Pemphigus and Bullous Pemphigoid in Indian Skin: A Study done in Tertiary Hospital of Bihar

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Abstract:
Clinical diagnosis of Pemphigus and Bullous pemphigoid relies heavily on the appearance of the bulla which varies depending on multiple factors like age of the lesion, itching, previous or self application of steroids, and most importantly skin tone. Most of the medical literature is derived from studies in the western population and hence leaves a large gap in the knowledge of presentation of these diseases in the Indian population. An accurate representation of these varied clinical presentations is required to enable the clinicians to make a competent diagnosis.

Introduction

Cutaneous disorders have different presentations and pigmentary sequelae depending on the skin tone of patient. Most medical literature use images that feature the white skin in which the presentation differs vastly from the non-white skin. Indian skin is largely underrepresented in the medical literature, which leads to difficulty in correct recognition and treatment of these diseases. Studies have shown incidence of pemphigus in India varies from that in the western countries with incidence being 0.09 to 1.8% among out patients. This is found to be higher than the available data from Germany, France and lower Tunisia. [1] Studies from North India show increased incidence from Punjab and Haryana. [2] Global reports for Bullous pemphigoid show incidence of 2.4 and 21 cases per million people, where as reports from Indian population is sparse. [3-4]

Case 1
A 62 yr old male presented with pruritis followed by blistering on extremities since 6 months which were found to be tense and lacked erythema upon physical examination (Fig 1, A). Direct immunofluorescence showed IgG positivity at the dermoepidermal junction in serrated pattern suggestive of Bullous pemphigoid (Fig. 1; B) and Histopathology showed presence of sub epidermal cleft and dermis infiltrated predominantly by
eosinophils confirming the diagnosis of Bullous pemphigoid (Fig 1, C). The patient was put on treatment with oral steroids. Post recovery hyperpigmentation could be seen in the areas of the blisters (Fig 1, D).

Figure 1: Case 1 (A) tense blisters on leg lacking erythema (B) H&E stained at low magnification showing sub epidermal clefts and eosinophilic infiltrates. (C) DIF showing 4+ positive serrated pattern of IgG at dermoepidermal junction (D) Post recovery hyperpigmentation

Case 2
A 55 yr old male presented with pruritis which was followed by blisters on trunk and upper extremities since 4 months. On physical examination multiple bulla and erosions were found on the back, chest and arms without any erythema (Fig 2, A). A clinical diagnosis of Bullous pemphigoid was made. On histopathology, section showed sub epidermal cleft and dermis markedly infiltrated by eosinophils as well as eosinophils infiltrating the cleft space confirming the diagnosis of Bullous pemphigoid (Fig 2, B). Patient was put on oral steroids and presented post recovery with hypo pigmented patch with follicular prominence with pigmentation (Fig 2, C)
Case 3

A 29 yr old female presented with complaint of pruritic blisters on hands and legs since 1 months. On physical examination, multiple erosions of skin were seen which did not have an erythematous base (Fig. 3, A). DIF showed 2+ IgG deposits in the intercellular spaces of epidermis suggesting diagnosis of Pemphigus (Fig. 3, B). On histopathological examination, supra basal cleft containing acantholytic cells along with mixed inflammatory cell infiltrate in dermis were seen suggesting a diagnosis of Pemphigus vulgaris (Fig. 3, C). Patient was started on oral corticosteroids and Rituximab. Post recovery the blisters healed leaving behind areas of hyperpigmentation (Fig. 4, D).

Figure 2: Case 2 (A) Blisters and erosions on upper arm and chest lacking erythema (B) Scanner image of H&E section showing subepidermal cleft and dermis markedly infiltrated by eosinophils (C) Post treatment - hypo pigmented patch with follicular pigmentation

Figure 3: Case 3 (A) erosions present on hand lacking erythematous base (B) DIF showing 2+ IgG deposits in intercellular spaces of epidermis (C) H&E stained section on scanner view showing supra basal cleft containing acantholytic cells and dermis infiltrated by mixed inflammatory infiltrate( D) Post treatment hyperpigmentation.
Case 4
A 72 yr old female, known case of hypertension, diabetes mellitus and tuberculosis, presented with oral erosions since 15 days, redness around the eyes since 3 days and blisters on back lacking erythema and chest since 3 days. On physical examination, multiple flaccid bullae and vesicles were seen (Fig. 4, A & B). The DIF results were negative but the histopathology showed supra basal split containing acantholytic cells, neutrophils and eosinophils, and dermis showing perivascular and periadnexal mixed inflammatory infiltrates confirming the diagnosis of Pemphigus vulgaris (Fig. 4, C). Patient was treated with oral corticosteroids and Rituximab. Post treatment the cutaneous blisters healed with hyperpigmentation where as mucosal erosions healed without any pigmentation (Fig. 4 D & E).

