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Case Report

A rare case of oral pemphigus vulgaris in a teenage patient: Early diagnosis and successful conservative management

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Abstract

Pemphigus is a chronic, serious potentially life-threatening autoimmune condition which leads to painful blistering and erosion of the skin and mucous membrane. It occurs when the body produces autoantibodies that target desmosomal glycoproteins on the surface of keratinocytes. The immune response disrupts cell-to-cell adhesion, resulting in intraepithelial clefting and blister formation. In pemphigus vulgaris, 80–90% of individuals experience oral lesions, with oral symptoms being the initial indication in approximately 60% of cases. Clinically, affected individuals often present with painful oral ulcers and dysphagia. Accurate diagnosis relies on a combination of clinical examination, histological evaluation, and immunopathological testing. Early identification and management of these oral signs are vital, as timely treatment may prevent skin complications. Initiating therapy during the oral phase often leads to better disease control and increases the likelihood of early remission. This report discusses a 13 years old female patient who presented with recurrent oral ulcers over the inner side of lip, cheek, tongue and swallowing difficulties since 1 year. She was subsequently diagnosed as pemphigus vulgaris. The condition was detected early and managed effectively with lower dose systemic steroids and nonsurgical periodontal therapy within shorter treatment duration. Dental practitioners should be well-informed about the clinical signs and symptoms of pemphigus vulgaris. Early diagnosis and timely intervention play a critical role in controlling disease progression and improving overall prognosis.

Keywords: Pemphigus Vulgaris, Autoimmune condition, Nikolsky sign, Direct immunofluorescence, Desmoglein, Corticosteroid therapy.

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1. Introduction

Autoimmune bullous disorders are conditions caused by autoantibodies that lead to blister formation on the skin and/or mucous membranes.[1] These disorders are classified into two main categories—pemphigus and pemphigoid—based on the depth at which blisters form.[2] Pemphigus represents a group of rare, chronic diseases affecting both skin and mucosa, and is marked by intraepithelial blistering. This occurs due to autoantibodies targeting components of the desmosome-tonofilament complex, which is essential for maintaining keratinocyte adhesion.[3] Several clinical and pathological variants of pemphigus have been identified, including pemphigus vulgaris (PV), IgA pemphigus, pemphigus foliaceus (PF), and paraneoplastic pemphigus (PNP).[4] Among these, pemphigus vulgaris is the most prevalent, representing over 80% of all cases.[5]

Its global incidence ranges from 0.76 to 16 cases per million annually, with higher prevalence noted among individuals of Ashkenazi Jewish and Mediterranean backgrounds.[6] Although it affects both genders, a slight female predominance has been observed.[7] Although uncommon, instances have also been reported in both children and elderly individuals.[8] Clinically, pemphigus vulgaris often begins with blisters that rupture easily, resulting in painful erosions, particularly in the oropharyngeal region, which is commonly the first site of involvement.[9] Pemphigus vulgaris lesions can appear anywhere within the oral cavity, but the buccal mucosa is most frequently affected. Other commonly involved areas include the palate, tongue, and lips, while the gingiva is the least affected. When the gums are involved, desquamative gingivitis is the most typical presentation.[5] In many individuals, oral manifestations are followed by the appearance of skin lesions.[10] The disease mechanism

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involves IgG autoantibodies against desmoglein 3 and desmoglein 1—cadherin family proteins critical for the adhesion of cells in stratified squamous epithelium.[3] Identifying oral pemphigus vulgaris early is crucial, as timely intervention can help prevent the condition from progressing to involve the skin. However, delays in diagnosis exceeding six months are frequently observed.[11] In some cases, the disease may remain confined to the oral cavity for up to one year, which can result in misdiagnosis or inadequate treatment of this potentially life-threatening condition. This article presents a clinical case of oral pemphigus vulgaris along with its successful management.

2. Case Report

A 13-year-old female presented to the Outpatient Department of Periodontics with a primary complaint of recurrent multiple painful oral ulcers over the inner side of lip, cheek and tongue persisting for past one year. She also reported difficulty in swallowing and burning sensation when consuming hot or spicy foods. Symptoms aggravated during stress and menstrual period as reported by the patient.

