

Genital Bullous Pemphigoid – A Rare Clinical Equivocacy

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Abstract: Pemphigoid is a sub group of auto immune bullous skin disorders. This includes the most common entity Bullous pemphigoid (BP). Genital bullous pemphigoid (GBP) is a localized variant of BP occurring most frequently in paediatric and female patients and rarely in males. We report one case of GBP in a male. A 65-year-old man presented with fluid filled lesion over the body predominantly over the genitals since 4 months. Vesicles ruptured within 1 day, healed with scarring and was associated with intense itching. There was remission and relapse of the lesions every 2 weeks since then without involving any other mucosal areas. Although GBP in males is rare, the clinical presentation should induce suspicion as quick and accurate diagnosis is imperative to implement effective therapy and achieve remission.

Key Words: Pemphigoid, BP, GBP

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I. Introduction

Pemphigoid is a sub group of immunobullous skin disorders. This includes the most common entity Bullous pemphigoid (BP). Genital bullous pemphigoid (GBP) is a localized variant of BP occurring most frequently in paediatric and female patients and rarely in males. We report one case of GBP in a male.¹

Bullous pemphigoid (BP) is a rare, autoimmune, chronic skin disorder characterized by blistering, urticarial lesions (hives) and itching. Less commonly these blisters can involve the mucous membranes including the eyes, oral mucosa, and genital mucosa.² It typically presents in older adults as a generalized intensely itchy blistering skin condition.³

The first symptom of BP is usually redness and itching of the skin. Within weeks to months, thin-walled, tense blisters with clear fluid centers (bullae) appear on the arms and legs (flexor surfaces), in the armpits (axillae), on the abdomen, and/or in the skinfolds of the groin. Mucous membranes may also be involved but are less commonly seen than skin blisters.^{4,5}

The blisters are usually tense (tight), and contain clear or blood-tinged fluid; they do not rupture easily with gentle contact. If the blisters do rupture, pain may occur but healing is usually rapid and resolves without scarring.⁶

Bullous pemphigoid usually itches and in its early phase urticarial (hives) lesions may be present before blisters are noted.

The goal of therapy is to reduce symptoms (itching and formation of new blisters). Local skin care with antibacterial ointment to cover eroded blisters is recommended for all patients to decrease the likelihood of developing a secondary bacterial infection. High potency topical corticosteroid creams are typically used as first-line treatment.⁷ If topical application is not an appropriate option (for example, the patient is non-compliant), oral corticosteroids, such as prednisone, may be considered. Since the cumulative effects of long-term corticosteroid therapy are undesirable, treatment aims at the lowest dose over the shortest period of time.⁸ A 2010 Cochrane review summarized the available treatment data for BP and concluded that very potent topical steroids are effective and safe treatments for BP, but their use in extensive disease may be limited by side effects and practical factors, such as the need to cover large areas with ointment. Initial doses of prednisolone greater than 0.75 mg per kg per day do not give additional benefit and could cause more adverse reactions. Doses lower than 0.75 mg per kg per day may be adequate to control disease and reduce likelihood and severity of potential side effects.

An anti-inflammatory antibiotic, doxycycline, has been studied to treat BP. The benefit of using non-steroidal agents is their superior safety profile. This is often combined with vitamin B3 also called niacinamide.

II. Case Report

A 65-year-old man presented with fluid filled lesion over the body predominantly over the genitals since 4 months. Vesicles ruptured within 1 day, healed with scarring and was associated with intense itching. There was remission and relapse of the lesions every 2 weeks since then without involving any other mucosal areas.

On examination there were multiple vesicles and erosions present over the scrotum and shaft of the penis and over the prepuce. Crusted erosions present over the scrotum. Few vesicles present over the medial aspect of the thighs. Multiple hyperpigmented macules and crusted erosions present over forearms, arms & few over abdomen, chest and thighs, Skin biopsy for DIF and Histopathology were done.

Chief Complaints

➤ Fluid filled lesion over the body predominantly over the genitals since 4 months and aggravated since 4 days.