![Figure 4: Case 4 (A) Oral erosions (B) Blisters on back lacking erythema (C) H & E stained section on scanner view showing supra basal cleft (D) Post treatment healing with hyperpigmentation of cutaneous blisters (E) and mucosal erosions healing without pigmentation.](image)

Case 5
A 45 year old female presented with erosions on face, neck and chest since 15 days (Fig. 5, A). DIF was done which showed 2+ IgG deposits in intercellular spaces of epidermis forming fishnet pattern which was suggestive of Pemphigus vulgaris (Fig. 5, B). On histopathology, papillary dermis lined by a layer of basal epidermal cells was seen with a few acantholytic cells lying above it was seen. The dermis showed perivascular infiltration of mixed inflammatory cell infiltrate confirming the diagnosis of Pemphigus vulgaris (Fig. 5, C). The patient was treated with oral corticosteroids and Rituximab after which the erosions healed with hyperpigmentation (Fig. 5, D).
Case 6

A 52 year old female presented with erosions on back and chest since 10 days (Fig. 6, A). DIF was done which showed 2+ IgG deposits in intercellular spaces of epidermis forming fishnet pattern which was suggestive of Pemphigus (Fig. 6, B). Histopathology from the erosion showed papillary dermis lined by a layer of basal epidermal cells with a few acantholytic cells lying above it. The dermis showed perivascular infiltration of mixed inflammatory cell infiltrate confirming the diagnosis of Pemphigus vulgaris (Fig. 6, C). The patient was treated with oral corticosteroids and Rituximab after which the erosions healed with hyperpigmentation (Fig. 6, D)

Figure 5: Case 5 (A) Skin erosions (B) DIF showing 2+ deposits of IgG in fishnet pattern (C) H&E stained section on low power showing papillary dermis lined by basal epidermal cells (D) Post treatment hyperpigmentation

Figure 6: Case 6 (A) Skin erosions (B) DIF showing 2+ deposits of IgG in fishnet pattern (C) H&E stained section on low power showing papillary dermis lined by basal epidermal cells (D) Post treatment hyperpigmentation
Case 7
50 yr old male presented with multiple tiny blisters on the back. On examinations, the bullae were flaccid and lacked erythema (Fig. 7, A). DIF showed IgG 2+ positive in the intercellular spaces epidermis (Fig. 7, B). On histopathology, sub corneal cleft with mixed inflammatory infiltrate in the dermis was seen (Fig. 7, C) confirming the diagnosis of Pemphigus foliaceous. Patient was started on corticosteroids after which the bullae healed leaving behind hyper pigmented patch (Fig. 7, D).

![Figure 7](image1.png)

Figure 7: Case 7 (A) Blisters on back lacking erythema (B) DIF - IgG +ve (C) H&E stained section at scanner showing sub corneal split and dermis infiltrated by mixed inflammatory infiltrate (D) Post treatment hyper pigmentation

Case 8
40 yr old female presented with multiple tiny blisters on the extremities. On examinations, the bullae were flaccid and lacked erythema (Fig. 8, A). DIF showed IgG 2+ positive in the intercellular spaces epidermis (Fig. 8, B). Histopathology shows sub corneal cleft. (Fig. 8, C) confirming the diagnosis of Pemphigus foliaceous. Patient was started on corticosteroids after which the bullae healed leaving behind hyper pigmented patch (Fig. 8, D)

![Figure 8](image2.png)

Figure 8: Case 8 (A) Blisters on extremities lacking erythema (B) DIF - IgG +ve (C) H&E stained section at scanner showing sub corneal split (D) Post treatment hyper pigmentation
Discussion

Diagnosis of a skin pathology depends heavily on the appearance of the lesion. The Indian population has a more pigmented skin tone than the western population and hence a widely different presentation. Since most of the medical literature on bullous disorders is based on research in the western countries, there is a lack of proper representation of the Indian skin making doctors less equipped to effectively diagnose these lesions.

In cases 1 & 2, the patients complain of pruritis before appearance of blister but no erythema, even though the histopathology reveals heavy infiltration by eosinophils. Post recovery, the repigmentation of Bullous pemphigoid shows typical presentation. The lesion shows hypopigmented patch initially, probably due to loss of basal layer. The reglementation occurs in spotted manner, most likely from the stem cells in the appendages [5]. This can be used as a diagnostic sign to identify patients with history of bullous pemphigoid.

In cases 3, 4, 5 & 6 diagnosed to be Pemphigus vulgaris, patients complained of pruritic lesion but presented with no erythema, whereas the dermis showed the presence of inflammatory infiltrates in the dermis. After recovery, the pigmentary sequelae of pemphigus vulgaris is more dramatic than others probably due to intact basal layer. In case 7 & 8, similar pattern is seen.

Conclusion

The bullous disorders have a less erythematous presentation and leave behind a more pigmented sequelae than those represented in the medical literature. To ensure proper diagnosis and increased competence of medical professionals, variety of clinical images from all backgrounds should be used for training and education[6]. The post recovery pigmentation should be included in the differential diagnosis of pigmented lesions of skin.

References