On examination, crusted and irregular ulcers were observed on the lips. Widespread erosions and ulcerations of varying sizes with irregular edges and surrounding swelling were present on the upper and lower labial gingiva, bilateral buccal mucosa, and ventral surfaces of the tongue. Generalized gingival inflammation was present in both maxillary and mandibular gingiva. An erosive area was noted on palatal aspect of 13, 14, 15, 16 region, thick scrapable coating noted on dorsum of tongue associated with poor oral hygiene **(Figure 1A-1D)**. When tangential pressure was applied to the lesions, peripheral extension was noted, indicating a positive Nikolsky sign.

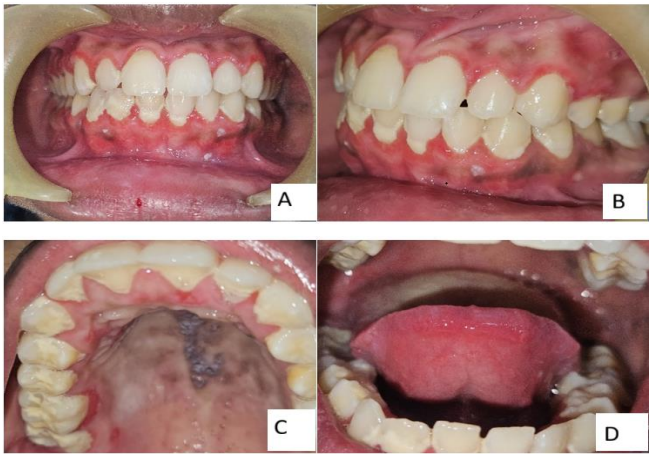

Figure 1: A: Pre-operative gingival lesion; **B:** Pre-operative gingival lesion; **C:** Pre-operative palatal lesion; **D:** Pre-operative tongue lesion

2.1. Diagnosis and treatment

Blood tests and biochemical evaluations were carried out and medications were prescribed for topical application. At 5th day review, patient showed symptomatic improvement and was advised to continue the medications. She was then referred to the Department of Dermatology where she was advised to undergo HSV 1,2 IgG and IgM and the report was negative and the following medications were prescribed for 2 weeks:

1. Syr. Mucaine gel 10ml TDS (Combination of Oxetacine, Aluminium hydroxide and Milk of magnesia)
2. Coolora mouthwash TDS (Benzydamine hydrochloride BP 0.15%w/v)
3. T. Clotroche 10mg TID (Clotrimazole 10mg)

After consultation in the Department of Oral Medicine patient was referred back to Department of Periodontics for oral prophylaxis and gingival biopsy. After obtaining informed consent from her guardian, Professional mechanical plaque removal (PMPR) was performed. The biopsy procedure was carried out under strict aseptic conditions. A 5 mm disposable punch biopsy instrument was used to harvest tissue from clinically normal-appearing gingiva adjacent to the active lesion (perilesional site), which is essential to preserve intact epithelial-connective tissue junctions for accurate immunofluorescence results.

Local anesthesia was achieved using 2% lidocaine with 1:100,000 epinephrine. Care was taken to avoid injecting directly into the biopsy site to prevent tissue distortion. The punch was gently rotated through the mucosa in relation to 22 and 23 until it reached the underlying connective tissue. The specimen was carefully elevated using tissue forceps and excised at the base with iris scissors to avoid mechanical trauma (**Figure 2**).