History of Presenting illness

- Started as a vesicle over the scrotum which ruptured within one day on its own leaving behind raw area which healed with scarring.
- H/o itching severe in intensity present throughout the day and more at night.
- No h/o weight loss/loose stools/similar complaints in family/drug intake prior to lesions.
- Multiple vesicles and erosions were present over the scrotum, shaft of the penis and thighs.
- Nikolsky Sign – Negative



Figure 1: Genital Bullous Pemphigoid

HISTOPATHOLOGY

Subepidermal bullae seen showing proteinaceous material admixed with lymphocytes, eosinophils and occasional neutrophils. Dermis shows perivascular lymphocytic infiltration admixed with eosinophils.

DIF: There was linear staining of basement membrane zone with IgG and C3.

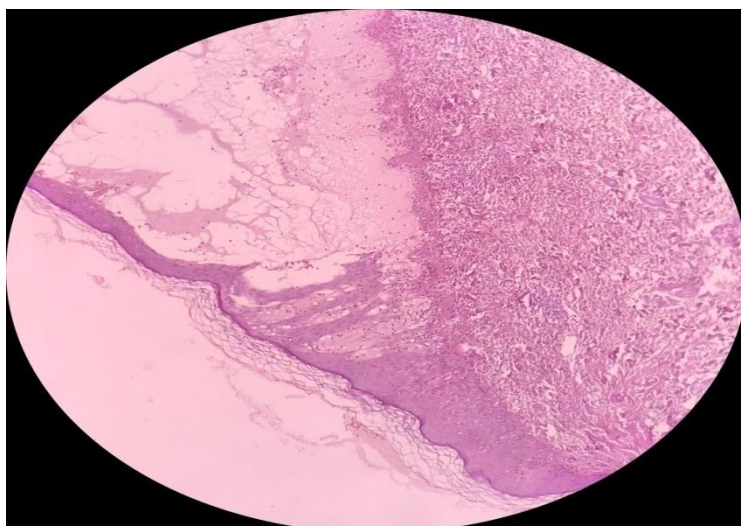


Figure 2: Dermal infiltration by inflammatory cells

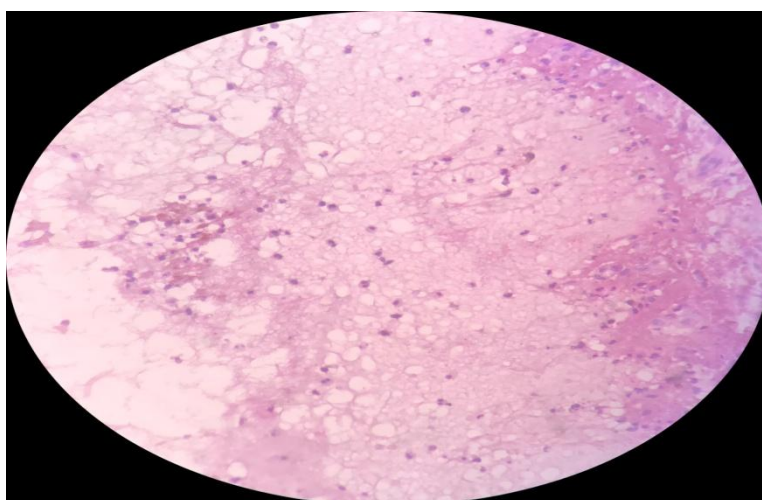


Figure 3: Eosinophils

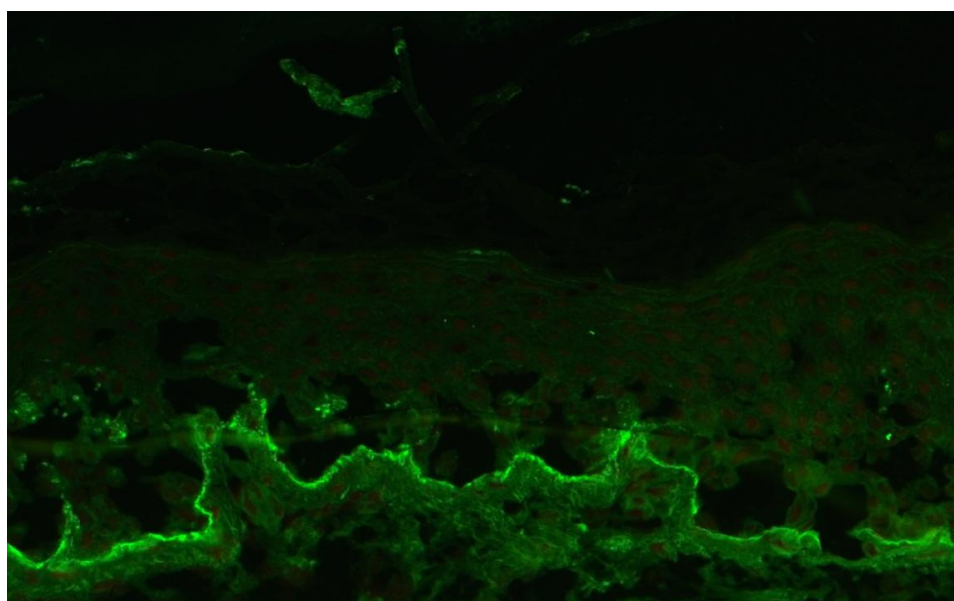


Figure 4: Linear staining of basement membrane zone with IgG and C3.

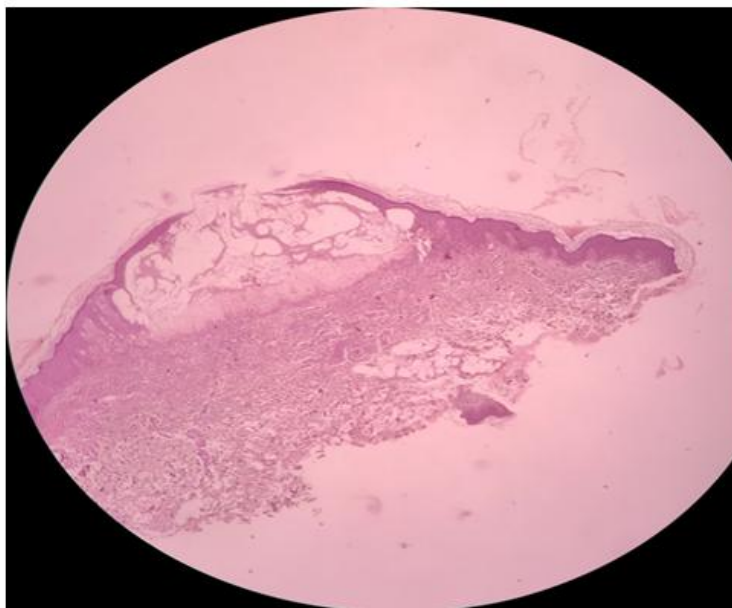


Figure 5: Sub epidermal blisters with eosinophils

III. Discussion

BP is an autoimmune, subepidermal blistering condition associated with immune-pathological reactivity against the BP180 or BP230 hemi- desmosomal components of the basement membrane. The clinical consequences are detachment of the epidermis from the dermis and the formation of subepidermal blisters. BP can be generalized, affecting the trunk and proximal limbs, or it can be localized. Localized BP is a rare subgroup, affecting 16–29% of patients. It is currently unknown why patients develop localized disease only, but factors such as trauma, ultraviolet light and hydrostatic pressure have been proposed.¹¹

Localized BP is a rare autoimmune subepidermal bullous disorder characterized by chronic intermittent eruptions that affect only a restricted area of the body.⁹

Clinically, the lesions of localized BP are similar to those of the generalized type. They can be urticarial, vesicular, bullous or eczematous. The diagnosis is usually delayed because they can mimic many localized bullous diseases.

Localized BP should be considered in cases in which localized bullae or erosions are chronic and recurrent. A correct diagnosis requires a high index of suspicion as well as confirmation with histology and immunofluorescence studies.¹⁰

IV. Conclusion

Although GBP in males is rare, the clinical presentation should induce suspicion as quick and accurate diagnosis is imperative to implement effective therapy and achieve remission.

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