

## Case Report

# A Case Study on Pemphigus Vulgaris

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### Abstract:

*Pemphigus vulgaris is a chronic autoimmune mucocutaneous disease that is primitively demonstrated in the form of intraoral lesions extending to other mucous membranes and the skin. The origin of pemphigus vulgaris is unknown, even though the disease has prompted considerable interest. It is marked by the existence of circulating IgG antibodies directed against desmosomes, which are skin components that retain particular layers of skin bound to one another. When desmosomes are attacked, the skin layer separates, and the clinical presentation resembles a blister. Oral lesions are vesicubullous at first but easily rupture, with new bullae attempting to form as the older one's rupture and ulcerate. A biopsy of perilesional tissue and histological and immunostaining examination is required for a correct diagnosis. The current standard of treatment relies mainly on systemic immunosuppression with corticosteroids, azathioprine, or other adjuvants, but newer therapies with possibly fewer adverse effects also seem promising.*

**Keywords:** Pemphigus, Autoimmune, Dental Awareness.

### Introduction:

Pemphigus Vulgaris is an autoimmune intra-epidermal bullous lesion that causes skin and mucous membrane blisters and sores. The term "pemphigus" is derived from the Greek word "pemphix" (blister or bubble), and "vulgaris" origins from the Latin word "vulgaris" (common)<sup>1,2</sup>. Pemphigus is a potentially fatal condition characterized by skin and mucous membrane erosions and blisters<sup>3</sup>. Pemphigus is a rare disease with an incidence rate ranging from 0.5 to 3.2 per 100,000 per year<sup>4</sup>. Pemphigus Vulgaris is a chronic mucocutaneous disease that starts in the oral cavity and later may spread to the skin or other mucous membranes<sup>5</sup>. It is exceptionally vital that oral health professionals have always been capable of recognizing the oral symptoms of Pemphigus vulgaris and treating and referring patients appropriately because it is a life-threatening condition<sup>6</sup>. Pemphigus vulgaris usually occurs in patients between the ages of 30 and 60. The lesions are usually painful. It is characterized by the development of flaccid, easily ruptured intraepithelial bullae on apparently normal skin and mucous membranes. During the course of the disease, the oral cavity is frequently affected. Intraoral lesions can appear in up to 50% of patients without simultaneous involvement of the skin in the disease<sup>5</sup>. The lesions tend to occur most frequently on the buccal and palatal mucosa and the gingiva. This article describes a patient with oral ulceration for two months, pain during swallowing for six months, and blisters on different parts of the body for one month, in whom Pemphigus Vulgaris was diagnosed six

months after the symptoms developed. The laboratory tests were used to confirm the diagnosis and the therapeutic options.

### Case Report:

A 70-year-old male patient was admitted in the Medicine department of the Bangladesh Medical College Hospital with chief complaints of pain during swallowing for six months, oral ulceration for two months, and blisters on various body parts for one month. The present complaint started 6 months back as blisters associated with pain and increased salivation. Gradually blisters started developing on the chest, abdomen. Later it involved the scalp, face, axilla, groin, upper and lower limbs. History of the present illness was itching, increased salivation and hoarseness of voice. He had no significant past illness. He took the following drugs- Inj. ceftriaxone, tab prednisolone, tab methotrexate, tab cefuroxime + clavulanic acid combination, tab metronidazole, tab cefixime, tab folic acid, tab levothyroxine sodium, cap DE lansoprazole, tab bilastine, tab domperidone, vitamin B-Complex, Miconazole 2% oral gel, triamcinolone oral paste, and mupirocin 2% ointment. Patient was a non-smoker, non-alcoholic, did not take betel leaves or betel nuts, did not maintain oral hygiene. There was no history of similar illness in his family. Rest of the family members were in good health. Skin examination revealed multiple erosions and crusts with erythematous base present in scalp, face, chest, back, abdomen, axilla, groin and both limbs. Two flaccid bullae were seen on back of the chest. Bullae contained clear fluid, easily ruptured on slight pressure, and formed an erosive skin lesion. Oral cavity showed erythematous lesions with ulceration present bilaterally on the buccal mucosa extending to retromolar region, right and left borders of the tongue, posterior hard palate and soft palate. There is yellowish color patch seen on dorsal surface of the tongue. No abnormality was detected on hair, nail and conjunctivae.

