ISSN : (Online): 1898-7249
Abbreviation: J Derm Cse Rep
DOI: 10.61705/jdcr.18.3.2025.51.55

Journal of Dermatological Case Reports

Extensive Milia Formation in Bullous Pemphigoid: A Rare Healing Sequela

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Keywords:
Bullous Pemphigoid, Milia, Autoimmune blistering disorder, Direct immunofluorescence, Sequelae of Bullous Pemphigoid

Abstract:

Background: Bullous pemphigoid (BP) is the most common autoimmune subepidermal blistering disorder, predominantly affecting the elderly. Healing usually occurs with post-inflammatory hyper- or hypopigmentation, but the development of milia is rare and more often associated with epidermolysis bullosa acquisita (EBA).

Case Presentation: We report a 60-year-old woman with chronic bullous pemphigoid, who, during the healing phase, developed widespread and numerous milia diffusely involving the trunk and extremities. Milia were present over more than 40% of body surface area, with hundreds of dome-shaped pearly white papules ranging from 1–5 mm, clustered over previously involved sites, including the arms, legs, dorsum of feet, trunk, and peri-umbilical region. Diagnosis was confirmed by skin biopsy demonstrating keratin-filled cysts.

Conclusion: Milia formation, although uncommon in BP, may occur extensively and mimic other immunobullous disorders such as EBA. Awareness of this rare sequela is essential to avoid diagnostic confusion and to guide appropriate patient counseling

Received : 11-08-2025 **Revised : 26-08-2025** **Accepted: 05-09-2025** **Published : 12-09-2025**

Introduction

Bullous pemphigoid (BP) is the most frequent autoimmune subepidermal blistering disease in elderly patients, characterized by intensely pruritic urticarial lesions followed by tense bullae. It is caused by autoantibodies directed against hemidesmosomal proteins BP180 and BP230, leading to complement activation and dermoepidermal separation. Healing generally occurs with pigmentary alteration, without scarring or cystic lesions.

Milia—small, keratin-filled epidermoid cysts—are typically seen in epidermolysis bullosa acquisita (EBA), mucous membrane pemphigoid, and other blistering dermatoses. Their occurrence in BP is considered rare, though recent studies suggest a prevalence of 7–8%. Extensive or diffuse milia formation in BP is distinctly uncommon.

We present a case of a 60-year-old woman with chronic BP, who developed widespread milia across multiple body regions during healing—a unique and diagnostically challenging evolution.

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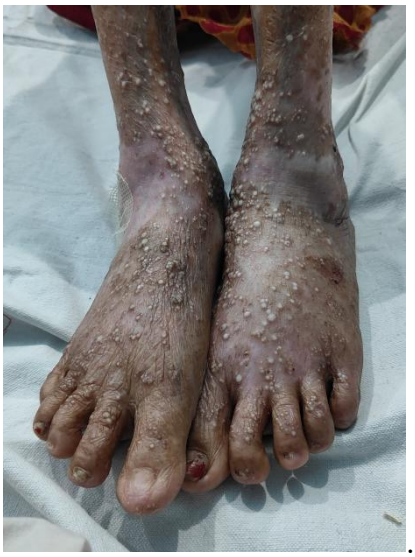
Case Presentation

A 60-year-old woman presented with a three-year history of recurrent, generalized, pruritic blistering eruptions, beginning behind the ears and later involving the trunk and extremities. The lesions were tense bullae measuring 0.5–3 cm, some intact and others ruptured with crusting. Mucosal involvement was initially absent, but she later developed dysphagia to solids, persistent pruritus, and facial swelling. Nikolsky sign was negative.



