# Toxic epidermal necrolysis-like bullous pemphigoid: a rare case report

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Bullous pemphigoid is the most common type of subepidermal autoimmune bullous disease. It usually affects older people; isolated cases among people younger than 65 have been reported with various presentations, usually more severe and active than those seen in the elderly. Toxic epidermal necrolysis (TEN) is a potentially life-threatening dermatologic disorder characterized by widespread necrosis and bullous detachment of the epidermis and mucous membranes resulting in exfoliation and possible sepsis. Various morphological variants of bullous pemphigoid have been reported, with reports of the TEN-like variant of bullous pemphigoid being scarce. In this study, we report a case of bullous pemphigoid with TEN-like presentation in a middle-aged female.

**Keywords:** bullous pemphigoid, toxic epidermal necrolysis, middle-aged female

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## INTRODUCTION

Bullous pemphigoid (BP) is an autoimmune subepidermal bullous dermatosis that generally affects people older than 70 [1]. Lesions start as itchy urticarial lesions mostly along the abdomen followed by the extremities, eventually erupting to form tense blisters with sero-hemorrhagic content [2]. Confluent bullae with large areas of denuded epidermis can mimic the epidermal sloughing classically seen in toxic epidermal necrolysis (TEN) [3]. Little has been documented about TEN-like bullous pemphigoid. This case highlights the vigilance required by healthcare workers in recognizing the change in the disease course with prompt management to prevent undue complications.

## CASE PRESENTATION

A 34-year-old uneducated female farmer presented to the skin emergency department with multiple fluid-filled lesions over the entire body since seven days earlier and oral intolerance to food since five days earlier. She was alright seven days back before she developed red-colored, intensely itchy lesions over the dorsum of the hand, which progressed to involve the trunk, both extremities, and face, followed by the appearance of tense blisters and the development of oral lesions. On general examination, the patient had pallor with left pedal edema. On dermatological examination, there were multiple tense bullae on an erythematous urticarial base with widespread erosions over the trunk, both limbs, and face

(Figure 1). We admitted the patient with differential diagnoses of bullous pemphigoid and erythema multiforme. Routine laboratory investigations were normal except for anemia. Histopathology showed subepidermal blister formation consistent with bullous pemphigoid. We started her on injectable dexamethasone 6 mg with supportive therapy. Still, 10-12 new blisters emerged every day. Five days after admission, the patient developed generalized peeling of the skin over her entire body with tenderness involving more than 50% of the body surface area. On examination, there was peeling of the skin with a positive pseudo-Nikolsky sign (Figure 2). In view of this, we clinically reviewed her differentials and took a skin biopsy, which was again suggestive of bullous pemphigoid, with no necrotic keratinocytes (Figure 3). Direct immunofluorescence was negative, and electron microscopy re-confirmed the diagnosis as bullous pemphigoid. The lesions had improved by day 17

