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Pemphigus vulgaris – Value of Diagnosis in Dental Setup

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KEYWORDS

Autoantibodies, Autoimmune, Bullae, Corticosteroids.

A B S T R A C T

Pemphigus is a group of potentially life-threatening autoimmune diseases characterized by cutaneous and/or mucosal blistering. *Pemphigus vulgaris* is the most common variant and is characterized by circulating IgG antibodies directed against desmoglein 3 with about half the patients also having desmoglein 1 autoantibodies. Involvement of the oral mucosa is common and in most cases precedes skin lesions. Most patients are initially misdiagnosed and improperly treated for months or even years. Dental professionals must be sufficiently familiar with the clinical manifestations of *Pemphigus vulgaris* to ensure early diagnosis and treatment, since this in turn determines the prognosis and course of the disease. Here we report a case series of 3 *Pemphigus vulgaris* cases with preceeding oral manifestations before skin lesions along with successful treatment with steroids as well as steroid sparing therapies.

Introduction

Pemphigus is a group of potentially life threatening autoimmune diseases characterized by cutaneous and/or mucosal blistering, *Pemphigus vulgaris* (PV) being most common variant, is characterized by circulating IgG antibodies directed against desmoglein 3 (Dsg 3), with about half the patients also having Dsg 1 autoantibodies (Harman *et al.,* 2003; Scully and Mignogna, 2008a, Solanki *et al.,* 2012). There is fairly strong genetic background to pemphigus linkage to HLA class II alleles and ethnic groups such as Ashkenazi Jews and those of Mediterranean and Indian origin are commonly affected. Pemphigus affects 0.1-/0.5 patients per 100000 populations per year (Black *et al.,* 2005). The peak incidence of PV occurs between the 4th and 6th decades of life with male to female ratio of 1:2 (Tamgadge *et al.,* 2011; Shamin *et al.,* 2008).

Clinically, the oral lesions are characterized by blisters that rapidly rupture, resulting in painful erosions. Any area in the oral cavity can be involved, the soft palate, buccal mucosa and lips are predominantly affected. (Tamgadge *et al.,* 2011; Shamin *et al.,* 2008) Apart from oral mucosa PV may also involve other mucosal surfaces like larynx, pharynx, gastrointestinal, eye, and genital mucosa. Cutaneous lesions may be isolated or generalized flaccid blisters that express citrine or hemorrhagic liquid when ruptured. Cutaneous lesions when ruptured leave extensive erosive areas (Ferreira *et al.,* 2013). The Nikolky's sign can be positive. The PV oral lesions may precede the cutaneous lesions by several months (Mignogna *et al.,* 2000; Weinberg *et al.,* 1997).

The diagnosis of PV can be made based on the clinical and histopathological characteristics of the disease (Ferreira *et al.,* 2013). Histologically the hallmark of PV is acantholysis. Cytological examination of freshly formed bullae is widely used in the diagnosis of PV. Although the presence of Tzanck cells is an important finding, there is no absolute specificity for pemphigus (Mignogna *et al.,* 2000). Demonstration of immnuoglobulins IgG and complement in intercellular space by direct immunofluroscence (DIF) is a very reliable test for PV. Indirect immunofluroscence (IIF) studies helps to detect circulating autoantibodies in the patient's serum (Shamin *et al.,* 2008). Enzyme linked immunosorbant assay (ELISA) is available for direct measurement of Dsg 1 and Dsg 3 antibodies in serum which offer advantages over IIF (Harman *et al.,* 2003).

The mortality rate associated with PV was 75% on average before the introduction of corticosteroids (Harman *et al.,* 2003) mainly from dehydration or secondary systemic infections (Black *et al.,* 2005) in the early 1950s (Harman *et al.,* 2003). Therefore early diagnosis and treatment are essential (Weinberg *et al.,* 1997). As the oral mucosa is often the first affected site in most of the cases, dental professionals plays a critical role in diagnosing and managing oral lesions of PV (Shamin *et al.,* 2008).

The treatment of PV consists of immunosuppressive therapy with systemic corticosteroids, with azathioprine or other adjuvant or alternatives. The newer therapies have potentially fewer adverse effects and also appear promising (Black *et al.,* 2005; Ferreira *et al.,* 2013).

Case report 1

A female patient aged 45 years reported to outpatient department of our college. Patient presented with a chief complaint of burning sensation in her mouth from past 3 months, inability to open her mouth since past 1 month. Also complaints of blisters in her body from past 15 days. Patient first noticed itching and burning sensation in her mouth 3 months ago. After about 1 month she noticed appearance of wounds on right cheek mucosa of her mouth. She reported that these wounds gradually increased to involve her entire mouth. She also reported that whenever she spits there was a shedding of tissue tags from her saliva. She also suffered from other symptoms like difficulty in eating and speaking. She noticed development of multiple blisters on abdomen, back, upper and lower limbs; 15 days before she reported to us. Past dental history revealed that she received a treatment from an oral surgeon. She was provisionally diagnosed to have chronic generalized periodontitis/ leukoplakia/lichen planus. But her condition did not improve even after the treatment. Medical and family histories were non contributory. She was a

chronic pan chewer from 8-10 years, she use to keep the quid equally on both sides of the oral cavity for 5 minutes. General physical examination revealed multiple blisters, present on her abdomen, back, upper and lower limbs. Bilateral submandibular lymphadenopathy was present. On oral examination generalized gingival inflammation and generalized bleeding and probing were present. On inspection diffuse erythematous areas, irregular in shape, size and margins were present involving right and left buccal mucosae (Figure 1 and 2), maxillary and mandibular labial mucosae, soft palate, ventral, dorsum and lateral aspects of the tongue (Figure 3) and gingivae. Irregular epithelial tissue tags surrounded the erosive areas; few areas of sloughing were also seen on buccal mucosae (Figure 4). On palpation generalized tenderness of mucosae was elicited, epithelial tags were scarpable, bleeding on manipulation was present, mouth opening was reduced to 22 mm. On her skin multiple well circumscribed intact bullae measuring about 2-3 mm to 1 cm large were present on the abdomen, back, upper and lower limbs (Figure 5 and 6). Few ruptured bullae were seen on the back with irregular erythematous area (Figure 7). On palpation the bullae were non tender, tense and flaccid. On rupturing the bullae a clear serous discharge were seen. However positive Nikolky's sign could not be elicited with skin lesions. But intraorally superficial epithelium got separated on application of little pressure. Based on history and thorough clinical examination a provisional diagnosis of *Pemphigus vulgaris* was made. A list of differential diagnosis included bullous and mucosal pemphigoid, epidermolysis aquisita, erythema multiforme and bullous/erosive lichen planus. Patient was subjected to intraoral biopsy, Tzanck test and skin biopsy which yielded Tzanck cells and intraepithelial and intraepidermal suprabasal bulla (Figure 8). The diagnosis of PV was confirmed following histopathology. Patient management was done in co-ordination with a dermatologist. Patient received tab Prednisolone (20 mg) 3 times a day for 1 month in tapering dosages, Silver Sulphadiazine cream (1%), Chlorhexidine (0.2%) mouth wash three applications for 2 weeks, tab Cetrizine (5 mg) + Pseudoephedrine (120 mg) twice a day for 2 weeks. Patient was recalled after 1 week for evaluation and for tapering of dosages.

