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CASE REPORT

PEMPHIGUS VULGARIS OF ORAL CAVITY: A CASE REPORT WITH CLINICAL DIFFERENTIAL DIAGNOSIS

Gurdeep Singh,* Sanya Bhatia., Abhishek Singhvi., Ripon Md. Chowdhury and Ashish Joshi

Vyas Dental College & Hospital, Vyas Institutes of Higher Education (RVS Jodhpur), Near Kudi Haud, NH-62, Pali Road, Jodhpur (Rajasthan)-342005

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ABSTRACT

Pemphigus Vulgaris (PV) is a chronic muco-cutaneous and potentially life threatening autoimmune disease. Its initial manifestations are in the form of intra oral lesions, which are blisters and erosions of mucous membrane and later spread to skin. Dental professionals must be sufficiently familiar with the clinical features of the pemphigus vulgaris to ensure early diagnosis and treatment which will help to determine the prognosis and course of the disease. This article present a case report of uncommon age, unknown etiology, along with differential diagnosis (DD) of PV.

INTRODUCTION

Pemphigus is a chronic inflammatory and potentially life threatening Vesiculobullous autoimmune disease. It can be classified into six types- Vulgaris, Vegetans, Erythematosus, Foliaceus, Paraneoplastic and IgA pemphigus.[1] Two types involve the oral mucosa *i.e.* PV and Pemphigus Vegetans and out of these two, vulgaris is the most prevalent type.[2]The term PV is derived from Latin word *pemphix* (blister or bubble) and*vulgaris*.[3] Annually there are 1-5 cases of pemphigus per 1,00,000 population with male to female ratio 1:2 [5] affecting in between 5th-6th decade of life[4] and disease is common in certain Mediterranean groups, Asians (Indian and Japanese) and Jews, reason being strong genetic predisposition linked to HLA class II alleles, Other etiological initiating factors include drugs (Captopril, Rifampicin, Phenyl Butazone, penicillamine), emotional stress, radiation, viruses (HHV 8).[3,5]

Initially oral lesions are predominant and later they involve the skin. The oral lesions involve buccal, palatal, lingual and labial mucosa. Buccal mucosa is the most common intra oral site. Gingival involvement is rare, but if it occurs Desquamative gingivitis is the commonest manifestation of this disease. [4] Clinically the lesions are characterized by the blisters that rapidly rupture and result in painful erosions.[6]

Histopathology reveals intra epithelial vesicle formation which is due to the acantholysis of epithelial cells along with free floating Tzanck cell which is one of the feature of disease.[1] Confirmatory diagnosis is given by both Direct and Indirect Immunofluorescence which demonstrates deposit of autoantibodies (IgG) throughout the epithelium.[7] The autoantibodies alter the intra cellular links of stratum spinosum which results in destruction of desmosomal junctions.[5] This case report describes the case of a patient complaining episodes of painful ulceration in oral cavity along with considerable discomfort and alter masticatory functions from past 1 year with unknown etiology who was diagnosed as PV.

Case Report

A 21 year old female patient, resident of Jalori gate Jodhpur reported to the Department of Oral Pathology & Microbiology of Vyas dental college & Hospital with chief complaint of painful ulceration on check and tongue since last 1 year. History of present illness reveals that the patient first noticed a small ulcer on right buccal mucosa which was painful. She visited a private dentist, and was advised topical ointments and pain killers. Patient got relief from pain and ulcer. 2 months later patient again noticed an ulcer on lateral surface of tongue for which dentist prescribed systemic steroid (Prednisone10mg) along with topical corticosteroids. There was temporarily relief with the treatment given for 3-4 months, but the patient kept experiencing multiple ulcerations at

various sites in the oral cavity frequently. One week back pain became severe, continuous in nature along with burning sensation and she was not able to masticate food and swallow properly. Her family history was not relevant. On intraoral examination, there was a erosive lesion present on right buccal mucosa between 46 and 47 (FDI tooth numbering system) and similar ulcer was noticed on buccal mucosa opposite right lower canines (Figure 1).

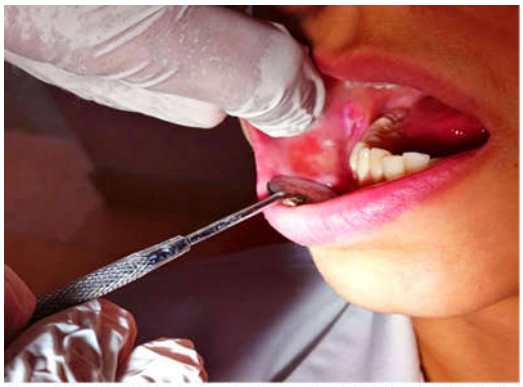

Figure 1: Deep ulceration in plane of occlusion in relation to 46 & 47, and erosive lesion in relation to right lower canine.

