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Pemphigus Vulgaris: A Case Report

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ABSTRACT: Pemphigus Vulgaris (PV) is an autoimmune, potentially life-threatening disease causing blisters and erosions of the skin and oral mucous membrane.
We present here a case report of Pemphigus Vulgaris of gingiva, oral mucosa and skin in a female patient with age of 42 year old. Perilesional incision was taken and histopathological study was done which shown comprise intraepithelial cleft and Tzanck cells .The oral lesions were treated with Prednisolone 30 mg twice daily and tapered gradually along with topical steroids , benzydamine mouthwash .
KEYWORDS: Desquamative gingivitis, Pemphigus Vulgaris.

I. INTRODUCTION

Pemphigus is a group of autoimmune diseases characterized by formation of intraepithelial bullae in skin and its mucous membrane [1]. and Ig –A. Pemphigus have been described[2] . It most commonly occurs in a 4 th , 5 th and 6th decade of life and more predilection seen in females than males [3] .

A number of dermatological diseases may present gingival manifestations sometimes in the form of desquamative lesions of the gingiva or gingival ulcerations [4]. We report here a case of pemphigus vulgaris which is involving lower lip, gingiva and right retromolar area in adult female patient.

II.CASE REPORT

A 42 year old female patient housewife by occupation reported to the department of Oral Medicine and Radiology with a chief complaint of bleeding of gums and inability to eat food due the lesions at gingiva since 6 months.

Patient was apparently alright before 6 months, then she noticed erosions on her right and left buccal mucosa, gingiva and she also had burning sensation which aggravates on eating hot and spicy food. She was not able to eat solid food and had difficulty in swallowing. She also has bleeding from gums while brushing .Later she noticed itching of the skin. So, she visited the department of oral medicine and radiology.

Patient does not have any type of serious illness ,no history of hospitalization or blood transfusion and no history any drug ,food stuff, pollen grains dust allergies and family history was clear.

Patient had extraction with lower right first molar 1 year ago. Bleeding of gingiva since 6 months, for that she visited to private dental clinic for the same problem .she was on medication for the same from last 1 month.

The patient had a proper oral hygiene habit without any deleterious habits.

On general physical examination, the patient was moderately built and nourished with vital signs within normal limits.

On Intra –oral examination, Desquamation of marginal gingiva at both maxillary and mandibular anterior and posterior region.

There are two main forms of pemphigus vulgaris and pemphigus foliaceus but from two decades non-classical form of pemphigus .They are para-neoplastic pemphigus, pemphigus hertiformis.

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Figure 1: Desquamation of marginal gingiva at both maxillary and mandibular anterior region.

On second visit, multiple erosion seen at lower lip, right buccal mucosa and mandibular right third molar region were inspected. The erosions are shallow reddish in colour surrounded by erythematous border.

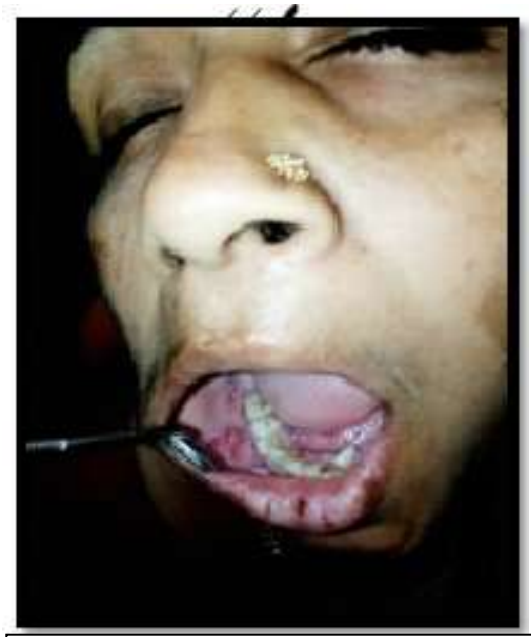

Figure 2: Multiple erosion seen at lower lip, right buccal mucosa, mandibular right 3 rd molar region.