1st Follow up

Patient did not report after 1 week, but reported after 4 months with itching sensation in her mouth. She also gave history of shedding of tissue tags from her mouth. The bullae over abdomen were completely healed (Figure 9), bullae over the back were partially healed with hypopigmentation (Figure 10), erosions over the tongue and labial mucosa were completely healed (Figure 11), erosion on right buccal mucosa was partially healed (Figure 12) and erosions on left buccal mucosa were completely healed (Figure 13). The dosages of tab Prednisolone were tapered. Traimcenelone acetonide (0.1%) in orabase was prescribed while other medications were stopped and patient was recalled after 1 week.

2nd Follow up

But patient reported after 2 months with continuous itching sensation. She presented with single intact vesicle measuring 2-3 mm in size in left retromolar mucosa (Figure 14). Skin lesions were completely healed (Figure 15). Later on patient reported whenever itching sensation reoccurred. She was asked to continue steroid treatment in lower doses for about 2 weeks. Thereafter patient was

maintained on topical steroids. No reoccurrence of lesions were reported on skin as well as on oral mucosa.

Case report 2

A 48 year old female patient reported to our outpatient department with a chief complaint of recurrent ulcerations in mouth since 6 months with skin blisters since 5 months. Patient reported multiple recurrent ulceration in her cheek mucosa bilaterally and at lower labial mucosa which occurred suddenly and then rapidly increased in frequency of occurrence within 2 months of onset with initially being less in number, frequency, size and associated symptoms. Ulcers occur within every 10-15 days and persist for a week and ruptures without any intervention. Patient also reported burning sensation on consuming spicy food. She reported severe skin blisters and scabs on various areas of her body. After a month of onset of mucosal lesions she consulted a local physician and was given a topical ointment. Her past dental, medical, family and habit histories were non contributory. Physical examination of patient revealed multiple irregular thick macules with scabbing which were evident on front, nape and right lateral neck surface extending till lumbar region of back (Figure 16 and 17). A solitary lesion was present on left breast (Figure 18). Lesions were of varying sizes and were scattered with ruptured vesicles with moistened surface as a result of release of a clear fluid from vesicles. There was evidence of solitary grayish black lesion on right hypochondriac-epigastric junction having a single large intact bulla measuring about 4x5 cm in size with 2-3 intact vesicles (Figure 19). Overlying skin surface appeared lacerated, erythematous and was covered by a greenish pseudomembranous slough and appeared as a scab with moistened surface (Figure 19). All the lesions on the body appeared rough surfaced, encrusted and pigmented which were associated with itching. There was negative Nikolky's sign but positive Asbo Hansen's sign on skin lesions. (Figure 20)

On intraoral examination, multiple ill defined erosive lesions were evident with buccal mucosae bilaterally (Figure 21 and 22) and lower labial mucosa (Figure 23) with single erosive lesion on upper labial mucosa in relation to 22, 23 teeth region (Figure 24). Lesions were covered by blisters in some areas and ruptured bullae in other areas having moistened surface. Periphery of ruptured bullae showed tissue tags that could be peeled off with gauze (Figure 25). There was also evidence of multiple ill defined erosive lesions on hard and soft palate junction (Figure 26). On the basis of history and clinical examination a provisional diagnosis of PV was given and list of differential diagnosis included bullous and mucosal pemphigoid, erythema multiforme and bullous/erosive lichen planus. Skin and intraoral biopsy was performed revealed acantholytic Tzanck cells with suprabasilar split confirming the diagnosis of PV (Figure 27). Patient management was done in consent with dermatologist and was prescribed tab Wysolone (20 mg) 2 times a day for 10 days which was to be tapered to (20) mg once a day for 10 days, Tab Endoxan (cyclophosphamide) (50 mg) once a day for 20 days, tab Levasiz (levocetrizine) once at night (5 mg) for 20 days, Tenovate (clobetasol proprionate) gel twice daily on encrustation for 1 month, Topinate gel for mucosal lesions twice daily for 1 month. Patient was recalled after 10 days for evaluation.

1st Follow up

Complete healing of intraoral lesions was found except a pseudomembranous tissue

tag on lower labial mucosa (Figure 28, 29). Skin lesions were partially healed with dry scaly papules with no new bullae formation (Figure 30, 31, 32). Patient was again recalled after 10 days.

2nd Follow up

Further healing of all lesions with no new bullae formation was evident. Patient was advised to maintain body and intraoral hygiene and to continue same treatment regimen for 1 month.

Case report 3

A 45 year old female reported to our outpatient department with a chief complaint of multiple ulcerations in her mouth since 6 months. Patient reported a single ulcer on palate which was followed by eruption of multiple ulcers involving the entire oral cavity which increased in size gradually. These lesions were preceded by fluid filled blisters and were associated with burning sensation, pain and difficulty in speaking, eating and swallowing due to involvement of esophageal mucosa. She also reported formation of new multiple fluid filled blisters in mouth once in every 15 days. They ruptured following trauma from mastication leaving behind irregular tissue tags, visible in saliva while spitting. Patient consulted number of doctors for the same but the condition remained undiagnosed. There were no relevant dental, medical, family and habit histories. Patient's physical examination did not reveal any significant findings or skin involvement. On intraoral examination, multiple ill defined erosive lesions were seen bilaterally with buccal mucosae, floor of the mouth, dorsum and ventral surface of tongue, hard and soft palate junction and attached gingiva of variable sizes. Tissue tags around ruptured bullae with sloughing were evident with surface of lesions on left buccal mucosa and ventral surface of tongue (Figure 33, 34, 35). On the basis of history and clinical examination a provisional diagnosis of PV was made. The list of differential diagnosis included mucous membrane pemphigoid, erosive/bullous lichen planus and recurrent major apthous ulcers. Patient was subjected to intraoral biopsy and direct immunofluroscence examination. Histopathological report was negative for pemphigus, DIF revealed IgG antibodies and C3 complement factor confirming the diagnosis of PV (Figure 36). The patient's treatment included tab Prednisolone (80) mg in 3 divided doses (40, 20,20 mg) for 15 days, tab Aciloc (150 mg) 2 times a day for 7 days, Tantum mouth rinse to be used 3 times a day for 7 days. Patient was recalled for evaluation after 15 days and patient was told to notice the new bullae formation.

