Case Report
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Oral Pemphigus Vulgaris with Cutaneous Manifestations: A Case Report

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Abstract

Pemphigus is a life threatening mucocutaneous disease characterized by production of auto-antibodies against desmosomal glycoproteins. This results in the formation of intra epithelial blisters. The etiology of this disease remains obscure, although the presence of autoantibodies confirms autoimmune origin. The initial manifestation is frequently the development of intraoral lesions, and later, may involve other mucous membranes and skin. As oral lesions are first to appear, dental professionals must be well acquainted with the clinical presentation of pemphigus vulgaris to ensure timely diagnosis and appropriate treatment. In this article, we report a case of 54- year-old male, who was diagnosed with pemphigus vulgaris.

Keywords: Dental Professionals, Oral Manifestations, Pemphigus Vulgaris, Tzanck Cells

1. Introduction

Pemphigus is an autoimmune, intraepithelial blistering disease of the skin and mucous membranes in which autoantibodies target keratinocyte antigens, leading to acantholysis and blister formation[1]. Pemphigus was first named in 1791 by Wichman and is derived from Greek word "Pemphix" which means bubble or blister[2]. It has 6 variants: Pemphigus Vulgaris (PV), pemphigus vegetans, pemphigus foliaceus, pemphigus erythematosus, Paraneoplastic Pemphigus (PNP) and drug related pemphigus[2]. Pemphigus Vulgaris is the most common type. It has oral lesions as an initial manifestation in 50 percent cases[3].

2. Case Report

A 54-year-old male came with a chief complaint of ulcerations and burning sensation in the oral cavity with blister formation on skin since 1 month. There was no significant personal and family history.

On physical examination, the patient had moderate built. Few skin lesions were present on back and extremities. Dorsal surface of left hand had fluid filled vesicle which was surrounded by erythematous area. (Figure 1) Intra oral examination revealed multiple ulcers on right and left buccal mucosa, dorsal surface of tongue, hard palate and soft palate. Irregular erythematous areas were also seen on labial mucosa and gingiva. (Figure 2) Bleeding was present on manipulation and Nikolsky sign was positive. Based upon history and clinical features a provisional diagnosis of Pemphigus vulgaris was made. Mucous membrane pemphigoid and erosive lichen planus were included in the differential diagnosis.

Routine blood investigations were performed and were found to be within normal limits. Cytological smear was taken from the oral lesion and sent for Papanicolaou Staining (PAP Stain). An incisional biopsy was performed from the perilesional area for histopathological examination. Cytosmear showed desquamated epithelial cells along with group of acantholytic cells (Tzanck cells). Tzanck cells were round in shape having large hyperchromatic nuclei

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Figure 1. Dorsal surface of left-hand showing fluid filled vesicle which is surrounded by erythematous area. Few healed lesions are also evident.



Figure 2. Multiple ulcers seen on dorsal surface of tongue, hard palate and soft palate. Irregular erythematous areas are also seen on labial mucosa.

with prominent nucleoli and thin rim of cytoplasm at the periphery of the cell. (Figure 3) Histopathological examination revealed parakeratinized stratified squamous epithelium exhibiting suprabasal split. This suprabasilar split showed the presence of tzanck cells along with fibrinous exudate and inflammatory cells. Inflammatory cells found in the exudate were chiefly lymphocytes and eosinophils. Underlying connective tissue stroma was also infiltrated by moderate amount of chronic inflammatory cells. (Figure 4) A definitive diagnosis of PV was made based upon the above findings.

Oral predinisolone (60mg/day) was prescribed. Dose of prednisolone was tapered at regular intervals and patient was kept under observation. The patient showed complete remission at 1 month of follow up.