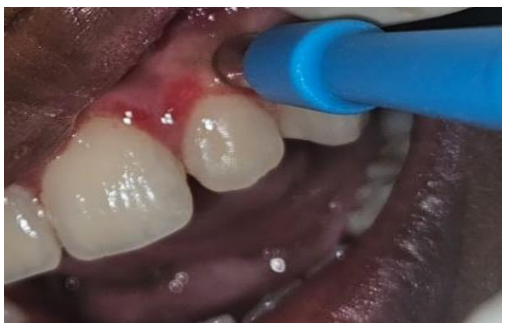

Figure 2: Gingival punch biopsy

The tissue sample was immediately placed in Michel's transport medium (pH 7.0), specifically designed to preserve antigen-antibody complexes for DIF analysis. The specimen was then transported promptly to a specialized immunopathology laboratory at Department of Dermatology, Manipal Medical College under refrigerated conditions. DIF detected circulating autoantibodies against desmoglein 1 and 3 (intercellular staining of epidermis with IgG and C3). The diagnosis of pemphigus vulgaris was confirmed (**Figure 3**).

Initial treatment included:

1. T. Prednisolone 25mg OD x 3 weeks
2. Coolora mouthwash BD (Benzydamine hydrochloride BP 0.15%w/v) x 3 weeks
3. Syrup Potklor 1tsp OD (Potassium chloride 1.5g) x 3 weeks
4. Shelcal 250mg BD (Elemental Calcium 250mg+Vitamin D3 IP 125 IU) x 3 weeks.

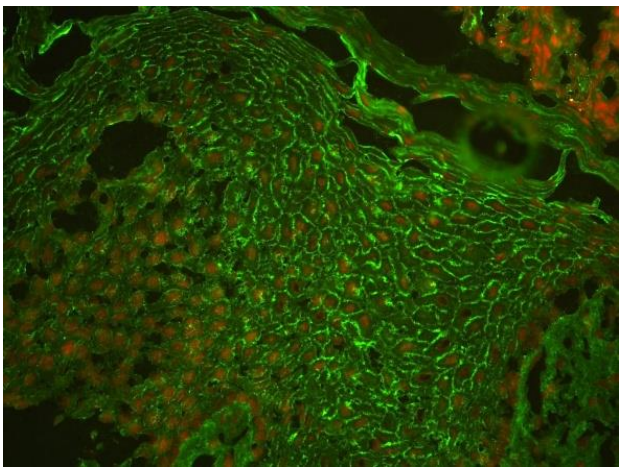

Figure 3: Direct immunofluorescence image

2.2. Follow-up and outcome

At the first follow-up, a marked reduction in ulcerated areas was observed. By the second visit, the lesions had shown significant healing and probiotic supplements (Darolac sachet) were prescribed. Over next two months period, the same medications were continued. After two months, PMPR was repeated to improve oral hygiene, steroid dose tapered to 20mg, iron and folic acid supplements were also prescribed. The patient is currently maintained on 20mg prednisolone. No new lesions have been reported since (**Figure 4**A,B).

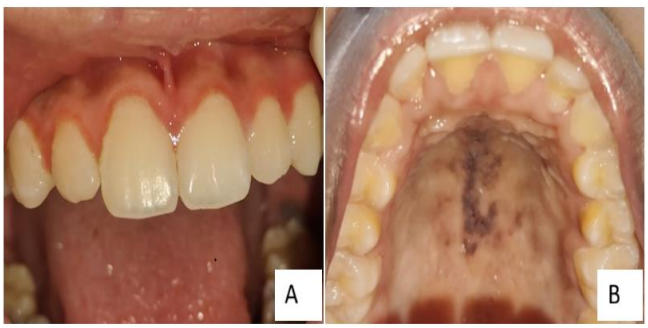

Figure 4: A: Three months post-operative view showing resolution of gingival lesion; **B:** Three months post- operative view showing resolution of palatal lesion

3. Discussion

Pemphigus is a potentially fatal autoimmune condition that affects both the skin and mucous membranes characterized by acantholysis (loss of keratinocyte-keratinocyte adhesion). It is triggered by circulating autoantibodies that target intercellular adhesion proteins. Its hallmark feature is the development of fragile blisters on otherwise normal-looking mucosa or skin. These blisters rupture quickly and spread outward, leaving large exposed areas. A key clinical feature of the disease is a positive Nikolsky sign—where slight tangential pressure applied to the skin or mucosa causes the top layers to shear away, indicating weakened cell adhesion. The primary types of pemphigus include pemphigus vulgaris, pemphigus vegetans, pemphigus foliaceus, pemphigus erythematosus, paraneoplastic pemphigus, and drug-induced pemphigus. Each variant is characterized by autoantibodies targeting specific cell surface antigens, leading to blister formation at varying depths within the epithelial layers. Among these, pemphigus vulgaris is the most prevalent, representing more than 80% of all cases.[12]

