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REVIEW ARTICLE **OPEN ACCESS**

Emerging Approaches for Management and Treatment of *Pemphigus vulgaris*: Steps to Improve Health

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ABSTRACT
Pemphigus vulgaris considered a social stigma, a chronic, autoimmune disorder affecting the health of the patients. The primary objective of treatment approaches is to control the relapse of the disease in the immune-compromised patient. It is a life-threatening disorder that causes intra-dermal lesions on the skin and oral mucosa, thereby causing difficulty swallowing food and speech. The conventional therapies for treatment remain challenging; adequate steps need to improve P.V.'s incidences. The treatment approaches for P.V. patients manage with corticosteroid therapy, which gives for long durations. Although the corticosteroids are associated with several side effects, the primary purpose of treatment strategies is to reduce the cumulative dose of corticosteroids in the blood. With the emergence of other therapies using intravenous rituximab, azathioprine is a highly potent and successful therapeutic agent in managing P.V. incidences. Pemphigus vulgaris presents a unique challenge for the patients that already have diabetes, suffering from high blood pressure, and other chronic infections. The newer therapeutic interventions are needed to improve the quality of life of immune-compromised patients. The rare occurrence of pemphigus incidences in fewer patients presents a challenge for practitioners to adopt newer therapeutic approaches to treat other infections and control relapse.
Keywords: Pemphigus vulgaris (P.V.), treatment, an autoimmune disorder, rituximab, relapse, health.

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INTRODUCTION

Pemphigus vulgaris is an autoimmune disorder characterized by blisters in oral mucosa epithelium and loss of keratinocytes from the skin surface. The mainstay for the therapy results in multiple complications and results in increased mortality. Systemic glucocorticoids like rituximab and other therapeutics azathioprine and dapsone are recommended for controlling the disease. Other therapeutics include a high dose of cyclophosphamide to fight against immune infections and minimize the risk of adverse effects. [1, 2]

EPIDEMIOLOGY

Pemphigus vulgaris is the most common; around 80% of patients suffer from alone Pemphigus. The occurrence of infection is more common in the Mediterranean and Jewish people. Women are more susceptible as compare to men's. The antigen responsible is (HLA) Human leukocyte antigen; other cases may be disease and medicine induced. [3]

SEVERITY OF DISEASE

PV accompanied by pain in oral mucosa, loss of body weight resulting in malnutrition, skin color changes, mood swings, loss of electrolyte and fluid, and hair loss.
The loss of cutaneous epidermis causes protein loss and bone loss and enhances other complication complications in compromised patients like diabetes and hypertension. There is no treatment available for the disease, but the symptomatic treatment given to control newer blisters' formation and prevent relapse. The complications make this disorder life-threatening, and 70-75% of the remaining untreated cases died within 3-6 years.

In Pemphigus, an autoimmune disorder, the body produces antibodies that destroy skin and oral mucosal cells. It is not contagious, and sometimes it is triggered by Angiotensin-converting enzyme inhibitors and antibiotics like penicillin. [4]

RISK FACTORS AND COMPLICATIONS OF *PEMPHIGUS VULGARIS*

The infected blisters in the mouth are painful, having pus cells and inflamed, red cells on the oral epithelium. The infected blisters are potentially life-threatening if untreated; the medical intervention of dermatologists immediately needed to reduce the severity of the infection. The disease has a stigma of frightening, loneliness, and mental confusion among infected patients. [5] The complications in Pemphigus Patients are explained in Figure. 1

DIAGNOSIS OF PEMPHIGUS PATIENT

- **Immunofluorescence testing along with Biopsy:** Helps to differentiate between oral lesions and Pemphigus lesions.
- **Histopathological studies:** Helps in detecting the size of vacuoles.
- **Blood Test:** To detect antibodies in blood using ELISA (enzyme-linked immune sorbent assay) test
- **Endoscopy:** Chest and Throat to find out blisters [6, 7, 8]