Figure 3. Cytosmear showing cluster of large round cells having hyperchromatic nuclei with prominent nucleoli and thin rim of cytoplasm. (Tzanck cells)

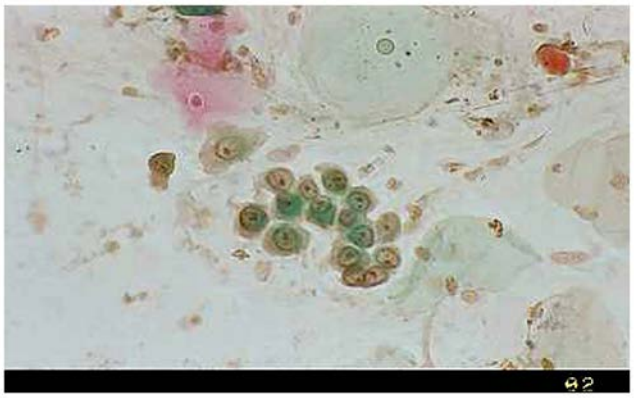


Figure 4. Photomicrograph showing parakeratinized stratified squamous epithelium with suprabasal split. This intraepithelial cleft showed the presence of acantholytic tzanck cells along with fibrinous exudate and inflammatory cells chiefly lymphocytes and eosinophils.

3. Discussion

The term "Vulgaris" in latin means common. PV is the commonest variant of the pemphigus group. It affects 1-5 patients per million populations per year with peak

incidence between fourth and sixth decade of life[3]. It is seen predominantly in females[3].

The blistering which is the characteristic feature of this disease occurs due to production of autoantibodies against desmoglein 1 and 3 which are cell adhesion molecules found in desmosomes. However, in oral mucosa, autoantibodies only against desmoglein 3 leads to suprabasal split. Another theory suggests that antibodies against acetylcholine receptors trigger acantholysis[1]. The IgG antibodies which target Dsg-3 are deposited at the periphery of the epithelial cells, resulting in "chicken wire/fishnet" appearance on viewing the biopsy sample with Direct Immunofluorescence (DIF). Rarely the lesion appears as blisters in the oral cavity. This is because the oral cavity is always subjected to trauma and the blisters formed in pemphigus have thin roof, which easily ruptures giving rise to painful ulcers and erosion[3]. Many times, it could be misdiagnosed as apthous stomatitis, erythema multiforme, or erosive lichen planus because other mucous membrane and skin involvement occurs late in PV. The mainstay of treatment for PV is centered on immunosuppression. Hence, the first line of therapy remains systemic corticosteroids. Treatment is administered as a loading phase, to control the disease, and a maintenance phase, which leads to consolidation and treatment tapering[4]. 0.5–2 mg/kg of prednisone is given as loading dose[5]. Depending on patient's response, the dose is gradually tapered[5]. Prolonged administration of systemic corticosteroids may lead to serious side effects. Adjuvant therapies, including immunosuppressive agents like dapsone, plasmapheresis and intravenous immunoglobulin provides alternate therapeutic options. Dental professionals play a critical role in diagnosing and managing oral lesions of PV. Timely recognition and therapy may prevent further progression of the disease leading to better prognosis and quality of life.

4. References

1. Grando SA. Pemphigus autoimmunity: Hypotheses and realities. Autoimmunity. 2012; 45:7–35. https://doi.org/10.3109/08916934.2011.606444. PMid:21939410. PMCid:PMC3251002
2. Rajendran R. Diseases of skin. Shafer's Textbook of Oral Pathology. 6th ed. Rajendran R, Sivapathasundaram B, editors, Philadelphia: Elsevier; 2009. p. 797-843.
3. Shamim T, Varghese VZ, Shameena PM, Suddha S. Pemphigus Vulgaris in oral cavity. Clinical analyses of 71 cases. Med Oral Pathol Buccal. 2008; 13:2622–26226.
4. Fellner MJ, Sapadin AN. Current therapy of pemphigus vulgaris. Mt Sinai J Med. 2001; 68(4–5):268–278.
5. Toth GG, Jonkman MF. Therapy of pemphigus. Clin Dermatol. 2001; 19(6):761–767 https://doi.org/10.1016/S0738-081X(00)00190-5