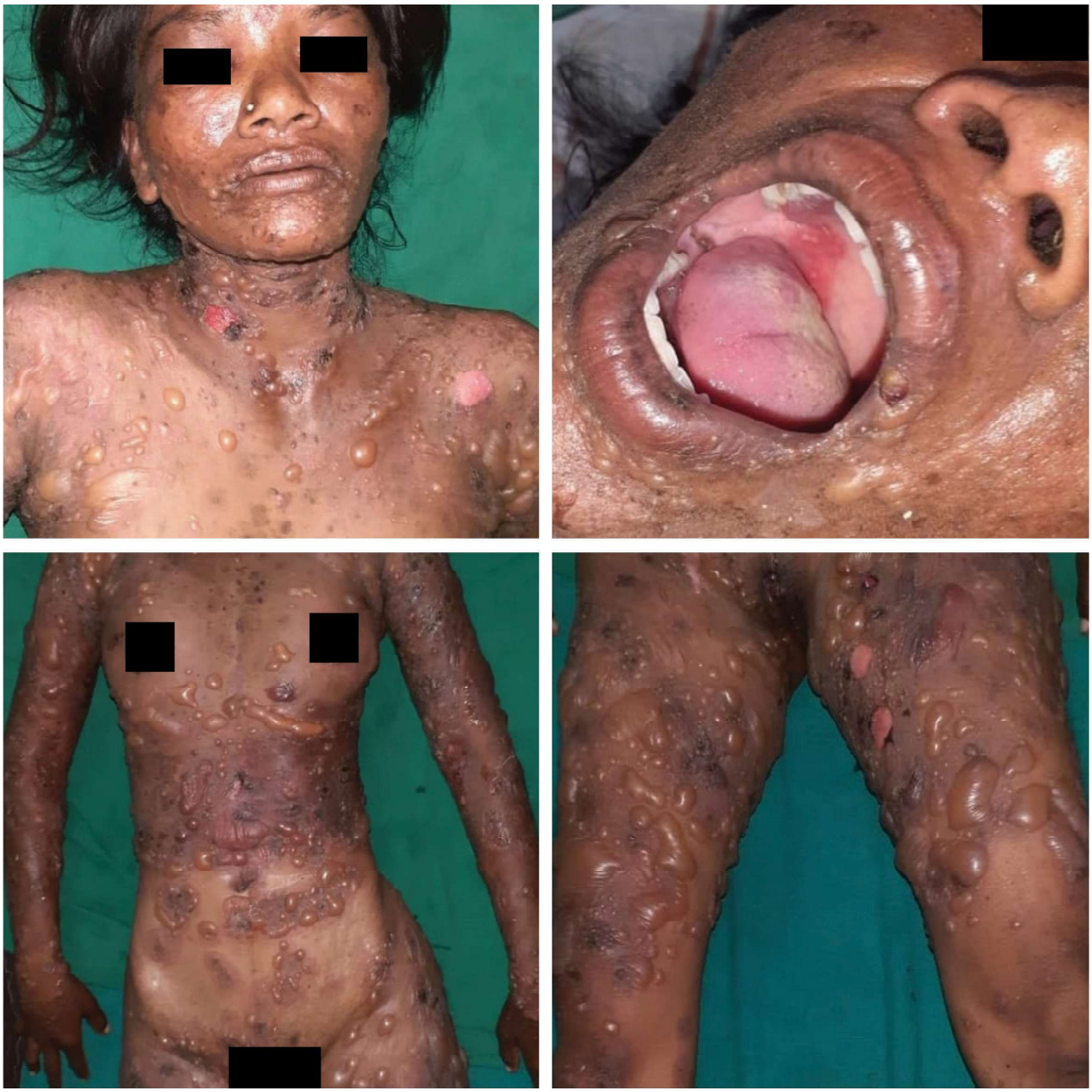

**Figure 1.** Day 1 of admission; multiple tense bullae on an erythematous urticarial base with widespread erosions over the trunk, limbs, and face