1st Follow up

Patient reported 1-2 new bullae formation which was ruptured within 2 days. Lesions were partially healed in hard and soft palate junction, dorsum and ventral surface of tongue, left buccal mucosa and floor of the mouth; with completely healed lesions with right buccal mucosa, attached gingiva and hard palate. Prednisolone dosage was tapered to 60 mg per day in 3 divided doses (40, 10, 10 mg) for 15 days along with other same medications. Patient was recalled after 15 days.

2nd Follow up

Patient again reported 1-2 new bullae formation which used was ruptured within 2 days. All intraoral lesions were completely healed except the lesions of hard and soft palate junction and dorsum of tongue. Prednisolone dosage was again tapered to 40 mg per day in two divided doses (20, 20 mg)

for 15 days along with other medications and was again recalled after 15 days.

3rd Follow up

All lesions were completely healed except the lesions on hard and soft palate junction.

Results and Discussion

Bullous lesions of the skin and oral mucosa were traditionally classified according to their clinical and histological patterns. In the '60s and '70s, it was demonstrated that several of these conditions were autoimmune diseases and the autoantigen localization was described by immunohistology. Many of these bullous lesions which have clinical features similar to those of autoimmune diseases, are actually caused by mutations in genes coding for proteins against which patients with autoimmune disease produce antibodies which includes pemphigus and other mucocutaneous autoimmune diseases (Dabelsteen, 1998). The word pemphigus originates from Greek word Pemphix, which translates in to blister or bubble which is associated with malignancy of lymphoid tissue. Pemphigus is a serious autoimmune disorder with mucocutaneous manifestations characterized by development of blisters on the skin and/or mucosal membrane. Six types of pemphigus has been established – Vulgaris, Vegetans, Erythematosus, Foliaceous, Paraneoplastic and pemphigus IgA (Ali *et al.,* 2011). *Pemphigus vulgaris* accounts for about two-thirds of all pemphigus cases and probably constitutes the most common bullous autoimmune disorder (Beissert *et al.,* 2006). The overall incidence of pemphigus worldwide is estimated at 0.076 to 5,100,000 persons per year (Santoro *et al.,* 2013) and in India 4.4 per million population per year which is found to be higher than available data from Germany and France (Kanwar *et al.,* 2011). The incidence of PV is higher in women (Santoro *et al.,* 2013) with peak incidence in 4-6th decade, however young patients less than 40 years of age has also been documented in the literature to be affected by PV (Kanwar *et al.,* 2011). All our 3 affected parent cases were in 4th decade.

Possible etiological factors could be diet rich in garlic, drugs like thiol containing groups such as captopril, penicillmaine, phenol drugs like rifampicin, diclofenac, other ACE inhibitors, viruses like herpesvirus 8, high exposure to pesticides and genetic mutations. PV may occasionally be associated with other autoimmune disorders such as rheumatoid arthritis, myasthenia gravis, lupus erythematosus and pernicious anemia (Robinson *et al.,* 1997; Scully *et al.,* 2002b). Epitehlial desmosomes are anchoring junctions to which keratin filaments bind. The transmembrane proteins of desmosome are desmogleins and desmocollins. Dsg 1 and Dsg 3 have so far been identified which react with pemphigus antibodies (Dabelsteen, 1998). All forms of pemphigus involve circulating autoantibodies that bind to keratinocytes, disrupting normal cell-cell adhesion within epithelium and producing acantholysis. Clinically, this process appears as a blister or vesiculobullous disintegration of involved areas of skin and mucous membranes (Robinson *et al.,* 1997). In PV initial lesions affect the oral mucosa where they may be present for more than 6 months before other lesions develop (Dabelsteen, 1998). Clinically the oral lesions consist of varying sized asymptomatic blisters having thin roofs which rapidly ruptures giving rise to painful and bleeding erosions. Deep form of PV involves multiple, irregular ulcers arising from healthy mucosa causing burning sensation and difficulty in eating (Bystrym, 2005). Common sites of

involvement includes buccal mucosa, palate, tongue, lips, attached gingiva causing desquamative gingivitis and then gradually involves entire oral cavity along with esophageal and laryngeal mucosa which may lead to dysphagia and airway obstruction. Other oral manifestations includes halitosis, sialorrhea and formation of brown to black crust and tissue tags at the periphery of ruptured bullae due to which patient reports blood along with tissue tags while spitting saliva (Bystrym, 2005; Martinez *et al.,* 2010). After weeks to months, the condition progresses with lesions appearing on skin usually involving scalp, face, back, upper chest and upper torso and gradually involving the entire body (Bystrym, 2005) Initially skin lesions show small blisters with thin roof which rupture in several days and replaced by sharply outlined superficial erosions with loose epidermis which tend to enlarge gradually at the edges forming large flaccid bullae. Lesions usually begin with multiple, pruritic, crusted and coin sized patches which usually are described as cornflakes. These crusts can easily be removed leaving superficial erosions. If left untreated lesions over the weeks to months, become confluent and resembles exfoliative erythroderma involving the entire skin surface. The above following features were present in our case 1 and case 2 with only oral lesions in case 3. Other clinical findings include nail dystrophy, paronchyia and sublingual hematomas (Santoro *et al.,* 2013). Pemphigus is associated with premature birth and fetal death (Bystrym, 2005).