Among the various theories proposed to explain the disease mechanism are the "Desmoglein Compensation Theory" and the "Multiple-Hit Hypothesis," both centered around the process of acantholysis, which is the loss of cohesion between keratinocytes.[13] Desmosomes, the structures responsible for intercellular adhesion in the skin, play a critical role in this process. These structures anchossr keratin filaments to the cell membranes of epithelial cells.[14] Two key desmosomal proteins involved in cell adhesion—desmoglein 1 (mainly in the skin) and desmoglein 3 (primarily in the oral mucosa)—are commonly targeted by IgG autoantibodies in pemphigus vulgaris (PV).[13] Pemphigus vulgaris can be further categorized into two subtypes: mucosal-dominant and mucocutaneous-dominant forms based on the specific autoantibodies produced against desmoglein proteins, namely desmoglein 1 (Dsg1) and desmoglein 3 (Dsg3). Patients with a Dsg3-positive/Dsg1-negative (Dsg3+/Dsg1−) antibody profile typically present with mucosal-dominant PV, while those with both Dsg3 and Dsg1 positivity tend to exhibit mucocutaneous involvement. However, the relationship between desmoglein antibody profiles and the clinical manifestation of mucosal or skin lesions can vary significantly among individuals with PV.[15]

Additionally, autoantibodies may also attack desmocollin[16] mitochondrial proteins,[17] and non-desmosomal antigens such as pemphaxin, alpha-9 acetylcholine receptors, and thyroperoxidase[18] leading to the breakdown of epithelial integrity. This antibody binding either triggers the activation of proteolytic enzymes or, as more recent research suggests, directly interferes with the adhesive function of desmoglein. This disruption leads to acantholysis, or the separation of epithelial cells, occurring mainly in the lower layers of the stratum spinosum. As a result, suprabasilar bullae form, progressively affecting larger areas of the skin and mucous membranes. Following the loss of intercellular glycoprotein attachments, spaces are created between epidermal cells, allowing fluid from the dermis to seep in. This results in cavity formation within the epidermis, producing intraepidermal clefts, vesicles, and bullae, characteristic of pemphigus. The detached epithelial cells, known as “Tzanck cells,” become free-floating within these vesicles. These cells

round up and display signs of degeneration, including swollen nuclei and intensified nuclear staining, as seen in cytological preparations.[14]

In approximately 60%–90% of cases, oral lesions are the initial indication of the condition. They typically begin as a classic blister on a non-inflamed surface, which soon ruptures to form a shallow, irregular ulcer. These lesions often appear first in areas prone to friction, such as the buccal mucosa along the biting surfaces, and may later involve the palate, gums, pharynx, larynx, esophagus, and conjunctiva, along with widespread skin involvement.[19] The blisters in the mouth have a fragile outer layer that easily breaks with minor trauma, resulting in persistent, painful ulcers, bleeding sores, and erosions that are slow to heal.[20] The patient also exhibited symptoms of desquamative gingivitis, resulting in considerable discomfort, greater plaque accumulation, and compromised oral hygiene. Taking a thorough patient history is crucial for differentiating pemphigus lesions from those caused by acute viral infections like herpes or erythema multiforme (EM).