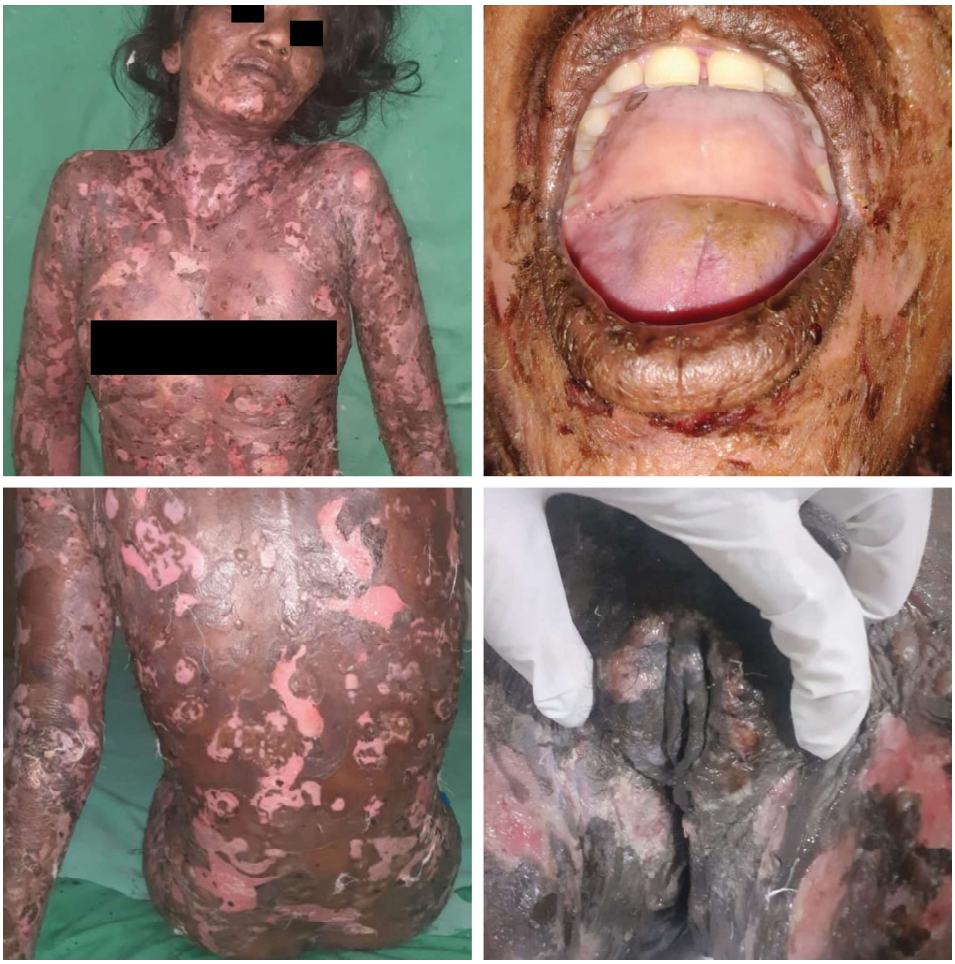

**Figure 2.** Day 5 of admission; generalized peeling of the skin over the entire body, involving more than 50% of the body surface area

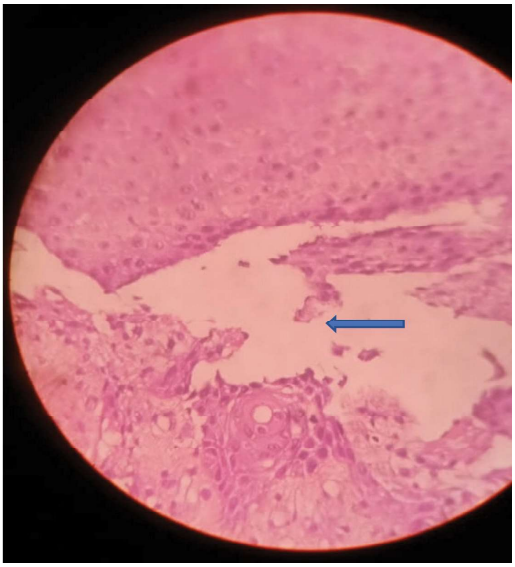

**Figure 3.** Histopathology section, with a blue arrow showing subepidermal splitting

of admission, evidenced by healing and crusting (Figure 4).

## DISCUSSION

Bullous pemphigoid is a blistering autoimmune disease caused by autoantibodies against the collagen XVII (BP180/BPAG2) and BP230 (BPAG1) components of the hemidesmosome [4]. Various morphological variants are described clinically. Common ones include classic, localized, nodular, and vegetating; erythrodermic-like, ecthyma-like [5], and TEN-like [6] are some of the rare ones.

Our patient had urticarial plaques with the development of multiple tense blisters at admission that rapidly evolved to tender confluent bullae and widespread epidermal detachment. Bullous pemphigoid features subepidermal blister formation with eosinophilic infiltrates, while in TEN, there is