Diagnostic modalities and clinical examinations that aid in diagnosis of PV includes Nikolsky's sign which refers to direct application of pressure on blister causing the extension of blister. The indirect Nikolsky's sign is positive when the application of friction on clinically normal skin induces a blister. Although being a viable test, but it is difficult to achieve positive results in most of the intraoral cases as with our 3 clinical cases (Santoro, 2013) Another is Asboe-hansens sign which is the enlargement of bullae by applying finger pressure, due to release of its contents through the surrounding epidermis which was positive with case 2 (Ganapati, 2014) The intensity and severity of disease can be assessed by 'Autoimmune bullous skin disorder intensity score' and 'Pemphigus Disease Area Index' (PDAI) (Santoro *et al.*, 2013). The tentative clinical diagnosis must be confirmed by means of complementary tests such as exfloliative cytology, histopathological study which was confirmed in case 1 and 2, DIF and IIF, which reveals Tzanck acantholytic cells, intracellular edema in the suprabasilar portion of stratum spinosum, IgG and C3 deposits in epithelial intracellular spaces as confirmed in case 3 and antibodies targeted to epithelial cell surface like IgG 4 and IgG 1 respectively (Ali, 2011).

Differential diagnosis of mucosal involvement includes acute herpetic stomatitis, apthous stomatitis, erosive/bullous lichen planus, mucous membrane pemphigoid and erythema multiforme. Cutaneous involvement includes pemphigus foliaceous, linear IgA bullous dermatoses, herpetiformis dermatitis, Steven Johnson syndrome, Hailey Hailey disease and Grover disease. Complications of PV include malnutrition and dehydration due to multiple ulcers. Staphylococcus septicemia because of poor hygiene and immunity due to long term steroid therapy treatment, respiratory failure and underlying neoplasms which leads to mortality with a rate of 3.58% (Kanwar, 2011). Dental complications include inadequate maintenance of oral hygiene, caused by pain and hemorrhage, which leads

to increase dental plaque and contribute to progression of periodontal diseases (Santoro *et al.,* 2013).

The therapies used to control pemphigus can be divided in to those that act rapidly and are usually use to control the activity of the disease and those with a delayed effect that are generally used in chronic management to decrease the need of systemic corticosteroids. The goal of therapy includes minimizing disease burden and improving quality of life. Treatment is usually divided in to 3 phases that includes control, consolidation and maintenance. Since there is much heterogeneity in the manifestation of disease and its response to therapy, treatment is tailored to the needs of individual patient (Scully, 2008a; Bystrym, 2005).

Fig.1 Diffuse erythematous areas on right buccal mucosa

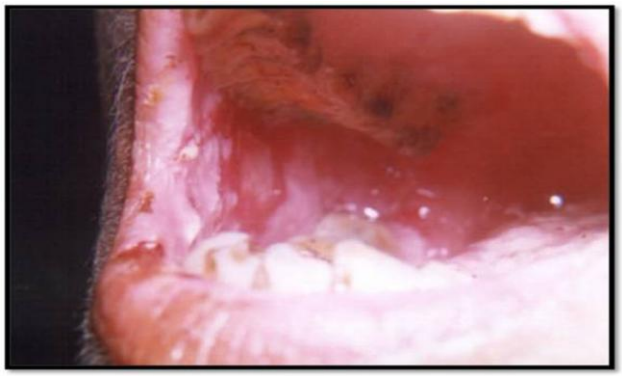


Fig.2 Diffuse erythematous areas on left buccal mucosa

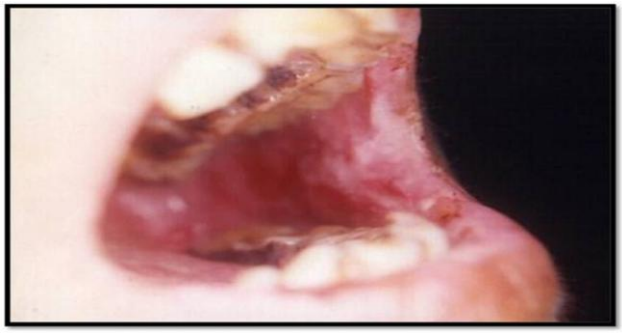


Fig.3 Irregular erosions on upper labial mucosa and dorsum of tongue

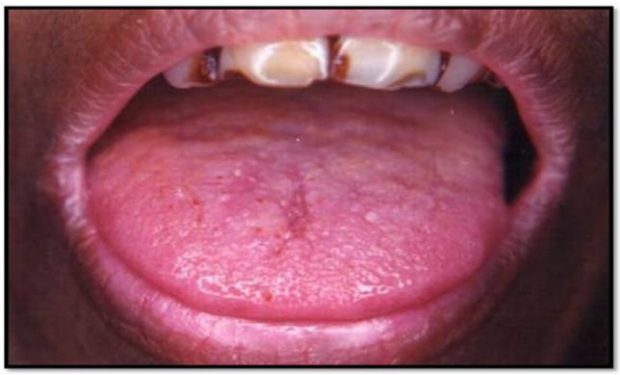


Fig.4 Epithelial tissue tags at the periphery of the erosions

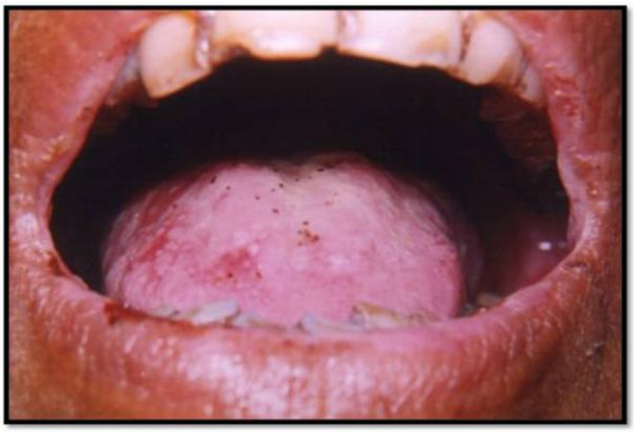


Fig.5 Multiple well-circumscribed, intact fluid-filled bullae on the abdomen

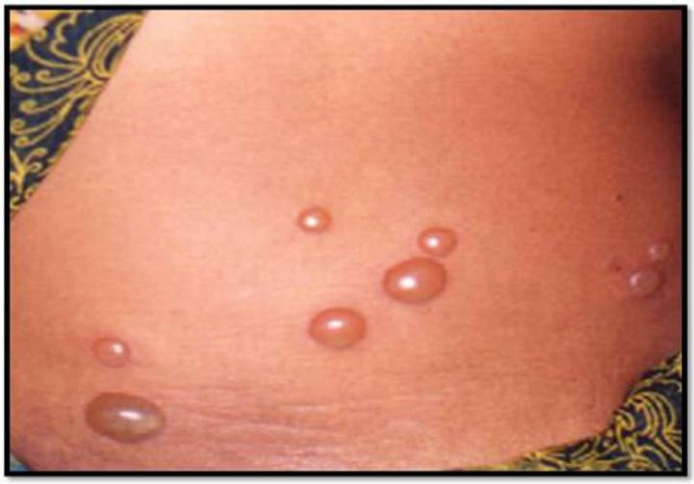


Fig.6 Multiple well-circumscribed, intact fluid-filled bullae in upper and lower limbs