Diagnosis of PV includes direct immunofluorescence (DIF), indirect immunofluorescence (IDIF), and the identification of anti-desmoglein antibodies. Ideally, a biopsy should be taken from a newly formed blister (less than 24 hours old) at the edge of the lesion to optimize diagnostic accuracy.[20] Characteristically, histopathology shows suprabasal acantholysis with basal cells appearing like a row of “tombstones,” which helps differentiate PV from other subepithelial blistering disorders such as mucous membrane pemphigoid, bullous lichen planus, and chronic ulcerative stomatitis. For immunofluorescence, a second biopsy is typically performed. DIF will show intercellular deposits of IgG and complement component C3, and sometimes IgA or IgM, displaying a "chicken wire" or "fishnet" pattern.[21] IDIF testing, which uses monkey esophagus as the substrate, can detect circulating antibodies in the blood, although it is less sensitive than DIF and may be used when biopsy isn't possible.[22] Enzyme-linked immunosorbent assay (ELISA) is commonly used to detect Dsg1 and Dsg3 antibodies in serum.Timely diagnosis is essential for effective disease control.

Early treatment allows for the use of lower drug doses over a shorter duration, minimizing complications. The main goals of therapy are to halt disease progression and prevent relapse. Treatment is typically carried out in two main stages: an initial loading phase aimed at achieving disease remission, followed by a maintenance phase, which includes both consolidation and gradual tapering of therapy.[20] Systemic and topical corticosteroids remain the gold standard in treatment, often combined with immunosuppressive agents that help reduce steroid dependency. Based on the patient's response, the medication dose is slowly reduced to the lowest effective amount, ideally taken once daily to limit adverse effects. Systemic prednisolone at a dose of 1–2 mg/kg/day should be started promptly, either alone or in combination with topical agents such as betamethasone, beclometasone, or triamcinolone acetonide. In stubborn or resistant cases, pulsed therapies like intravenous dexamethasone–cyclophosphamide have been employed. Additional treatment may include immunosuppressive agents that reduce steroid dependence, such as azathioprine, mycophenolate mofetil, methotrexate, and rituximab, a monoclonal antibody targeting CD20. In severe or refractory cases, advanced options like plasma exchange or extracorporeal photopheresis may be necessary.[23] Emerging therapies, such as intravenous immunoglobulins (IVIG) and targeted biologics, offer immunomodulatory effects and may be considered alternatives to traditional immunosuppressive treatments. If left untreated, pemphigus can be life-threatening, largely due to widespread skin involvement that compromises the epidermal barrier.

Oral prophylaxis is a vital component in the comprehensive management of pemphigus vulgaris (PV), particularly in patients with oral lesions, as the oral cavity is often the first and most persistent site of disease manifestation. Maintaining good oral hygiene helps reduce plaque-induced inflammation, supports mucosal healing, and prevents secondary infections. In contrast, poor oral hygiene and plaque accumulation can worsen inflammation, act as chronic irritants, and diminish the effectiveness of systemic therapies such as corticosteroids and immunosuppressants. Regular professional cleaning enhances the response to medical treatment and reduces pain, bleeding, and discomfort, ultimately improving the patient’s quality of life. Gentle, atraumatic oral hygiene procedures tailored to the patient's tolerance are recommended to avoid further mucosal trauma. Therefore, a multidisciplinary approach involving both dermatologists and dental professionals is essential, and routine oral prophylaxis should be integrated into the standard treatment protocol for effective long-term management of PV.[24]

4. Conclusion

It is essential for dental professionals to identify the clinical signs of pemphigus vulgaris (PV), as delayed diagnosis and treatment can result in significant morbidity, affecting 5%–10% of cases.[22] Diagnosing and managing pemphigus vulgaris are complex and requires the use of several diagnostic approaches, including clinical evaluation, histopathological analysis, and direct immunofluorescence testing— the gold standard for confirmation of the diagnosis. Establishing a well-structured treatment strategy is essential to enhance the patient’s overall quality of life. The primary objective should be to achieve rapid remission, reduce the frequency of relapses and minimize both hospitalizations and the adverse effects associated with therapy. Regular professional cleaning helps control local irritants, lowers the risk of secondary infections, and improves patient comfort and response to therapy. A multidisciplinary approach,

including dental and medical care, is critical for effective long-term management of PV.

5. Source of Funding

None.

6. Conflict of Interest

None.

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