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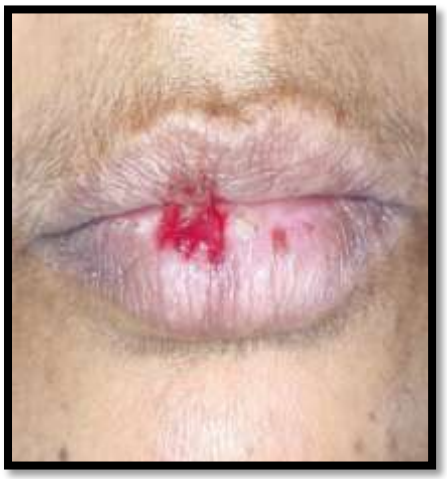

Figure 3: Erosion seen at lower lip.

The size of erosions, at lower lip was 1 ×1.5cm and at right buccal mucosa 1×2 cm and they were tender and bleeds easily on palpation.

Based on clinical signs and symptoms on intra-orally,the provisional diagnosis of desquamative gingivitis was given.

After routine hemogram, urine and blood sugar examination of the patient was taken up for incisional perilesional biopsy with intact epithelium .The sample kept in normal saline and sent for histopathological examination .The sections showed stratified squamous epithelium of oral cavity showing intraepithelial or suprabasal vesicle formation and 'Tzanck cells' within the suprabasilar spilt. Intraepithelial split was also noted .Then diagnosis of Pemphigus Vulgaris was confirmed.

The oral lesions were treated with clobetasol propionate 0.05% ointment, twice daily benzydamine mouthwash also given for burning and Tablet Prednisolone 30mg twice daily and tapered gradually.

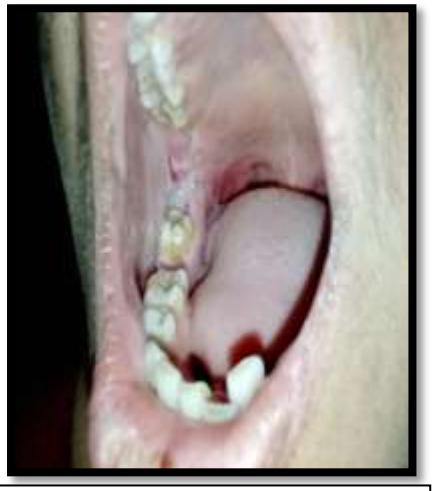

Figure 4: Healing of erosion seen lower right 3 rd molar region.

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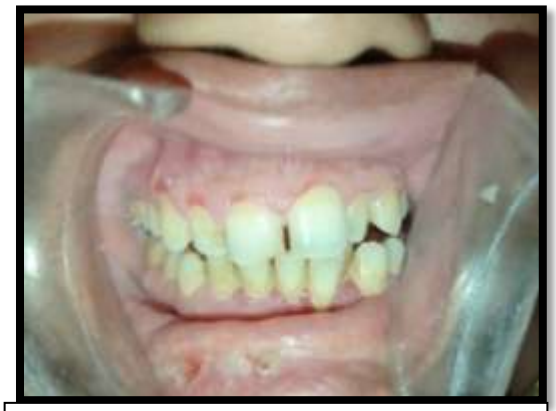

Figure 5: Healing of the lesion on the lower lip.

III.DISCUSSION

The word ‘‘pemphigus’’ has its roots in the Greek ‘pemphix’, which means blister; and ‘‘Vulgaris’’ is derived from Latin word which means ‘common’.

Pemphigus is defined as a group of disorders with autoimmune etiology which could be life threatening and clinical manifestations are mainly epithelial blistering affecting cutaneous and/or mucosal surfaces including mucosa of the mouth, nose, conjunctiva, genitals, esophagus, pharynx, and larynx.

The classic lesion of pemphigus is a thin – walled bulla arising on otherwise normal skin or mucosa, which rapidly breaks and continues to extend peripherally, eventually leaving large denuded areas. This disease also exhibits positive “Nikolsky’s sign” – the ability to induce peripheral extension of a blister and/or removal of epidermis as a consequence of applying tangential pressure with a finger or thumb to the affected skin, peri-lesional skin, or normal skin in patients affected or suspected with pemphigus[5].

