CASE STUDY

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# A case report on *Pemphigus vulgaris* – A chronic Autoimmune Mucocutaneous Disease

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**ABSTRACT:** *Pemphigus vulgaris* is a chronic autoimmune disease that clinically manifests in the form of intraoral lesions and then it disseminates to other mucous membranes and skin. The disease is scarier and more lethal when it is not diagnosed and treated in its initial stages. Middle-aged and elderly people were most commonly affected by this disease. *Pemphigus vulgaris* is a rare disease with an estimated worldwide annual incidence of 0.1 to 0.5 per 100,000. Here we have presented a case of a 45-year-old woman who is a resident of Madurai with a history of crusted erosions over her face, lips, and trunk. The patient complained that these erosions were painful, causing discomfort in her oral functions. The liver function test report of the patient reveals the declining trend of total protein, albumin, and globulin. From this report, the hypoproteinaemia nature of *Pemphigus vulgaris* is evident. Systemic corticosteroids along with immunosuppressants are the first-line drugs of choice in the therapeutic management of *Pemphigus vulgaris*. In this case, the selected drugs from these two categories were dexamethasone and azathioprine. Recent studies reveal the beneficial effects of rituximab, an anti-CD20 monoclonal antibody that will soon outweigh corticosteroids as first-line agents for the treatment of this disease. Patients require long-term follow-up in order to achieve remission and avoid recurrence.

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## INTRODUCTION:

*Pemphigus vulgaris* is a chronic autoimmune mucocutaneous disease. The disease initially manifests in the form of intraoral lesions; later it spreads to other mucous membranes and the skin [1]. It is significant to diagnose this condition early and should initiate the treatment as quickly as possible since it leads to serious involvement in other mucosal and cutaneous sites and can even be mortal [2]. This disease affects both genders and it is more prevalent in middle-aged and elderly patients. Systemic corticosteroid therapy leads to a dramatic positive prognosis of the condition, but

complications of medical therapy remain a concern [3]. The diagnosis was based on biopsy confirmation of intraepithelial vesicle formation, acantholysis, and the presence of Tzanck cells [4]. *Pemphigus vulgaris* is a rare disease with an estimated worldwide annual incidence of 0.1-0.5 per 100,000. It occurs in all racial and ethnic groups where Ashkenazi Jews have the highest incidence. Even though it is more commonly found in middle-aged people, a few cases have been reported in children also [5].

The pathogenesis of this disease is manifested by the existence of circulating and tissue-bound autoantibodies against the keratinocyte cell surface desmosomal molecules desmoglein 3 (Dsg3) and desmoglein 1 (Dsg1). Dsg3 and Dsg1 are cell adhesion molecules thatbelong to the cadherin superfamily. These autoantibodies cause loss of cell-cell adhesion between epithelial cells, which results in supra-basilar intraepithelial vesicle formation [6]. Presenting a case study of a mid-age woman patient with *Pemphigus Vulgaris* and ruling out the diagnostic and treatment approaches of the concerned case along with pharmacist intervention are the main objectives of this article.

## CASE REPORT:

A 45-year-old woman who is a resident of Madurai, Tamilnadu was referred to the dermatology department with a history of crusted erosions over the face, lips, and trunk. The patient subjectively presented that these erosions were painful, causing discomfort in her oral functions. The patient's family history and personal history were found to be normal with no abnormalities. The patient was a known case of '*Pemphigus Vulgaris*' on treatment. On intraoral examination, crusted erosions were observed. Crusted erosions were also observed on the face, lips, and trunk. No skin lesions were found. The laboratory investigation report in the case report is mentioned in Table 1.

The patient was referred to the clinical department for treatment. Initially, the patient was advised to take the drugs according to the drug chart. The patient's drug chart is provided in Table 2. Lupituss syrup 5 ml thrice daily was prescribed and it was withdrawn after a few days. Sparing a week, Sucrafil O Gel suspension 2 teaspoons thrice daily was added to the prescription, and following that Sebawashes Shampoo, Dilute Hexidine Mouth wash (after every meal), and Liquid Paraffin for Lips were also added to the prescription. Tablet Livogen had been withdrawn from the prescription. Injection Dexamethasone (dose 2 ml) was replaced with a 1.5 ml dose.