Figure(a) : Showing extensive milia over feet

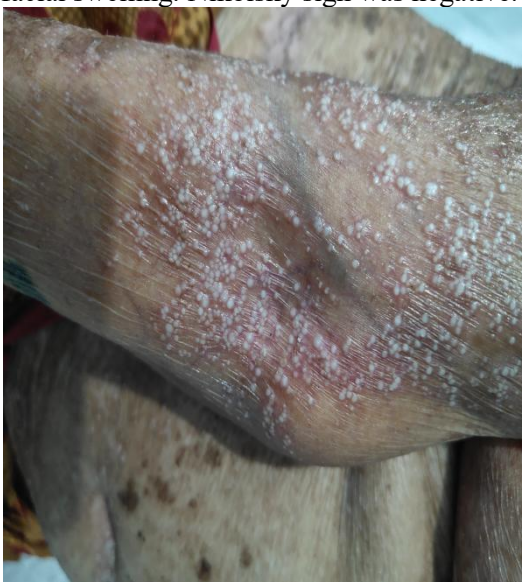


Figure (b) : Showing extensive milia over elbows (extensor aspect)

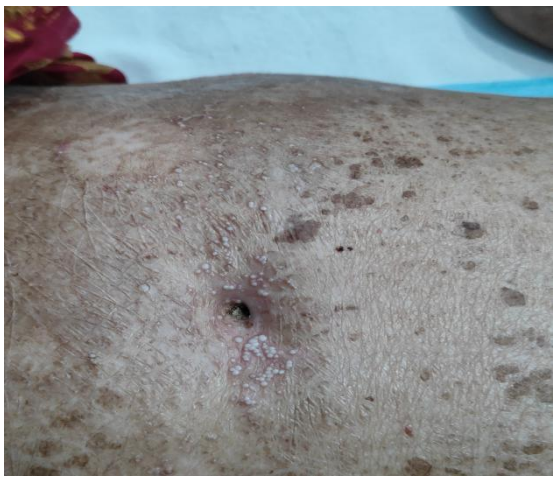


Figure (c) : Showing milia at peri-umbilical area

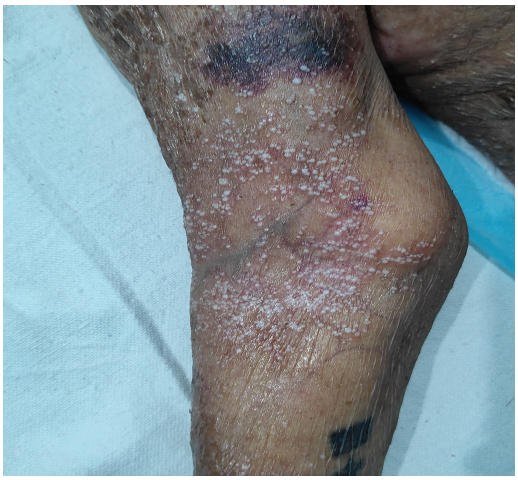


Figure (d) : Showing milia over flexural aspect of elbow joint

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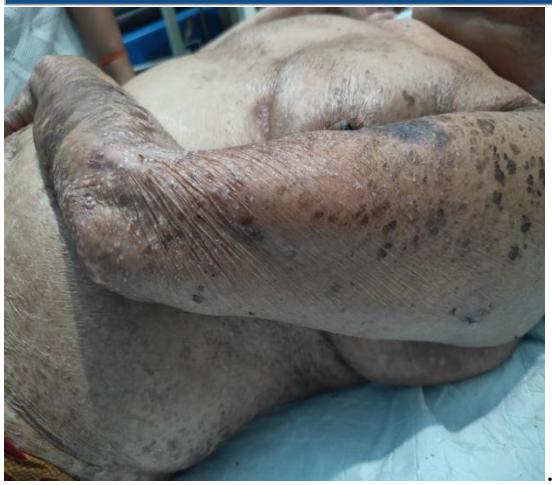

Figure (e) : Showing erythema, superficial crusting and milia over arm,elbow and forearm.

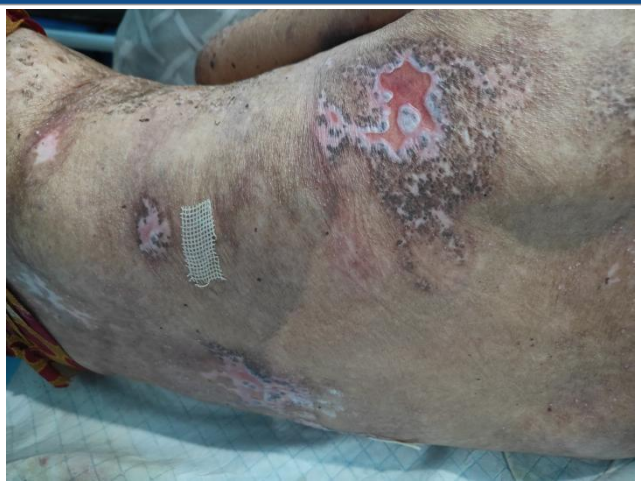

Figure (f) : Showing superficial erosion,and scattered milia over back.

