An Unusual Intersection of Dermatology and Cardiology: Urticarial Bullous Pemphigoid and Microvascular Angina in a 68-Year-Old Male from Western India – Case Report.

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Abstract:
Bullous pemphigoid (BP) is an autoimmune subepidermal blistering disease. Urticarial BP, an uncommon variant, presents with pruritic urticarial plaques and erosions rather than tense bullae. We describe a 68-year-old male from Western India with pre-existing ischemic heart disease (microvascular angina) who presented with a 7-month history of pruritic rash and burning sensations affecting his extremities and buttocks. A skin biopsy confirmed urticarial BP. The patient achieved remission within two months following treatment with a combination of systemic immunosuppressants and topical corticosteroids.

Keywords: Bullous pemphigoid, Urticarial bullous pemphigoid, Autoimmune blistering disease, Case report, Ischemic heart disease, Microvascular angina

Introduction:

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering disease typically characterized by tense bullae. Urticarial BP, a rare variant, is characterized by pruritic urticarial plaques and erosions rather than the classic tense blisters [1]. Representing 10-20% of BP cases [2], urticarial BP can pose diagnostic challenges due to its atypical presentation. We present a case of urticarial BP in a 68-year-old male with ischemic heart disease (microvascular angina), emphasizing the importance of including this variant in the differential diagnosis of chronic urticarial eruptions.

Case Presentation:

A 68-year-old male from Western India, with a known history of controlled hypertension, diabetes mellitus, and ischemic heart disease (microvascular angina) on conventional medications, presented with a 7-month history of an itchy rash and burning sensations on his upper and lower limbs, as well as his buttocks. He denied any past history of similar lesions, photosensitivity, family history of blistering disorders or recent medication changes. Prior treatment with oral and topical antifungals provided no relief.

Physical examination revealed scattered plaques and erosions on the scalp vertex, extensor surfaces of the both arms and forearms, and anterior aspects of the both lower limbs. Similar lesions, characterized by hyperpigmented plaques with erythematous, irregular borders, were noted on the left buttock. Mucosal involvement (oral, genital, and conjunctival) was absent. Peripheral nerves were non tender and not thickened. Sensory, Motor examination and Muscle Strength testing were within normal limits.

Laboratory investigations, including complete blood count, liver and renal function tests, Mantoux test, and chest radiograph, were unremarkable. The absolute eosinophil count was within normal limits. ECG showed non-specific ST-T changes and a positive stress test for ischemia and coronary angiography showed normal epicardial vessels but reduced coronary flow reserve confirmed the diagnosis of microvascular angina.

Skin biopsies from a plaque on the left forearm and a vesicle on the left second finger demonstrated subepidermal vesiculation with neutrophils and a mild to moderate superficial dermal lymphocytic infiltrate, consistent with BP.

Based on the clinical and histopathological findings, a diagnosis of urticarial BP was established. Treatment was initiated with Tablet Azathioprine 50 mg daily, Tablet Doxycycline 100 mg daily, Tablet Levocetirizine 10 mg daily, short course of Tablet Prednisolone 20 mg after breakfast and Tablet Mirtazapine 7.5 mg at bedtime, Tablet Pantoprazole 40 mg daily, and topical Betamethasone cream. The patient's microvascular angina was deemed stable, managed with conventional medications including calcium channel blockers and beta-blockers as needed.

The patient responded favourably, achieving complete resolution of lesions within two months and remaining in remission for the subsequent two months.

Discussion:

Urticarial bullous pemphigoid is a rare variant of BP, accounting for only 10-20% of cases [1, 2]. Its clinical presentation of pruritic urticarial plaques and erosions, often without tense bullae [3], can lead to diagnostic confusion.

The pathogenesis of urticarial BP likely involves autoantibodies targeting hemi-desmosomal proteins BP180 and BP230, triggering complement activation and inflammatory cell recruitment, ultimately resulting in the characteristic skin lesions [4].

The atypical presentation of urticarial BP can mimic other dermatological conditions such as chronic urticaria, eczema, or drug eruptions [5], potentially delaying diagnosis, as observed in this case.

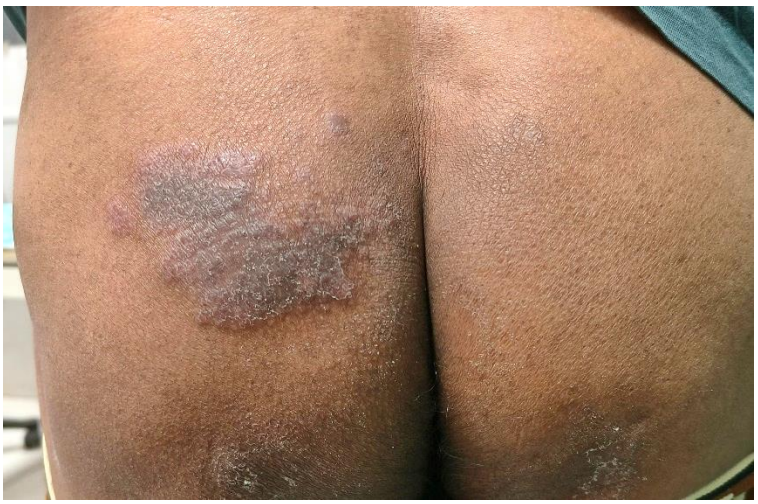

Fig. A – Hyperpigmented plaques with erythematous borders with scales on the left buttock.