Lesions were tender on palpation with no spontaneous bleeding. Two ulcerative lesion were noticed on left lateral border of tongue (Figure 2).

Figure 2: Two well demarcated ulcerative lesion on anterior left lateral border of tongue

According to clinical findings and symptomsdifferential diagnosis Lichen Planus, Lichenoid reaction, RecurrentAphthous Stomatitis, Allergic Stomatitis and mucocutaneous lesions were proposed. Complete blood count (CBC) was conducted and all parameters were in normal range. A cytological smear was prepared followed by Incisional Biopsy on right buccal mucosa after taking consent from the patient. Both the specimens were sent to oral pathology lab for further diagnosis. Histological examination revealed acantholysis with some free floating epithelial cells and suprabasilar epithelial split (Figure 3).

The histopathological investigation suggested mucocutaneous lesion which was confirmed as pemphigus vulgaris on immunofluorescence (Figure 4) using fluorescein label monospecific antisera. Treatment plan was discussed with patient and informed consent was taken which mentioned adverse effects of medication, risk associated with medication and alternative treatment plan.

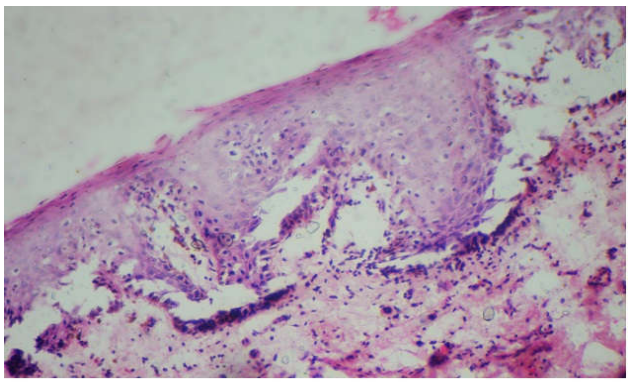

Figure 3: (H & E stain 10 X 10 X = 100 X) showing supra basilar acantholysis

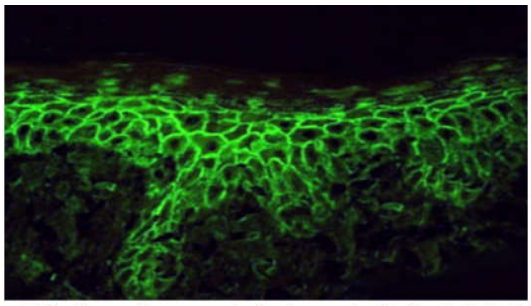

Figure 4: Direct immunofluorescence showing fluorescent antibodies attached to the cell membrane of the epithelium

Treatment included diet modification (soft diet) and soft toothbrush to minimize local trauma. Analgesic mouth wash Benzydamine hydrochloride 0.15% (Tantum oral rinse) prior to eating or brushing along with antiseptic mouthwash Chlorhexidine gluconate 0.2% (Hexidine) twice daily (for 15 days).Topical application of Triamcinolone Acetonide 0.1% dental paste thrice daily over the lesion (Tess 0.1%),with systemic corticosteroids Prednisolone 30mg once daily (Nucort). After consultation with Physician immunosuppressive drug Methotrexate 10mg weekly (Imutrex-10) for 2 months as initial treatment. After 2 months patient was recalled and examined, ulcers have healed with no new ulcer formation. Patient is responding to the treatment and is under observation and was put on prophylactic dose in which topical application of analgesic and mouthwashes was same but reduction in systemic corticosteroids Prednisolone 10mg once daily, Methotrexate 7.5mg weekly for next 1 months was prescribed. Patient was advised to undergo CBC and liver function test (LFT) before the next visit.

DISCUSSION

Many lesions of the oral cavity can possibly cause ulcerations and erosions. Some of these have common clinical appearances posing a great challenge to the clinician in their diagnosis. Many lesions of ulcerative lesions can be initially misdiagnosed and treated incorrectly for months leading to patient discomfort and possible side effects caused by medications. In the present case, a list of DD was considered which were systematically eliminated and hence demands the need to be discussed.