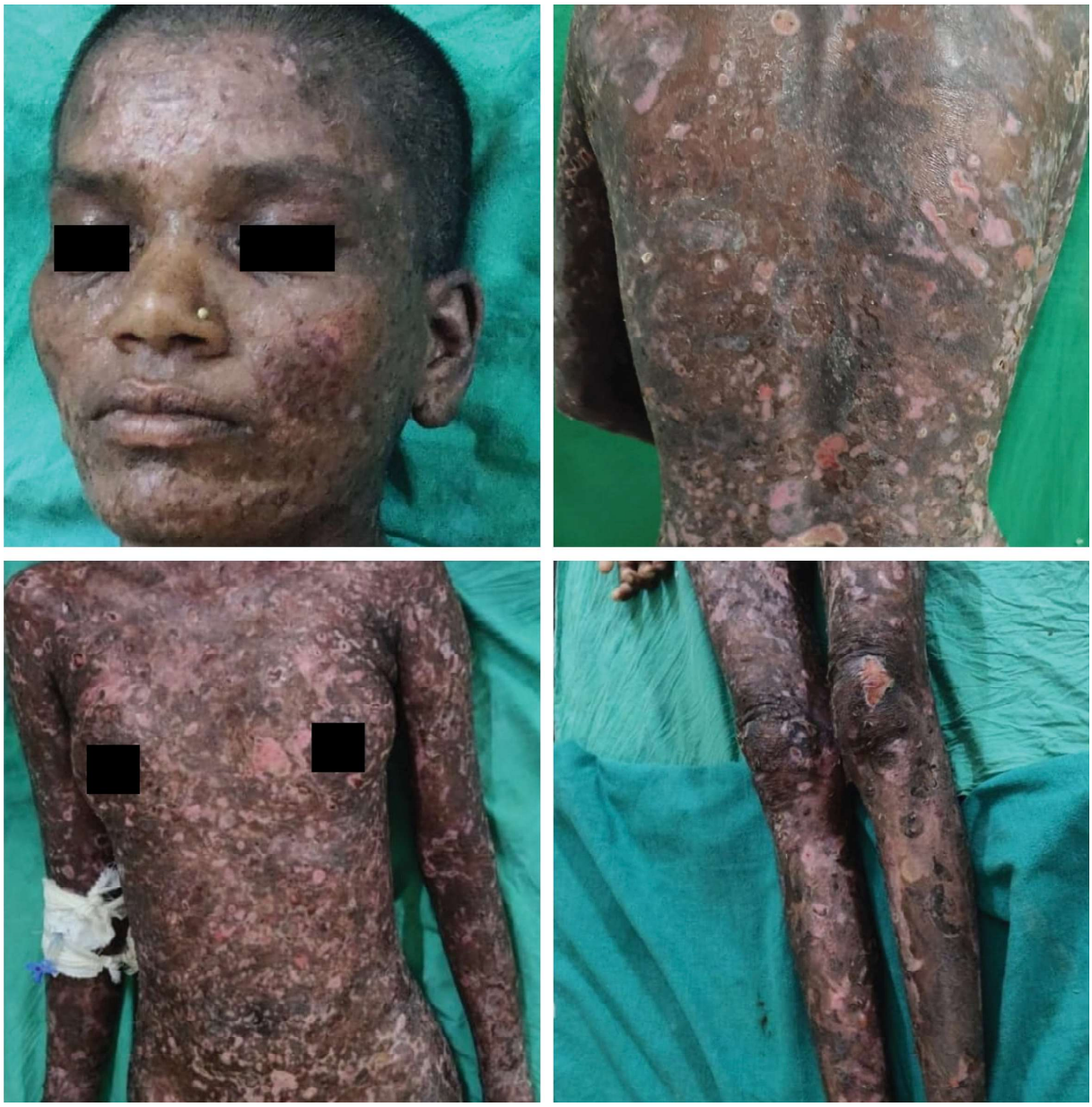

**Figure 4.** Day 17 of admission; improved lesions, evidenced by healing and crusting

subepidermal bullae, loss of the entire epidermis, and the presence of necrotic keratinocytes. As the initial presentation was confirmed as bullous pemphigoid, the course of the disease altered rapidly to evolve into generalized peeling. Also, histopathology specimens at different times of the disease course correlated with eosinophil-rich sub-epidermal blisters seen in bullous pemphigoid. Negative immunofluorescence studies, as in this case, occur in 4% of patients with bullous pemphigoid [7]. It was electron microscopy that further confirmed the diagnosis as bullous pemphigoid.

Bullous pemphigoid presents with urticarial papules or eczematous plaques, whereas TEN begins with tender, dusky, ill-defined erythematous patches. The classic bullous pemphigoid lesion is a 1–3 cm tense bulla on an erythematous base,

differentiating bullous pemphigoid from TEN. Mucosal involvement occurs almost always (90%) in TEN, but is only seen in roughly 20% of bullous pemphigoid patients [8].

## CONCLUSION

Bullous pemphigoid usually presents as red itchy lesions with urticarial plaques followed by tense blisters. TEN-like BP is a rare presentation that was seen in our case.

**Conflict of interest:** None declared.

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