

A rare case of pemphigus vulgaris associated with generalized pustular psoriasis: A case report

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Abstract Pemphigus vulgaris, chronic autoimmune blistering disorder characterized by flaccid vesiculobullous eruption. Generalized pustular psoriasis, characterized by sterile pustules on erythematous background. For the very first time, we are reporting a case of pemphigus vulgaris associated with generalized pustular psoriasis in our community.

Key words

Pemphigus vulgaris, pustular psoriasis.

Introduction

Pemphigus vulgaris is a chronic autoimmune disease of intraepidermal cell adhesions resulting in flaccid blisters and erosions involving the skin and mucosa.¹ It is caused by autoantibodies against Desmoglein-3(Dsg-3) and Desmoglein- 1(Dsg-1). Pemphigus affects 0.1-0.5 patients per 100,000 population per year.² Histopathology shows supra basal split with acantholytic cells. Direct immunofluorescence (DIF) shows deposition of IgG and C3 in the intercellular spaces in a 'fishnet' pattern. Treatment includes systemic steroids and immunosuppressant's like azathioprine, mycophenolate mofetil, IV immunoglobulins, and Rituximab etc.³ Generalized pustular psoriasis is an uncommon variant of psoriasis characterized by widespread pustules on an erythematous background.⁴

Patients of pustular psoriasis are characterized

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by having a history of psoriasis and may have residual lesions of psoriasis vulgaris. Histological epidermal changes are similar to those of psoriasis vulgaris, with parakeratosis and elongation of rete ridges. There is a dermal and epidermal infiltration with spongiform pustules of Kojog.⁴

In literature, many case reports have described the coexistence of psoriasis and autoimmune bullous diseases especially with bullous pemphigoid.⁵ Only one case of Pemphigus vulgaris along with psoriasis was reported.⁶ We are reporting a rare case of pemphigus vulgaris associated with generalized pustular psoriasis first time in Pakistan.

Case report

A 16 years old unmarried female presented to us with pustular eruption all over body, more pronounced on intertriginous areas. She also has small erosions on her back. There was no mucosal involvement. We considered bullous impetigo, IgA pemphigus, subcorneal pustular dermatosis as differentials and IV antibiotics along with dapsone were started.



Figure 1 Initial presentation: Discrete pustules coalescing to form bullae on trunk.



Figure 2 Flaccid vesicles, bullae and erosions on trunk.

Routine investigations with skin biopsy done. 1st biopsy taken from fresh pustule came out to be non-specific. 2nd biopsy was done and histopathological examination showed subcorneal split with acantholytic cells and eosinophils. These findings were not consistent with clinical picture.

Patient's skin lesions were not improving with antibiotics and dapsone. Biopsy was repeated again, sent for histopathology and DIF. According to direct immunofluorescence typical findings of pemphigus vulgaris were shown. Oral corticosteroids with azathioprine started and patient improved dramatically within two

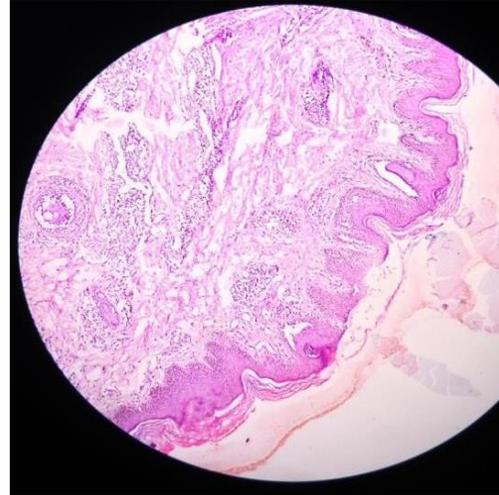


Figure 3 2nd biopsy showed subcorneal split, acantholytic cells along with eosinophilic spongiosis favouring pemphigus foliaceus.