Recurrent aphthous stomatitis (RAS) is a disease synonymous with oral ulcers. Clinically, 3 forms of RAS are seen- Minor, Major and Herpitiform. RAS Minor is the most common form in which the ulcers are less than 1cm in size and heal within 7-14 days without scarring. Traumatic ulcers on the other hand occur as a result of physical, chemical and thermal injuries to oral tissues due to cheek bite, ill-fitting prosthesis, aspirin burns and hot liquid or food. Although the etiology of RAS is

unknown, researchers have suggested several factors that contribute to its occurrence such as haemolyticus streptococcus infection, nutritional deficiency, hormonal imbalance, food allergies and stress. RAS is almost always associated with pain and can occur at any site in the oral cavity except gingival and hard palate. Traumatic ulcers depending on the cause site of occurrence vary but are seen mostly on buccal mucosa.[8,9] A negative history for any traumatic injury, a diffuse erythematous lesion and no response to topical corticosteroid treatment can serve easy differentiating factor from the present case.

Allergic stomatitis may present as minor localised ulcerations to wide diffuse erythematous pruritic lesions. In the oral cavity allergens that are responsible for this type of reaction include dental material like acrylic resins, prosthesis like dentures, metal crowns, other things like latex, resins, hygiene products and food products. [10] In the present case allergic stomatitis can be excluded as a diagnosis as no dental prosthesis and restoration was within the oral cavity on examination and the history for known allergic reaction was not relevant.

Lichen planus is a T-cell mediated chronic inflammatory autoimmune mucocutaneous disease with varying clinical presentations. Reticular pattern is the most common clinical form appearing bilaterally as grey or white velvety lines known as Wickham's straie. Erosive LP, although not as common as reticular form but is more significant as the lesions are symptomatic. Clinically seen as atrophic, erythematous areas sometimes bordered by white striae. [11, 12] In the present case erosions are more widely spread & distributed throughout the oral cavity. Erosive LP cannot be eliminated on clinical basis from pemphigus thus making biopsy mandatory.
Candidiasis is the most common fungal infection affecting oral cavity caused by candida albicans, a normal commensal of the oral cavity. Candida can cause infection ranging from mild superficial mucosal lesion to widespread erosions in immune-compromised patients. A variety of clinical patterns are exhibited in candidiasis.[13] For the present case erythematous type was considered in the DD. Acute form is associated with long term antibiotic therapy & chronic forms include median rhomboid glossitis, angular cheilitis & denture stomatitis. As the etiology of erythematous candidiasis is specific in each form, it is easy to distinguish from mucocutaneous lesions like pemphigus.

Pemphigus is autoimmune disease causing intraepithelial blistering of skin and mucous membrane. In this the antibody IgG is directed against the kertainocyte cell surface molecule desmoglein1 and desmoglein3. The four clinical subsets of pemphigus include pemphigus vulgaris, pemphigus vegetans, pemphigus foliaceous, pemphigus erthematousus, paraneoplastic pemphigus and IgA pemphigus. PV accounts for 80% of all pemphigus cases. It is characterized by vesicles and bullae that rupture rapidly leaving behind raw denuded and ulcerated surface. [1,4,5] In our case the patient had only mucosal lesions with no cutaneous findings. PV typically affects mucous membrane first and is painful with slow healing. Histopathology shows presence of intraepithelial edema and characteristic T-Zanck cells.

Mucous membrane pemphigoid (MMP) is twice as commonly occurring mucocutaneous diseases as compared to PV. In this lesion, the auto antibodies are directed towards the various components of basement membrane leading to sub epithelial bullae formation. Although, blistering of these bullae is less but in the oral cavity chances for rupture is more as compare to any other site. These erosions are painful and present diffusely throughout the mouth. MMP show involvement of other mucosal sites and healing is by scarring. This is also called as cicatricial pemphigoid. [6,11] Our patient did not give any history of blisters but this can be missed making MMP a positive differential diagnosis.

Erythema multiforme is a self-limiting blistering muco-cutaneous disease. It has an acute onset with wide spectrum of severity. Cutaneous involvement is characterized by raised target all bull's eye lesions with concentric rings like appearance. Oral lesion present as large shallow erosion and ulceration which may be extremely painful. The etiology of these lesions is unknown and researchers believe it to be immune mediated.[11] Oral lesion without cutaneous involvement is questionable. However, the wide spread mucosal involvement makes it a potential differential diagnosis.

CONCLUSION

There is an extensive list of possible diseases that may present as an ulcerative lesion in the oral cavity. While most of these can be identified by inspection, diagnosis of some may be more elusive and perplexing. PV is one such lesion that requires prompt diagnosis for timely management as it is a serious and chronic autoimmune disease. Generally PV is treated with topical and systemic corticosteroids. However, the current regime includes the systemic immunosuppressant's adjuvant like methotrexate which should beconsideredwith regular follow up.

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