

# An assertive interventional study on pemphigus vulgaris

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## ABSTRACT

Pemphigus vulgaris (PV) is a rare but potentially life-threatening autoimmune blistering disease affecting the skin and mucous membranes. The disease is caused by circulating antibodies targeted against the desmogleins (desmosomal glycoproteins expressed on the epithelial cells of the mucosa and skin). Disruption of these adhesion proteins results in a loss of cohesion between cells leading to acantholysis and blister formation. The clinical result of this process is the easy rupture of the blisters, leaving behind painful sloughing erosions on the mucosa and/or skin. We report a case of severe PV in a 47-year-old woman presenting with widespread, painful, eroded mucocutaneous lesions. The uniqueness of the disease demanded the pharmaceutical care provided by a clinical pharmacist to successfully manage the problem. This report highlights the importance of an early interdisciplinary team approach to improve the outcome of patients suffering from Pemphigus vulgaris.

**Key Words:** Pemphigus vulgaris; Desmogleins; Acantholysis; Clinical pharmacist

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## INTRODUCTION

Pemphigus was derived from the Greek word ‘pemphix’ meaning bubble or blister and ‘vulgaris’ a Latin word meaning common [1]. Pemphigus vulgaris (PV) is a rare but potentially life-threatening autoimmune blistering disease affecting the skin and stratified squamous mucosa, i.e., larynx, esophagus, conjunctivae, urethra, vagina, cervix, and anal canal in severe cases. PV affects men and women equally. The incidence of pemphigus among the Indian population has varied widely from 0.09 to 1.8%. It was found to be higher than available data from Germany, France but lower than Tunisia.

In India, a significant proportion of patients have been younger than 40 years of age. This is in contrast to other parts of the world where pemphigus occurs between 40 to 60 years of age [2]. Adhesion between cells in the outer layer of skin and mucosa is important to maintain tissue integrity and act as a protective barrier against the external environment. Desmosomes are specific adhesion protein complexes (made up of two types of calcium-dependent glycoproteins called desmoglein and desmocollin) responsible for maintaining cell-to-cell adhesion in this layer. Pemphigus vulgaris (PV) is characterized by the loss of cellular adhesion (acantholysis) due to the formation of IgG autoantibodies against desmoglein 1 and 3 [3,4].

## CASE DESCRIPTION

A 47-year-old female was admitted to the Dermatology department with the chief complaints of oral ulcers, pain in mouth and throat, since 3-4 months, swelling over the nails, lesions over the groin, back neck, labia extended to the anus, per vaginal discharge with itching at perianal region since 2 months. She has taken treatment outside hospitals (Figures 1 and 2). No history of Diabetes and Hypertension. Personal and family histories were not significant. From the complaints, history and clinical examination, a provisional diagnosis of Pustular psoriasis, Secondary syphilis was made. The Final diagnosis of Pemphigus Vulgaris was made based on clinical and histopathological findings. (The consent was taken from the patient before enrolling into the study. Also, the Institutional

Ethics Committee of the college has granted the ethical approval for the study.)

## Investigations

Perilesional Biopsy shows intra epidermal supra basal acantholytic blistering dermatitis. Several acantholytic cells and neutrophils seen in blisters. The floor of the blister shows “tombstone” pattern with occasional acantholytic cells. Mild spongiosis with neutrophils present at the periphery of the blister.

Impression: Pemphigus vulgaris

Serum Australia antigen – Negative

HIV I and II – Negative

STS (serologic test for Syphilis) - Negative



Figure 1: Lesion at back neck .

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Figure 2: Paronychia.



Figure 4: Recovery of patient after withdrawing Clavulanic acid.

### Management

In the Inpatient department (IPD), on the first day, the patient was given with inj. Dexamethasone 8 mg BD, inj. cefotaxime 1g BD, inj. Amoxicillin + Clavulanic acid 625 mg BD, inj. Ranitidine 50 mg BD and diluted chlorhexidine mouthwash (Figures 3-5). On the second day, inj. Amoxicillin + Clavulanic acid was withdrawn from the treatment through the clinical pharmacist intervention while the rest of the medications were continued. On the third day of hospitalization, patient was discharged with the following medication with follow up advice after 2 weeks.