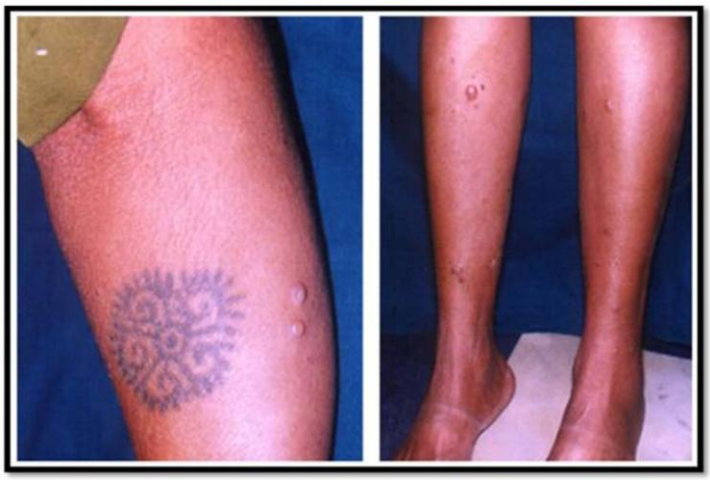


Fig.7 Ruptured bullae on an erythematous base

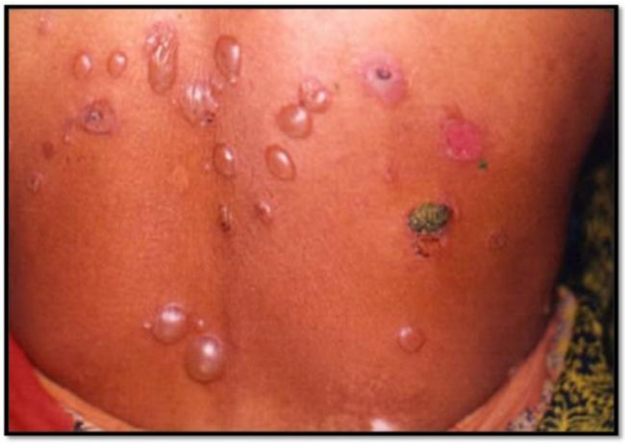


Fig.8 H & E sections showing - intra epithelial split & acantholytic cells

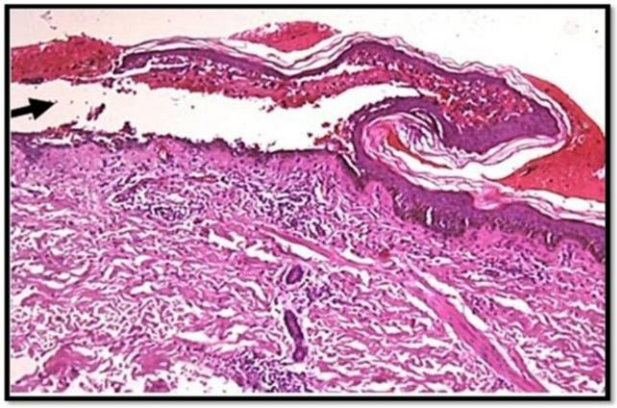


Fig.9 Healed bullae over abdomen

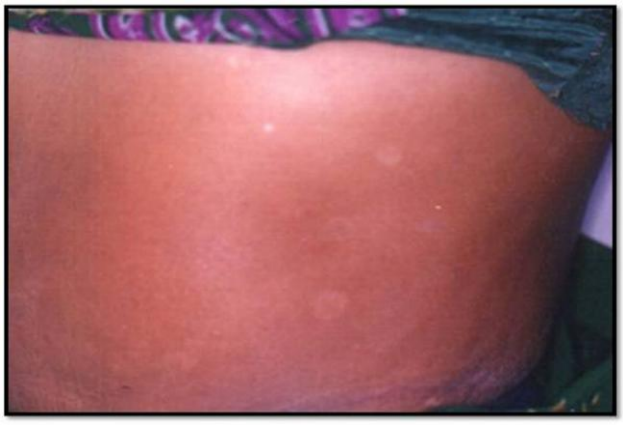


Fig.10 Partially healed bullae over back

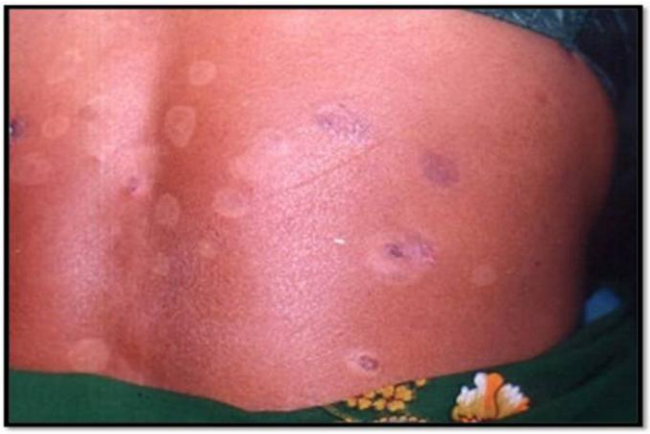


Fig.11 Healed erosions on labial mucosae

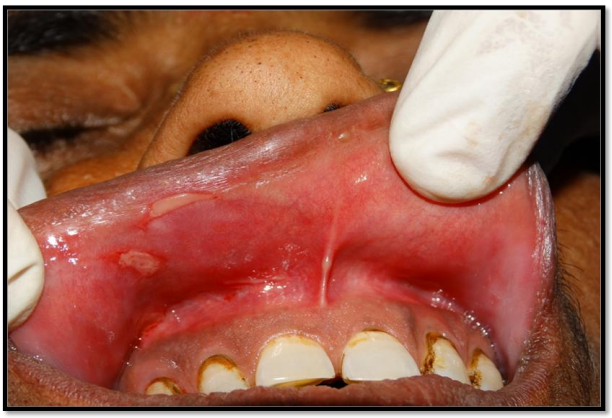


Fig.12 Partially healed erosions on right buccal mucosa

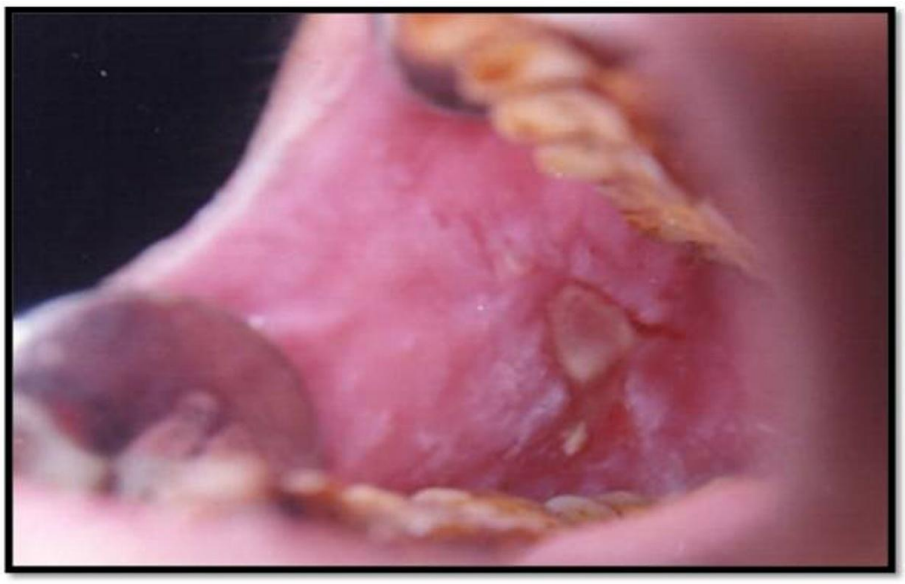


Fig.13 Partially healed erosions on left buccal mucosa

