.
- **Salt-split skin analysis:**
  - BP → IgG/C3 localizes to **roof (epidermal surface)**.
  - EBA → IgG/C3 localizes to **floor (dermal side)**.

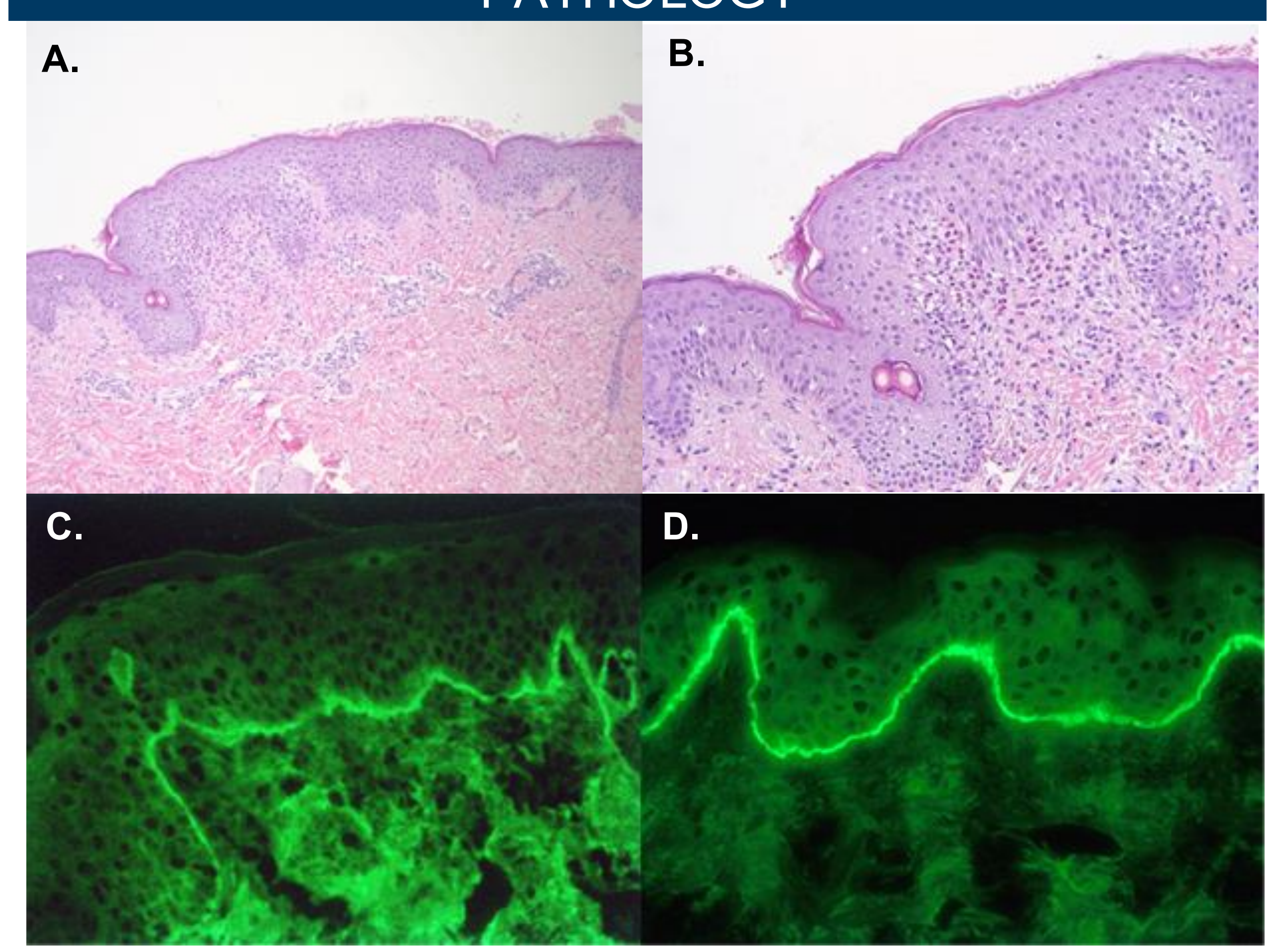


- The **red dashed line** marks the NaCl-induced split at the lamina lucida.
- **Bullous pemphigoid (BP):** IgG/C3 deposits on the **roof (epidermal side)**.
- **Epidermolysis bullosa acquisita (EBA):** IgG/C3 deposits on the **floor (dermal side)**.

Clinical Pictures

A. Tense bullae on the left foot. B. Healing, previously ruptured blisters on the right leg. B. Mixed new and resolving bullae on the left hand.

PATHOLOGY



E.

Antibody	Result	Reference Range	Interpretation
IgG BP180	125 U/mL	< 9 U/mL = negative	Positive
IgG BP230	0 U/mL	< 9 U/mL = negative	Negative

A. Spongiotic dermatitis and perivascular lymphohistiocytic infiltrate inflammation. B. papillary dermal edema with eosinophilic exocytosis. C. IgG linear dermoepidermal junction DIF. D. C3 linear dermoepidermal junction DIF. E. Results of ELISA assay.

Management & Outcomes

- **Treatment:** Prednisolone oral taper; 0.1% triamcinolone ointment.
- **Clinical Course:** No new blisters since treatment initiation. At follow-up, hands and feet were clear with mild erythematous macules at prior blister sites. Plan: dermatology follow-up after steroid taper; escalate to higher potency topical steroid or dapsone if recurrence occurs.

SIGNIFICANCE AND CONCLUSIONS

Rarity:
- Infantile bullous pemphigoid (BP) is extremely uncommon, with only a limited number of reported cases in the literature.

Clinical features:
- Tense bullae, often with acral involvement. Mucosal involvement is less frequent than in adults.

Diagnostic challenges:
- Significant overlap with infectious (impetigo), genetic (epidermolysis bullosa), and other autoimmune blistering disorders (linear IgA, epidermolysis bullosa acquisita). Histopathology and immunopathology (DIF, BP180/230 testing) are essential for confirmation.

Management:
- Most infants respond well to systemic corticosteroids.
- Steroid-sparing agents (e.g., dapsone, IVIG) may be needed in refractory or relapsing cases.

Prognosis:
- Typically favorable in infants compared to adult BP.
- Relapse is possible but long-term disease is uncommon.

Key teaching points:
- Always consider BP in the differential for infantile blistering eruptions.
- Early recognition prevents misdiagnosis and unnecessary treatments.
- Clinicopathologic correlation is the cornerstone of diagnosis.

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