Table 1: Medications on discharge.

Medication	Dose	Duration
	40 mg/day	2 weeks
Prednisolone	20 mg/day	Next 2 weeks
	10 mg/day	Next 2 weeks
Dapsone	100 mg/day	4 weeks
Terbinafine	250 mg/day	2 weeks
Triamcinolone ONT	0.1% w/w	
Ranitidine	150 mg BD	

The patient made a slow but gradual recovery over the next 4 weeks. Two months since discharge, the patient continues to be reviewed on regular basis by the dermatology team (Table 1). She has still prescribed dapsone and is on a tapering dose of prednisolone (currently 10 mg once daily). She has fortunately not suffered a recurrence of the disease till now.



Figure 3: Recovery of patient after withdrawing Amoxicillin.

### DISCUSSION AND CONCLUSION

PV can be either induced or triggered by certain drugs. (Pemphigus that continues after a patient stops taking a drug is referred to as triggered, whereas lesions that start healing soon after withdrawal are referred to as induced). The clinical, histological and immunofluorescence abnormalities of drug-induced pemphigus vulgaris are similar to those of the idiopathic variety. Drugs associated with PV include Penicillin, captopril, enalapril, penicillamine, thioprolone, interleukin-2 (IL-2), nifedipine, piroxicam, and rifampicin [5-7].

As Amoxicillin-Clavulanic acid is found to cause exacerbation of PV and drug-induced PV, it was withdrawn from the prescription through the clinical pharmacist intervention.

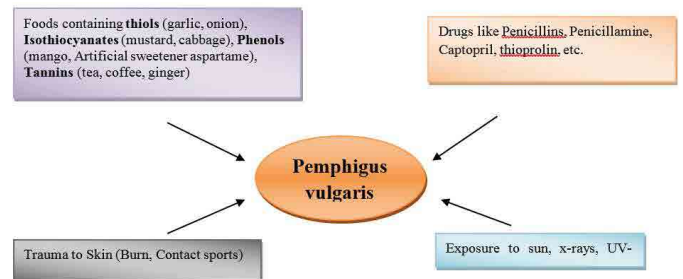


Figure 5: Factors exacerbating to Pemphigus vulgaris.

The present case study emphasizes the role of a clinical pharmacist in the management of PV through therapeutic interventions, consultation with other health care professionals and counseling of patient. The main objective in the management of the pemphigus vulgaris is to control the disease, prevent relapses and avoid adverse events associated with the prolonged use of steroids and immunosuppressive agents. Therefore the prevention and management of patients with PV requires coordination of care between dermatologist, physician and clinical pharmacist.

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### CONFLICT OF INTEREST

The author declares there is no conflict of interest.

## REFERENCES

1. Nagaraju B, Ramu A, Vidhyadhara S. A Case Study on Pemphigus Vulgaris. *IJOPP* 2016;9:4.
2. Kanwar AJ, De D. Pemphigus in India. *Indian J Dermatol Venereol Leprol* 2011;77:439-49.
3. Vinall C, Stevens L, McArdle P. Pemphigus vulgaris: a multidisciplinary approach to management. *BMJ Case Rep* 2013.
4. Kumar SJ, Nehru Anand SP, Gunasekaran N, et al. Oral pemphigus vulgaris: A case report with direct immunofluorescence study. *J Oral Maxillofac Pathol* 2016;20:549.
5. Pemphigus causes; pemphigus research. International Pemphigus and Pemphigoid Foundation (IPPF). CA, US. 2017.
6. James WD, Berger TG, Elston DM. *Andrew's Diseases of the Skin*. 12th edn, Philadelphia, Elsevier 2016;451-3.
7. Autoimmune Blistering Diseases; Risk Factors. International Pemphigus and Pemphigoid Foundation (IPPF). 2017;CA, US.