**Table 1. Clinical laboratory investigation report.**

| Tests | Test values | Reference value |
|---|---|---|
| Liver function test | | |
| 1) Total Protein | 5.5 g/dl | 6-8 g/dl |
| 2) Albumin | 3.1 g/dl | 3.5-5.5 g/dl |
| 3) Globulin | 2.4 g/dl | 2.5-3.5 g/dl |
| Urinalysis | | |
| 1) WBC | Present | |
| 2) Pus cells | 2-3 | |
| 3) Epithelial cells | 6-8 | |

## DISCUSSION:

*Pemphigus vulgaris* is a potentially life-threatening autoimmune disorder causing painful blisters on the skin and mucous membranes which show a female predominance. The reason for female preponderance is not well understood [7]. Pemphigus was derived from the Greek word, 'pemphix' which literally means blister. Among the three major variants of pemphigus, *pemphigus vulgaris* is the most common one [8].Apart from genetic factors, various environmental factors such as drugs (captopril, penicillamine), infectious conditions (herpes simplex virus, Epstein–Barr virus, etc.), pesticides, ultraviolet radiation, ionization radiation, thermal burns, stress, and food containing an allium, phenol, thiol, or urushiol have been reported as risk factors for the occurrence of *pemphigus vulgaris* [9]. There are threesub-typeswithin *pemphigus vulgaris,* namely, mucosal-dominant type, mucocutaneous type, and cutaneous type. This case of the 45-year-old female patient shows blisters in oral mucosa, face, and trunk regions and it belongs to the mucocutaneoussubtype.

The liver function test report of the patient reveals the declining trend of total protein, albumin, and globulin. From this report, the hypoproteinemia nature of the *pemphigus vulgaris* is evident. The urinalysis of the patient shows the presence of white blood cells, moderately high levels of epithelial cells, and the normal reference range of pus cells. These laboratory investigations are not ample for the confirmation of *pemphigus vulgaris*. The confirmatory test is the detection of autoantibodies in systemic circulation [10]. Some corroborative tests for detection of autoantibodies are Tzanck smear, histopathological examination, direct

immunofluorescence examination, and immunohistochemical examination [11].

Systemic corticosteroids along with immunosuppressants are the first-line drugs of choice in the therapeutic management of *pemphigus vulgaris* [12]. In this case, the selected drugs from these two categories are dexamethasone and azathioprine. The widespread medical literature, however, does not support the use of dexamethasone as a conventional monotherapy. It can be used either as a pulse therapy along with prednisolone, or prednisolone alone can be used to arrive at better outcomes [13]. Azathioprine 50 mg given as a tablet twice daily, is the best adjuvant for the treatment of *pemphigus vulgaris* according to the European Dermatology Forum guidelines [14].

The role of drug treatment other than dexamethasone and azathioprine in this patient can be justified if thorough examinations are done. The use of vaseline petroleum jelly, however, can be helpful to soothe lip ulcerations. Drug-drug interactions between calcium-containing shelcal/ doxycycline and pantoprazole/ ferrous fumarate-containing livogen should be addressed properly in order to evade unnecessary adverse drug effects.

## CONCLUSION:

*Pemphigus vulgaris* is an autoimmune life-threatening disease that causes blisters in both skin and mucous membrane. The management of this disease necessitates good collaboration between dermatologists and dentists. Therapeutic management of this disease is performed with systemic corticosteroids, and immunosuppressive drugs like azathioprine. Recent studies authenticate the advantageous effects of rituximab, an anti-CD20 monoclonal antibody that will soon replace corticosteroids as a first-line agent. It is a chronic debilitating disease with mortality of around 10 %, and septic shock is the main cause of death. Patients need long-term follow-up in order to achieve remission and avoid recurrence.

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