Investigations

Two punch biopsies from the arm and leg revealed subepidermal blistering with an eosinophil-rich dermal infiltrate. Direct immunofluorescence (DIF) demonstrated strong linear IgG and C3 deposition (3+) at the dermoepidermal junction, with weaker C3 positivity (1+). IgA was negative. These findings confirmed bullous pemphigoid.

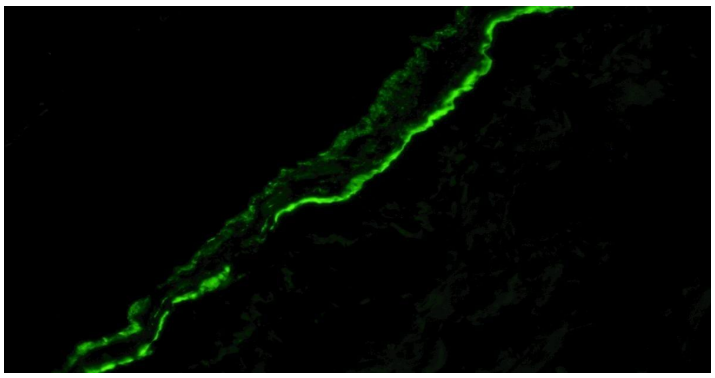

Figure (g): Showing DIF - Features were consistent with linear immune deposits.

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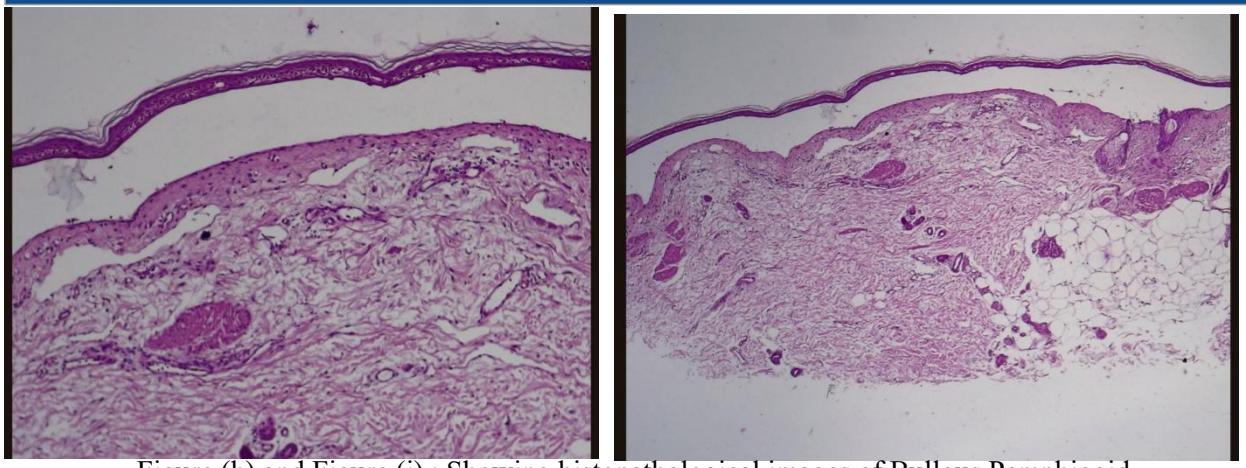

Figure (h) and Figure (i) : Showing histopathological images of Bullous Pemphigoid