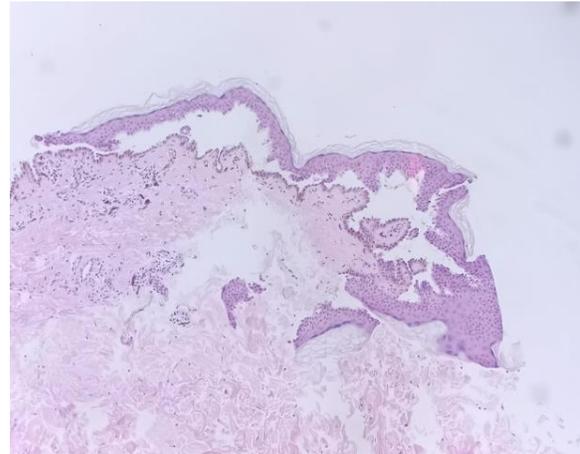


Figure 4 3rd biopsy showed supra basal split with tombstoning favouring pemphigus vulgaris.



Figure 5 Discrete pustules, few forming lakes of pus on erythematous base favouring pustular psoriasis.

weeks. Steroids were tapered. After one week typical lesions of pustular psoriasis erupted. Acetretin started and patient showed response within a week and discharged with steroids, azathioprine and acetretin.

Discussion

Pemphigus vulgaris clinically present as flaccid vesicles, bullae and erosions initially involving the oral cavity, later on affects the skin mainly trunk, scalp, axilla, umbilicus with large erosions.^{1,6} Our patients presented with pustules and pus containing flaccid bullae mainly in flexural areas, trunk and back.

On initial presentation, IgA pemphigus (subcorneal pustular variant), subcorneal pustular dermatosis, bullous impetigo were considered as differentials. Patients with IgA Pemphigus present with flaccid vesicles or pustules in axillary and groin areas. On histopathology there is acantholysis and neutrophilic infiltration.³ In Subcorneal pustular dermatosis, there is symmetrical sterile pustular eruption involving the trunk, intertriginous areas and flexor aspects of the limbs.^{4,7} Histopathology shows subcorneal collection of neutrophils and DIF is negative.^{4,8} Initially our patient present similar to IgA Pemphigus or subcorneal pustular dermatosis, that's why dapsone was started, but biopsy and Immunofluorescence confirmed the diagnosis of pemphigus vulgaris. Bullous impetigo considered due to pus containing bullae, which was excluded from differentials as the cultures showed no growth and patient didn't respond to antibiotic.⁴ Our case is a unique one with pustular form of pemphigus vulgaris as the literature showed only one case of this type.^{3,6,9} Biopsy and immunofluorescence findings ruled out IgA pemphigus, subcorneal variant and subcorneal pustular dermatosis.

Generalized pustular psoriasis is severe form of pustular psoriasis clinically present as discrete sterile pustules on a red fiery skin associated with fever, malaise, joint pain and can progress to erythroderma.^{4,5} Pustules can be discrete, forming lakes of pus and exfoliate as they dry but new wave can recur. Treatment includes use of retinoids e.g. acetretin, methotrexate, cyclosporine, TNF-alpha inhibitors.^{5,6}

Patients with psoriasis are frequently complicated with autoimmune bullous diseases, especially, Pemphigoid diseases.⁵ Mostly Pemphigus Erythematosus was reported to develop in Psoriasis patients.^{5,9}

According to a meta analysis a significant association was found between pemphigus and psoriasis.^{6,10} Physicians managing patients with pemphigus should aware of this comorbidity.¹⁰ We couldn't find any case uptill now in literature showing pemphigus vulgaris coexisting with generalized pustular psoriasis.^{5,8,10} We are reporting the first case in which patient with Pemphigus Vulgaris, coexisting with generalized pustular psoriasis successfully treated with steroids, acetretin and azathioprine.

Conclusion

Pemphigus Vulgaris can present with pustules and pus containing bullae and also can coexist with generalized pustular psoriasis. This is the first case report in our setup highlighting a rare pustular variant of pemphigus vulgaris associated with generalized pustular psoriasis.

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