Table 1.Treatment plan for *Pemphigus vulgaris* patients

Category	Drugs	Dose Regimen	Response Time	Treatment Success
Oral Corticosteroids	Prednisolone, Wysolone	45-60mg/day, no improvement, dose increase to 50-60%	Blister cessation occurs within 3 weeks, full healing 8 weeks	29% Patients showed improved symptoms
Pulse Intravenous corticosteroids	Methyl prednisolone, Dexamethasone	250-100mg	Recommended as adjuvant in severe cases,	2-3 months
	Prednisolone/Cyclosporine (5mg/kg)			
Adjuvant Therapy	Azathioprine	1-3mg/kg	Initial 3 weeks, blisters heal	28-45% improves
	Pulse cyclophosphamide, dexamethasone	100mg, increase dose if needed	Healing occurs within 4 weeks	48% patient's condition improved
	Methotrexate	125mg/week	Healing within 6 months	26 % improved
Anti-Inflammatory Drugs	Dapsone	250-100mg	Initial 3 weeks, blisters heal	20-32% condition improved
Modern Medicine	Gold	50mg/week	Healing in 10 weeks	28% improved, 17-35% patients suffer from withdrawal symptoms
	Immuno adsorption: Rituximab & Cyclophosphamide	1000mg in repeated dose 4 weeks	4 weeks	30-50%
	Rituximab	1000mg every 2 week, 375mg every week	Healing requires 3 weeks, stay in hospital	85-90% patients improved
	Cholinergic agonist Pyridostigmine	50-100mg every day	Healing 6 weeks	50% improved condition

***PEMPHIGUS VULGARIS*: LIFESTYLE MODIFICATION, HOME REMEDIES AND SUPPORTIVE CARE**

- The patients advised maintaining regular oral hygiene and skin conditions to improve health.

- Follow Guidelines of Dermatologist: Clean infected wounds regularly, apply OTC medication to minimize the wounds' pain.
- Gentle washing of skin surface: use mild soap or cleansers to clean the body surface, avoid excess rubbing, use warm water if needed
- Consult Dentist for Regular opinion: Maintain oral hygiene, use a soft brush for cleaning teeth and tongue surface, gargle with suitable antiseptics like betadine, rinse mouth 2-3 times a day, apply a local anaesthetic to reduce oral pains like orasore.
- Avoid immediate contact of Heat and U.V. rays: Prevention of the patient infected wound from direct contact of heat, may form new blisters.
- Avoid food items: Do not eat spicy items, may cause pain in the mouth, eat food after cooling at room temperature, wherever necessary, increase vitamin, protein rich foods, and boost fiber content in the diet.
- Home Remedies: Neem poultice on infected wounds for symptomatic relief, vasaka leaf for antiulcer, wound healing effects, Acacia for anti-inflammatory effects.
- Support therapy: Patients may suffer from stress and insomnia; proper counselling and continuous medical inventions help minimize patient suffering.
- Naturopathy and Yoga: Patients advised restricted-calorie diet having watermelon, sprouts, and leafy vegetables, increased electrolyte content in improving quality of life, and reducing comorbidities. Application of Neem oil and Turmeric paste on blisters provides supportive care along with relaxation exercise. [9, 10]

MEDICAL FOCUS ON TREATMENT PLAN

Depending upon the severity of the disease, the medications can be used alone or combined to improve patient conditions (Table 1). [11, 12, 13, 14, 15, 16]. The Schematic representation of P.V. patient therapy explained in Figure 2.