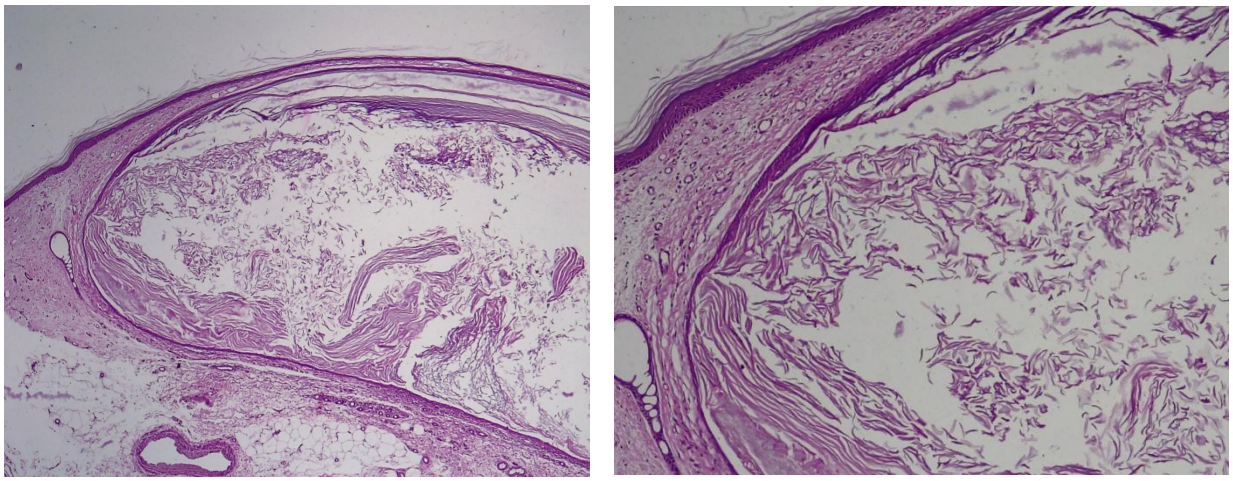

Figure (j) and Figure (k) : Showing histopathological images of Milia

Treatment and Clinical Course

The patient received intravenous dexamethasone in tapering doses, oral dapsone, and supportive treatment. Her bullous lesions gradually resolved with residual post-inflammatory pigmentary changes.

Development of Milia

During follow-up, after several weeks of healing, she presented with extensive diffuse milia over previously involved sites:
Upper extremities: Numerous 1–5 mm pearly white, dome-shaped papules densely scattered over forearms and elbows
Lower extremities: Profuse involvement of both legs, ankles, and dorsum of feet, with confluent clusters of milia
Trunk: Scattered and grouped lesions across the abdomen, particularly peri-umbilical, and over the back and chest
Distribution: In total, lesions covered >40% of body surface area, with several hundred papules evident on clinical examination.
Dermoscopy revealed bright white spherical structures consistent with keratinous cysts. A repeat biopsy confirmed dermal keratin-filled cysts, diagnostic of milia.

Discussion

Milia formation during BP healing is an unusual but increasingly recognized phenomenon. While BP usually resolves with pigmentary changes, a small proportion of patients develop secondary milia. The reported prevalence ranges from 7–8%.

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Pathogenesis

The exact mechanism is unclear, but hypotheses include:
Regeneration of disrupted adnexal structures (sweat glands, hair follicles) after subepidermal blistering, leading to keratin entrapment.
Immunogenetic predispositions such as HLA-DQ6.
Autoantibody profiles (anti-BP180 C-terminal, LAD-1) and high IgE levels have been implicated.

Clinical Significance

Milia are more commonly associated with EBA, where they appear with scarring, thus their presence in BP may cause diagnostic confusion. In our patient, the extensive, diffuse distribution over the trunk and extremities emphasized the importance of considering BP in the differential when milia are seen after blister resolution.
Compared to previously reported cases, the sheer number, distribution, and surface area involvement in our patient are particularly striking and highlight an extreme end of the BP–milia spectrum.

Conclusion

This case demonstrates an unusual healing outcome of bullous pemphigoid with diffuse, extensive milia formation covering over 40% of body surface area. Recognizing this rare sequela is important to avoid misdiagnosis with EBA and to appropriately counsel patients about disease prognosis.

Patient Consent
Informed consent was obtained. The patient understands that identifying details will not be published and that anonymity cannot be guaranteed.

Conflicts of Interest
None declared.

Financial Support
Nil.

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