Fig.14 Single intact vesicle in left retromolar area

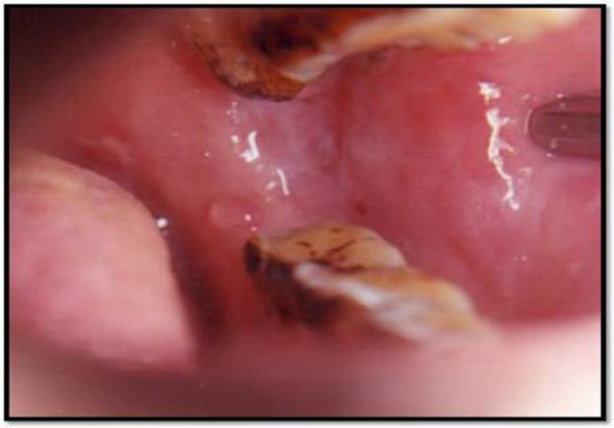


Fig.15 Completely healed skin lesions

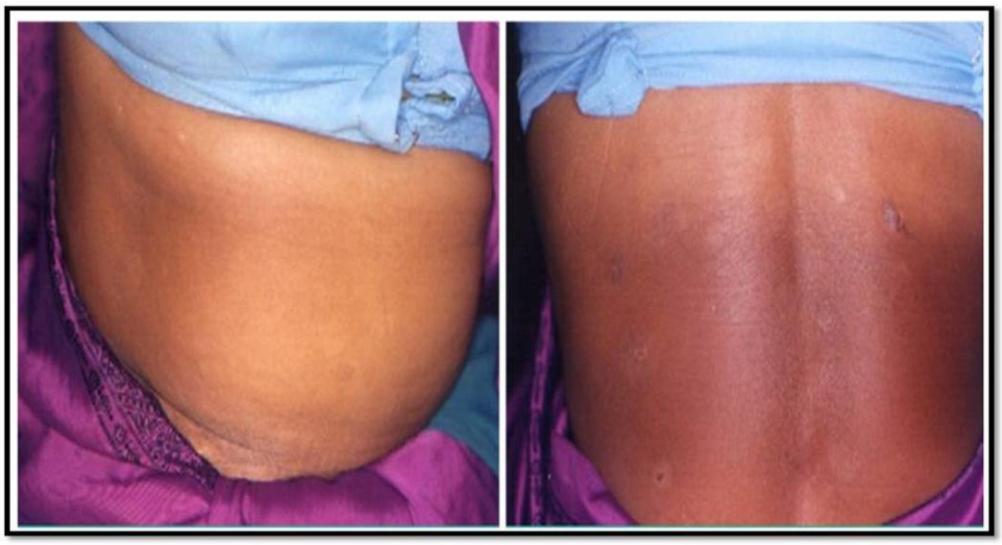


Fig.16 Multiple erythematous macules in front of neck

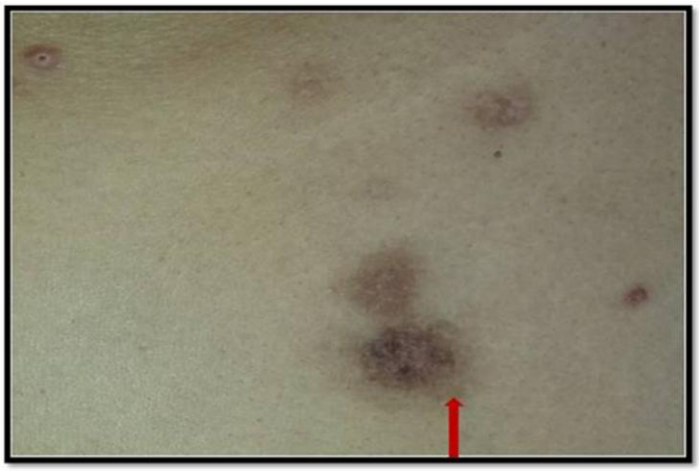


Fig.17 Black brown scab/papules on right lateral neck

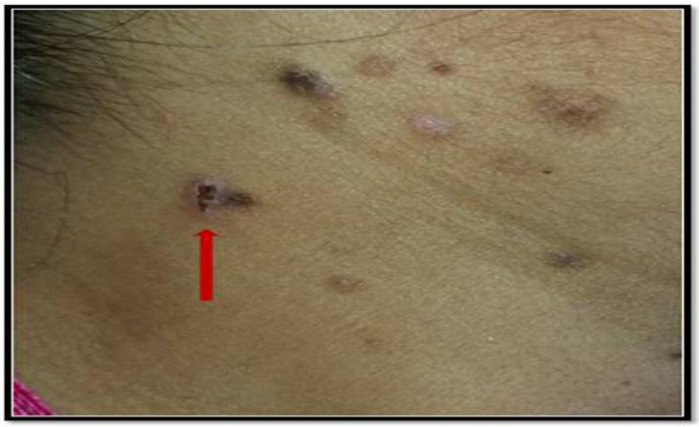


Fig.18 Brown/black scab on breast region

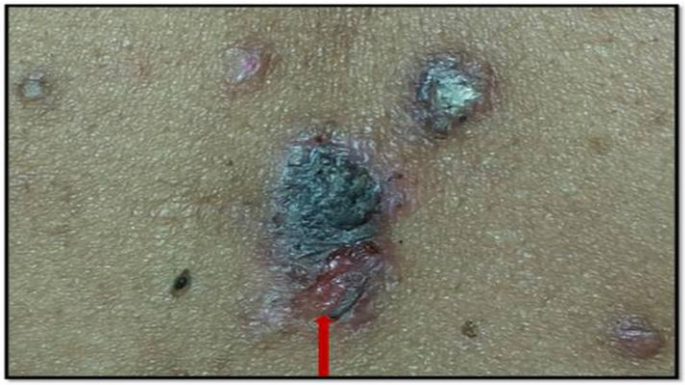


Fig.19 Large black brown encrustation on right abdominal region showing many intact fluid filled vesicles

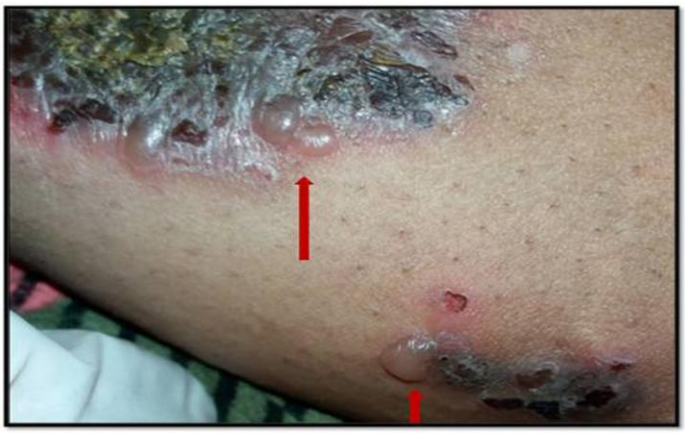