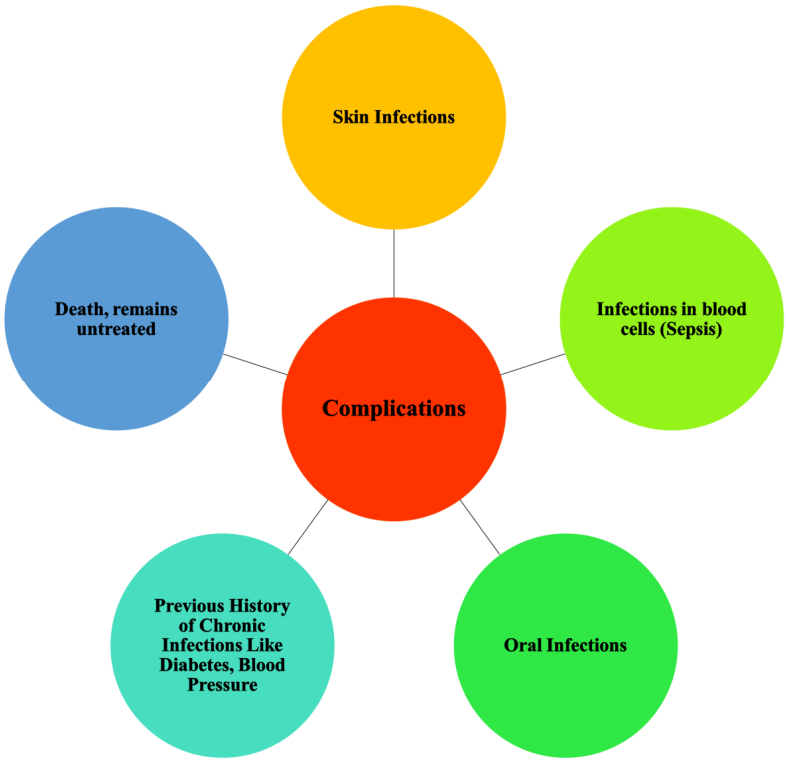

Figure 1. Complications in PV Patient

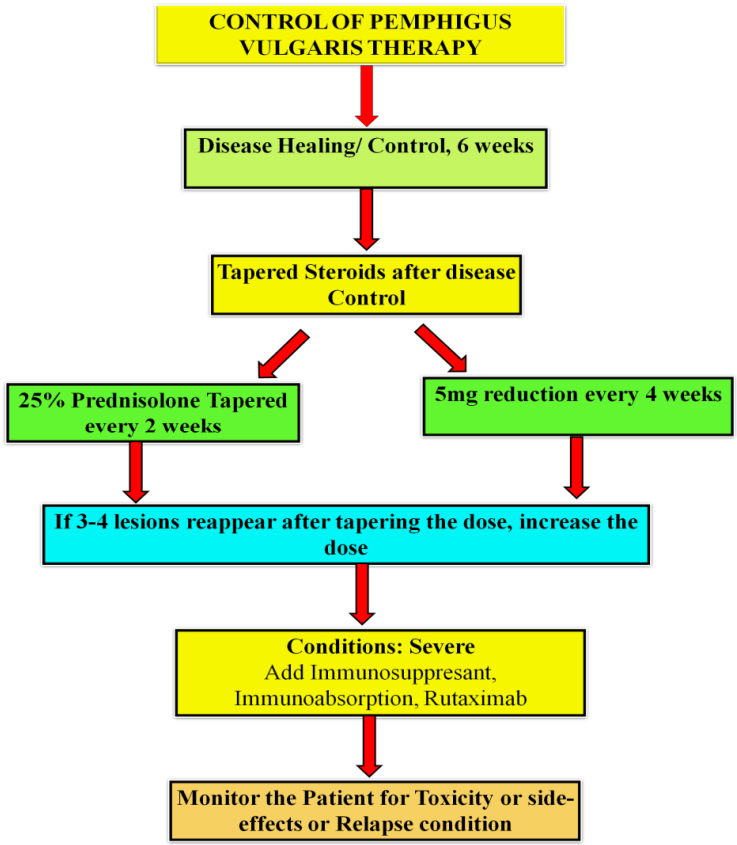

Figure 2. Schematic representation of Pemphigus Vulgaris Therapy

CONCLUSION

Pemphigus vulgaris is a rare disease, life-threatening disorder, and the fatality is steadily increasing with every passing moment. The damage in oral mucosa and the subcutaneous surface has an impact on the quality of life. Although many treatments have tried for the management of pemphigus patients, none of them showed absolute success. The multidisciplinary treatment approaches adopted and recommended for optimized patient care. The early prognosis and detection were vital for achieving adequate scientific measures for the management of Pemphigus. The steroidal therapy and pulse therapy with rituximab proved to be path-breaking in the affected patient's control. The selection of optimal dose using systemic steroids has also shown successful prognosis in the patient and regular exercise and yoga to minimize patients' suffering. The researchers focus on future findings on antigen-based approaches using novel therapies to manage Pemphigus for better health care.

ETHICS COMMITTEE APPROVAL AND PATIENT CONSENT

None

CONFLICT OF INTEREST

The author declared none conflict of interest

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