Pemphigus vulgaris pathophysiology seems to result from the deleterious actions of circulating auto-Abs, which are directed against desmosomal components, primarily desmoglein (Dsg3 and Dsg1), and lead to the loss of keratinocyte cell-cell adhesion within the epidermis, a phenomenon known as acantholysis [6].

In addition, blister formation in PV was suggested to result from increased secretion of pro-inflammatory mediators or other mechanisms, such as activation of specific muscarinic receptors expressed by keratinocytes, abnormalities in intercellular signaling or activation of apoptosis [7-8].

To initiate and perpetuate the disease process environmental factors could be required. Thus, drugs, viral infections (herpes simplex virus), physical agents, contact allergens, vaccinations, dietary factors, and psychological stressors have been implicated in the disease [9].

For the diagnosis of Pemphigus Vulgaris direct immunofluorescence (DIF) microscopy is considered as gold standard, which can detect tissue-bound auto antibodies. In pemphigus, DIF microscopy reveals intercellular binding of Ig G and/or C3 within the epidermis and/or epithelium [10].

To monitor and know the disease activity Enzyme-linked immunosorbent assays (ELISA) is useful.

Early diagnosis of the disease is very important in the treatment of pemphigus Vulgaris because without treatment, PV has a mortality rate ranging from 60 to 90% [11].

Systemic corticosteroids and adjuvant therapies have reduced the mortality rate of PV patients to approximately 10%. Corticosteroids suppress the patient immune system and prevent the production of pathogens.As there is significant decrease of pathogens in the skin and serum of patients, which clinically corresponds to the cessation of new vesicle formation and the maintenance of remission [12].

When corticosteroids are administered over long periods of time, side effects such as diabetes, hypertension or osteoporosis may occur. It is preferable to combine systemic corticosteroids with an immunosuppressive agent, in order to avoid side effects [13,14].

The first-line adjunctive immunosuppressive therapies in pemphigus are azathioprine and mycophenolate mofetil .These both drugs present steroid-sparing effect.

Azathioprine demonstrated a steroid –sparing effect when compared to prednisolone alone [15].ˢwhile mycophenolate mofetil has shown faster and more-durable treatment responses than placebo when added to prednisone regimens[16].

Intravenous immunoglobulins (IVIG) are considered as an adjuvant therapy in pemphigus [17].They neutralizes and slow down the production of circulating pemphigus antibodies [18].

Conventional immunosuppressants IVIG are effective at a dosage of 0.4 g/kg/day for 5 days, used as an adjunct to corticosteroid therapy once per month [19].

In addition, this therapy is considered the safest adjuvant treatment for pemphigus in pregnant women [20].

Plasmapheresis involves plasma exchange with albumin or fresh frozen plasma to remove serum Ig G antibodies. This procedure has been used to treat a variety of antibody mediated autoimmune disorders. This therapeutic alternative is generally used in patients who experience side effects in cortisone therapy, pregnant women or those who do not respond to conventional therapies [21].

IV. CONCLUSION

Being a dental professionals we should know the clinical features of pemphigus vulgaris for the accurate diagnosis of disease so that we could start the treatment at an early stage to minimize hospitalization , side effects due to steroids and the mortality rate associated with the disease .