Fig 20 Lesion covered by fluid of blister due to release of fluid from blister positive Asbo Hansen sign

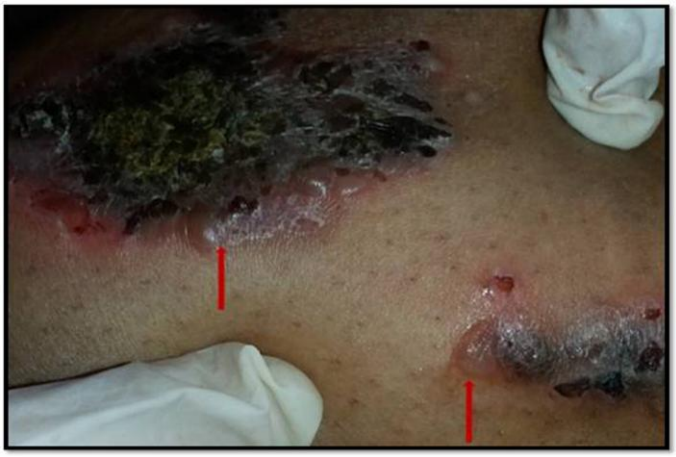


Fig 21 Diffuse irregular erythematous lesion with raw eroded areas on left buccal mucosa

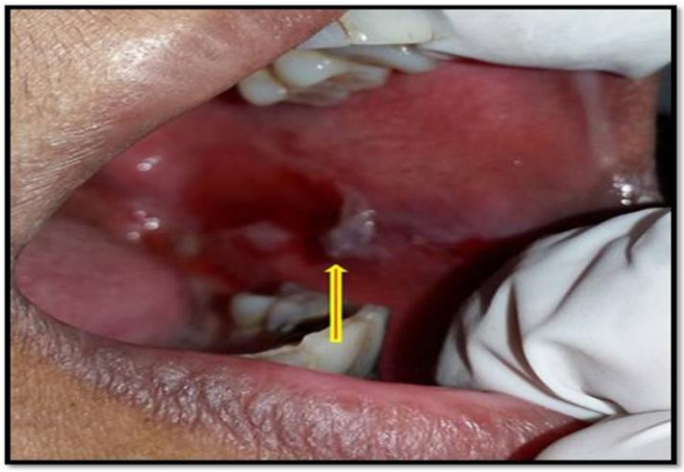


Fig 22 Diffuse irregular erythematous lesion with raw eroded areas on right buccal mucosa

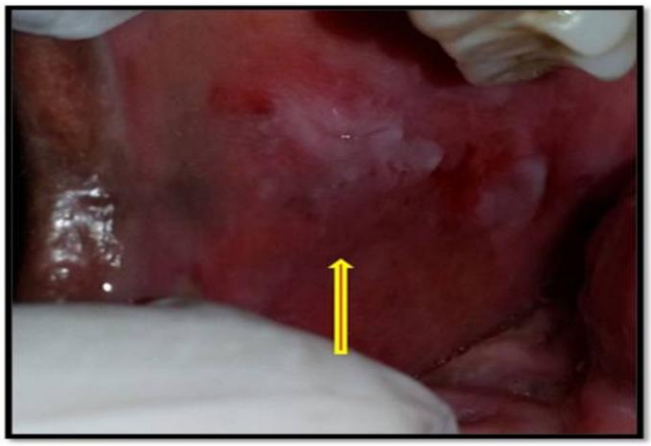


Fig 23 Extensive erythematous lesions with ruptured vesicle & bullae on lower labial mucosa

Fig 24 Solitary ill defined erythematous lesion on upper lip

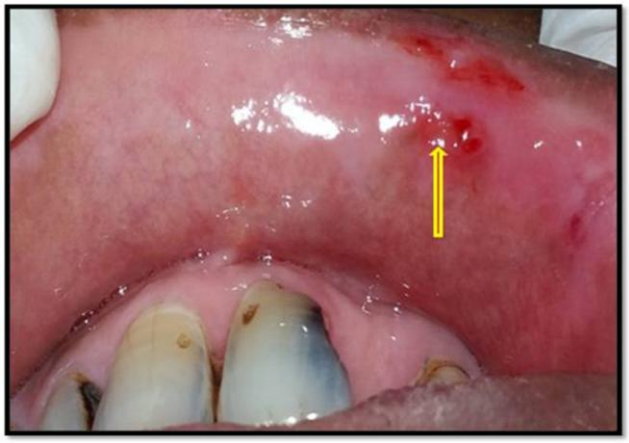


Fig 25 Tissue tags partially removable from the periphery of ruptured bullae/vesicles

