



## A Case on Pemphigus vulgaris with Superadded Infection, Hypoalbuminemia and Hyponatremia

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### ABSTRACT

Pemphigus vulgaris (PV) is a rare autoimmune disease that is characterized by painful blistering and erosion of the skin and mucous membrane. In the case in hand, a female patient of 44 years was presented with complaints of painful vesicles all over the body with erosions, with no history of fever, headache, or cough. The patient had previously been treated for the lesions over the body, considering them to be chicken pox. The patient was admitted to our hospital without any relief. A skin biopsy was done, a specimen was sent for direct immunofluorescence, and the reports were consistent with pemphigus vulgaris. Blood investigation reports revealed the presence of neutrophilic leucocytosis, elevated CRP, hyponatremia, and hypoalbuminemia. The treatment started with IV Dexamethasone (8mg q 8 hours) along with analgesics, IV fluids, and saline soakings. Secondary infections were managed with antibiotics, antibacterial creams, and other supportive medicines. The patient was provided with sodium supplementation and was advised to eat a protein-rich diet. During the hospital stay, the patient developed foul-smelling discharge from vesicles, and antibiotic treatment was escalated, suspecting pseudomonal infections. Antibiotic ear drops were added to the regimen for painful lesions in the ear and mouth. Artificial tears were added for painful eye lesions. The patient was discharged symptomatically better with sterile blood and pus cultures, healthy skin, and a tapered steroid dose. It is important to consider the nutritional requirements of the patient. Due to the painful nature of oral lesions, PV can result in an impaired nutritional status.

**Keywords:** Pemphigus vulgaris, autoimmune disease, Hypoalbuminemia, Hyponatremia, corticosteroids, direct immunofluorescence.

### INTRODUCTION

Pemphigus Vulgaris (PV) is a rare autoimmune disease that results in blisters over the cutaneous as well as the mucosal surfaces<sup>1</sup>. It can occur in both gender, and the mean age of onset is about 50-60 years. The exact etiology of PV is unknown, but patients with a genetic predisposition are more vulnerable. The studies have shown that PV is linked with Human Leukocyte Antigen class II alleles (HLA-II)<sup>2,3</sup>. Diet, stress, viral infections, environmental factors, medication, radiation therapy and allergens can cause immune dysregulations that may lead to PV. The occurrence of PV is also related to ethnicity and geography. Ashkenazi jews are more vulnerable to PV, whereas people in India, Southeast Europe and the Middle East are also at greater risk<sup>4,5</sup>.

PV is caused by autoantibodies, Immunoglobulin G (IgG) that target keratinocyte proteins (desmogleins), wherein there is a loss of keratinocyte-to-keratinocyte adhesion (Acantholysis) by the binding of the circulating IgG<sup>6,7</sup>. The diagnosis of PV is done with a skin biopsy. Direct immunofluorescence (DIF) is the hallmark of diagnosis of PV. PV is associated with 80% of intraoral blisters, 75% of the patients have cutaneous lesions with PV. The Nikolsky sign is seen with PV<sup>8</sup>. The first line treatment for PV is done with systemic corticosteroids, and it can take several weeks to achieve response<sup>9,10</sup>. Tapering of the steroid dose can be done when the symptoms improve<sup>11</sup>. Second line treatment is combination of corticosteroids with azathioprine or mycophenolate mofetil<sup>11</sup>. Third line

treatment includes IV Immunoglobulin (IVIG), cyclophosphamide, dapsone, immunoabsorption and methotrexate<sup>11,12</sup>. Patient education, medication compliance and close follow ups are important for the successful management of Pemphigus Vulgaris.

### Case Presentation

A 44 years old female patient was administered with case of blisters all over the body in the past 2 days with no history of fever, headache and cough. The event started previously with lesions over back and breasts and treatment were taken. The patient again went to hospital with difficulty in swallowing due to aphthous ulcers. Later small lesions were seen visible on the scalp and was treated as seborrheic dermatitis. The lesions began to start appearing on the forehead and stomach and was treated as chicken pox. The patient began to develop swelling tenderness and pain over both foot and had difficulty in walking. The lesions started to appear as blisters and started breaking off. The patient does not have any known co morbidities.

On local examinations, the patient had generalized bullae with erosion and healed vesicles were seen throughout the body. Dermatological opinions were taken and samples were sent for skin biopsy. The specimens were sent for histopathological examinations and direct immunofluorescence and the reports were consistent with pemphigus vulgaris. Lab advises were given to perform LFT, CBC, Electrolyte levels. The reports indicated with



patient having neutrophilic leucocytosis with neutrophil level of 74.7% and lymphocyte level of 17.2%, elevated CRP with 87.3 mg/L. The patient had reduced total protein level with 5.5g/dl and had hypoalbuminemia with albumin levels of 3g/dl. The patient had hyponatremia with sodium levels of 130mEq/L. The patient was administered with sodium supplementation and was advised for a protein rich diet.

The treatment started with corticosteroid which is the first line agent for PV. IV dexamethasone 8mg Q 8hrs was administered. The superadded infections associated with the disease were managed with antibiotics. Tab clindamycin 600mg TID, IV ceftriaxone 2g OD was administered. During the hospital stay she developed foul smelling discharge from vesicles and was escalated to Injection piperacillin + tazobactam suspecting pseudomonas infection. ENT consultation was done with painful lesions in ear and mouth and antibiotic ear drops ciprofloxacin were added to the regimen. Ophthalmology consultation was done for painful eye lesions and was advised for artificial tears. The patient was managed with analgesics tramadol acetaminophen, IV fluids, saline soakings, antibacterial creams and other supportive medicines.

At the time of discharge, the blood and pus cultures were sterile, skin health was gradually restored and steroid dose was tapered. The patient was discharged with medications like ciprofloxacin ear drops, refresh tears carboxy methylcellulose, Tab dexamethasone, clotrimazole mouth paint, mupirocin ointment along with analgesics. The patient was discharged symptomatically better. The patient was advised to contact immediately if any worsening of symptoms is seen.

## DISCUSSION

Pemphigus Vulgaris is an autoimmune disease that can be life threatening which can cause blistering and erosion on the oral and skin surface. Systemic Corticosteroids are the first line agents for the treatment of PV. But corticosteroids are well known for causing osteoporosis and other complications with long term use. Corticosteroids have been reported to cause fracture and osteonecrosis, therefore it is important to assess the fracture risk using the Fracture Risk Assessment tool (FRAX). According to Rai Arpita et al. the important aspect for PV is early diagnosis for which lower doses of the medication can be used<sup>13</sup>. Skin biopsy and Direct immunofluorescence are done for differential diagnosis of pemphigus vulgaris. In a retrospective cohort study done by Kridin et al. in Israel, the survival in patients with PV were lower when compared to the general population<sup>14</sup>. Therefore it is important to follow up and closely monitor the patients with PV.

## CONCLUSION

Pemphigus Vulgaris is a life-threatening autoimmune disease belonging to the pemphigus group of disease of skin and mucosae. The blistering and erosion of skin and mucosa associated with PV can cause pain and functional impairment. Systemic corticosteroids are the first line agents for the treatment of PV. But there are complications associated with long term therapy with steroids such as fractures, osteonecrosis and other complications like hyperglycemia, adrenal suppression, cataracts etc. Therefore, the ultimate goal of treatment should be complete remission with minimal risk associated with treatment for PV.

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## Ethical Approval

Ethical approval is not required at our institution to publish an anonymous case report.

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