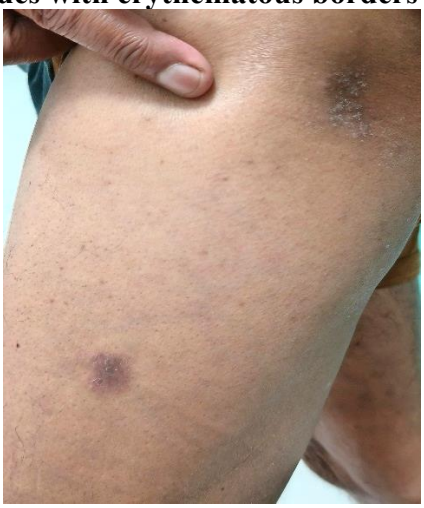

Fig. B – Hyperpigmented to erythematous Plaques with fine scales on lateral aspect of lower limb.

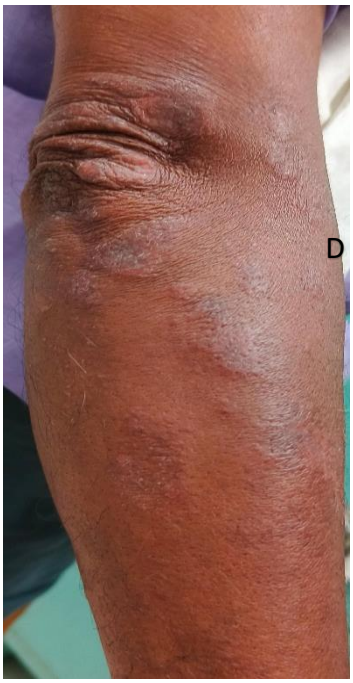

Fig. C – Well defined hyperpigmented plaques with scales at places on extensor aspect of forearms.

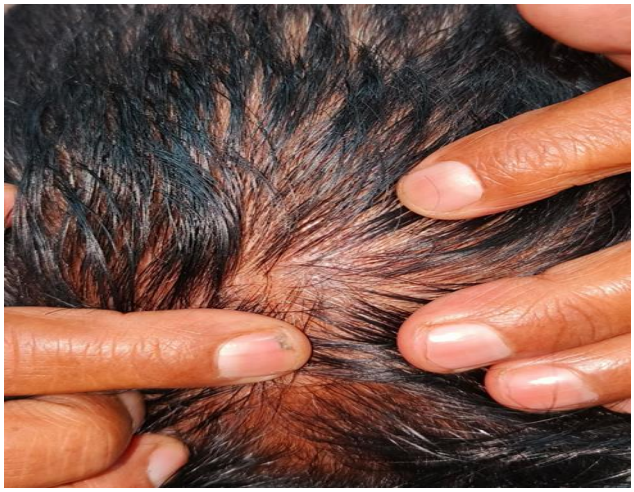

Fig. D – Ill-defined plaques and erosions on the vertex of Scalp.

Management of urticarial BP typically involves systemic immunosuppressants, including corticosteroids and steroid-sparing agents (e.g., azathioprine, mycophenolate mofetil), along with topical corticosteroids [6]. This case presents unique aspects: the rare combination of Urticarial BP and microvascular angina, the challenges in concurrent management, the potential for immunological links between the conditions, and treatment considerations and drug interactions. Our patient demonstrated a positive response to this therapeutic approach.

Conclusion:
Urticarial bullous pemphigoid (BP) is a rare variant of BP that poses diagnostic challenges due to its atypical presentation, often lacking the classic bullae. This case underscores the necessity of including urticarial BP in the differential diagnosis of chronic urticarial eruptions, especially when bullae are absent and in patients with comorbidities such as ischemic heart disease. The coexistence of microvascular angina in this patient further complicates the diagnostic process, highlighting the need for a comprehensive approach to care.

The successful resolution of the patient's skin lesions with systemic immunosuppressants and topical corticosteroids demonstrates the efficacy of this treatment strategy for urticarial BP. Nonetheless, further research is essential to understand the pathophysiological mechanisms underlying urticarial BP and its potential links to other medical conditions, including cardiovascular disease. This case report adds to the existing literature on urticarial BP and emphasizes the importance of heightened awareness and understanding of this rare condition among clinicians.

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Conflicts of interest:
The authors declare that they have no conflicts of interest.

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