REFERENCES

[1]. Rath SK, Reenesh M. Gingival pemphigus vulgaris preceding cutaneous lesion: A rare case report. J Indian Soc Periodontol 2012;16:588-91.
[2]. Pemphigus Vulgaris a d mucous membrane pemphigoid: Update on etiopathogenesis ,oral manifestations and management .J Clin Exp Dent 2011;3: e 246 -50 .
[3]. Lever WF and Talbott JH: Pemphigus: A historical study. Arch Derm Syphilol 46: 800-823, 1942.
[4]. Scully C, Challacombe SJ. Pemphigus vulgaris: Update on etiopathogenesis, oral manifestations, and management. Crit Rev Oral Biol Med 2002;13:397-408.
[5]. Salmanpour R, Shahkar H, Namazi MR, Rahman-Shenas MR. Epidemiology of pemphigus in South-Western Iran: A 10-year retrospective study (1991-2000). Int J Dermatol 2006;45:103-5.
[6]. Banerjee I, Bhowmik B, Maji A, Sinha R. Pemphigus vulgaris – A report of three cases and review of literature. J Family Med Prim Care 2018;7:1109-12.
[7]. Stanley JR and Amagai M: Pemphigus, bullous impetigo, and the staphylococcal scalded-skin syndrome. N Engl J Med 355: 1800-1810, 2006.
[8]. Grando SA: Pemphigus autoimmunity: Hypotheses and realities. Autoimmunity 45: 7-35, 2012.
[9]. Ahmed AR, Carrozzo M, Caux F, Cirillo N, Dmochowski M, Alonso AE, Gniadecki R, Hertl M, López-Zabalza MJ, Lotti R, et al: Monopathogenic vs multipathogenic explanations of pemphigus pathophysiology. Exp Dermatol 25: 839-846, 2016.
[10]. Mao X, Sano Y, Park JM and Payne AS: p38 MAPK activation is downstream of the loss of intercellular adhesion in pemphigus vulgaris. J Biol Chem 286: 1283-1291, 2011.
[11]. Shoimer I, Wong RX and Mydlarski PR: Pemphigus Vulgaris. In: Autoimmune Bullous Diseases. Sami N (ed). Springer International Publishing, pp1-18, 2016.
[12]. Bystryn JC and Rudolph JL: Pemphigus. Lancet 366: 61-73, 2005.
[13]. Kridin K: Emerging treatment options for the management of pemphigus vulgaris. Ther Clin Risk Manag 14: 757-778, 2018.
[14]. Harman KE, Brown D, Exton LS, Groves RW, Hampton PJ, Mohd Mustapa MF, Setterfield JF and Yesudian PD: British Association of Dermatologists' guidelines for the management of pemphigus vulgaris. Br J Dermatol 177: 1170-1201, 2017.
[15]. Caplan A, Fett N, Rosenbach M, Werth VP and Micheletti RG: Prevention and management of glucocorticoid-induced side effects: A comprehensive review: A review of glucocorticoid pharmacology and bone health. J Am Acad Dermatol 76: 1-9, 2017.
[16]. Almugairen N, Hospital V, Bedane C, Duvert-Lehembre S, PicardD, Tronquoy AF, Houivet E, D'incan M and Joly P: Assessment of the rate of long-term complete remission off therapy in patients with pemphigus treated with different regimens including medium- and high-dose corticosteroids. J Am Acad Dermatol 69: 583-588, 2013.
[17]. Beissert S, Mimouni D, Kanwar AJ, Solomons N, Kalia V and Anhalt GJ:

Treating pemphigus vulgaris with prednisone and mycophenolate mofetil: A multicenter, randomized, placebocontrolled trial. J Invest Dermatol 130: 2041-2048, 2010.

[18]. Porro AM, Hans Filho G and Santi GH: Consensus on the treatment of autoimmune bullous dermatoses: Pemphigus vulgaris and pemphigus foliaceus - Brazilian Society of Dermatology. An Bras Dermatol 94 (Suppl 1): 20-32, 2019.

[19]. Tavakolpour S, Mirsafaei HS and Delshad S: Management of pemphigus disease in pregnancy. Am J Reprod Immunol 77: e12601, 2017.

[20]. Harman KE, Brown D, Exton LS, Groves RW, Hampton PJ, Mohd Mustapa MF, Setterfield JF and Yesudian PD: British Association of Dermatologists' guidelines for the management of pemphigus vulgaris. Br J Dermatol 177: 1170-1201, 2017.

[21]. Guillaume JC, Roujeau JC, Morel P, Doutre MS, Guillot B, Lambert D, Lauret P, Lorette G, Prigent F, Triller R, et al: Controlled study of plasma exchange in pemphigus. Arch Dermatol 124: 1659-1663, 1988.