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Figures: Lesions in Pemphigus Vulgaris

On CBC examination, WBC:  $8.74 \times 10^9/L$ ; N:78%, L:19%, M:2%, E:1%, Hb:13.50 gm/dl; platelets:  $300.8 \times 10^9/l$ ; ESR: 15 mm in 1<sup>st</sup> hour: Creatinine: 1.08 mg/dl. On Serum Electrolytes examination,  $Na^+$ : 135 mmol/l;  $K^+$ :4.71 mmol/l,  $Cl^-$ :101mmol/l;  $HCO_3^-$ : 23mmol/l. Random blood glucose: 4.75mmol/l ; TSH:13.98 IU/ml. On skin biopsy, section showed skin, the epidermis showed intraepidermal bullae in microscopic examination. The dermis showed mild perivascular inflammatory infiltrate. Findings from Direct Immunofluorescence were Cryostat sections showed intracellular IgG (+) deposition in the epidermis in a fishnet-like appearance. No deposition of IgG, IgA or fibrinogen is seen. Based on objective and subjective parameters the patient suffered from Pemphigus Vulgaris along with Hypothyroidism. The patient was treated with the following medications: Infusion 5% DNS (Dextrose 5% + sodium chloride 0.9%), Ringers lactate solution, Tab prednisolone 20mg, Tab Azathioprine 50mg, Tab Esomeprazole 20mg, Cap Evening prime rose oil 50mg, Tab levothyroxine Sodium (50mcg), Tab. Calcium Carbonate + Vitamin D3 (500mg+200IU), Povidone Iodine 1% mouthwash, Triamcinolone Acetonide mouth prep, and Betamethasone 0.1% + Neomycin Sulphate 0.5% ointment.

#### Discussion:

Pemphigus Vulgaris is a classical catastrophically life-threatening, autoimmune, intraepithelial blistering disease of the skin and mucous membranes. There are five major categories of pemphigus: Pemphigus Vulgaris (PV), Pemphigus foliaceus, Paraneoplastic Pemphigus (PNP), drug-induced pemphigus and IgA pemphigus. The oral mucosa is the initial site of involvement in 70%-90% of the cases before the involvement of the skin and other mucosal sites. Nasal, conjunctival, pharyngeal, laryngeal, esophageal and genital mucosa are the other mucosal sites that may be affected<sup>7</sup>. The diagnosis must be confirmed as early as possible. When the skin blisters are present then the picture is distinctive. When oral ulcers indicate the disease but no significant blisters have been noticed, gently stroking the weakened epithelium of mucosa or skin can sometimes cause a vesicle or bulla to appear (Nikolsky's Sign). This disorder of the skin and the mucous membrane is characterized by acantholysis, initiated by circulatory antibodies to intercellular adhesion molecules. Mucocutaneous blisters are induced primarily by autoantibodies against Desmoglein (Dsg) 1 and 3, two proteins of the desmosomes that hold epithelial cells together. Mucosa is only dependent on Dsg3 for its integrity. The relative abundance of the two types of auto-antibody determines the relative effects on skin and mucosa. Circulating antibody can spread through the epithelium and bind to desmosomes and causes detachment of the cells from each other. And the epithelium loses its cohesion and disintegrates. The process starts in the supra-basal and the prickle cells form a vesicle in which fluid accumulates. Gradually vesicles enlarge to become bulla and eventually burst. Epithelial cells that have lost their attachments become rounded and fall into the bullae can be seen in a smear of the fluid in which they are known as

acantholytic or Tzanck cells<sup>8</sup>. As the initial manifestation starts from the oral mucosa so early diagnosis is very important for patient management. Early treatment could prevent the serious involvement of other mucosal and cutaneous sites and fatal complications. A definitive diagnosis of PV is made based on clinical, histopathological, and Direct Immunofluorescence (DIF) findings. DIF shows prominent intercellular deposition of antibodies directed against IgG. The IgG antibodies which target Dsg-3 are deposited at the periphery of the epithelial cells which causes an appearance that has been described as resembling “chicken wire” on viewing the biopsy sample with DIF<sup>9</sup>.

The management of PV can be done through local and systemic corticosteroid therapy. The treatment administration is done in two phases: the loading and maintenance phases. In the loading phase, controlling of the disease is done and in the maintenance phase consolidation and does tapering is done. Local treatments such as ointment and mouthwash are administered alone or combined with systemic treatment<sup>10</sup>. In case of substantial oral mucosal lesion or other skin involvement, systemic corticosteroid therapy is initiated straight away. Depending on the response the dose is gradually decreased. In case of prolong use of corticosteroid, some adjuvants such as Azathioprine is added to the regimen to reduce the complications of long-term corticosteroid therapy<sup>11</sup>.

#### **Conclusion:**

Before the emergence of proper treatment, PV was fatal with a mortality rate of up to 75%.<sup>12</sup> The rate has gradually decreased due to proper diagnosis and management. The mortality rate is now 5%-10% primarily due to the side effect of the therapy as the initial manifestation begins from oral mucosa even before the skin. Oral health professionals and Medicine specialists must be well-versed in the clinical manifestations of pemphigus vulgaris to ensure early diagnosis and treatment, as this results in a better prognosis, lower mortality, and a higher quality of life.

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