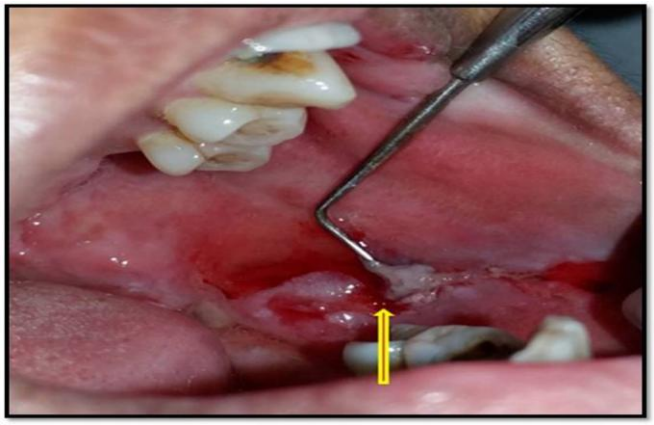


Fig 26 Diffuse erythematous lesion on palate without vesicles

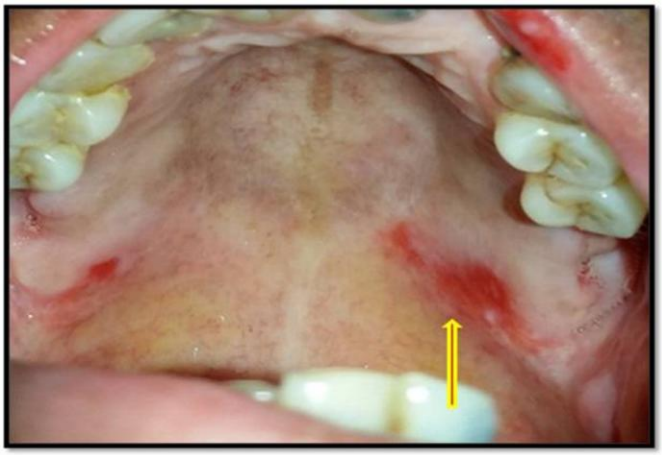


Fig 27 H&E section showing suprabasilar split and acantholytic cells

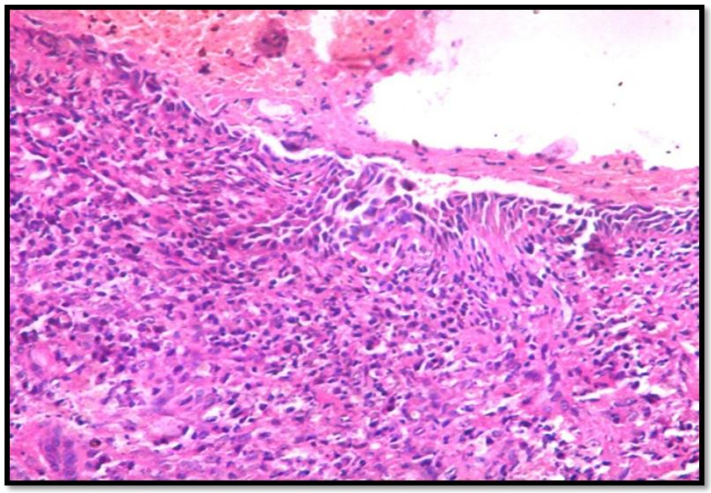


Fig 28 Completely healed lesions on buccal mucosa

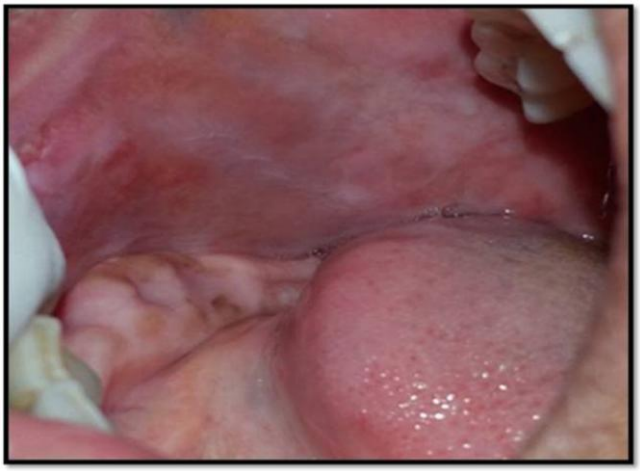


Fig 29 Partially healed lesions on lower labial mucosa

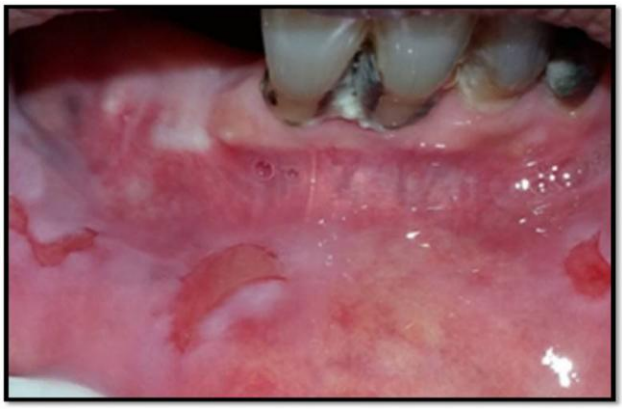


Fig 30 Partially healed lesions on neck

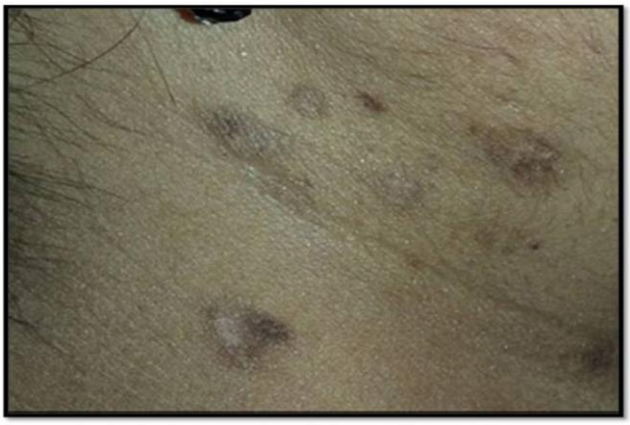


Fig 31 Partially healed lesion on breast

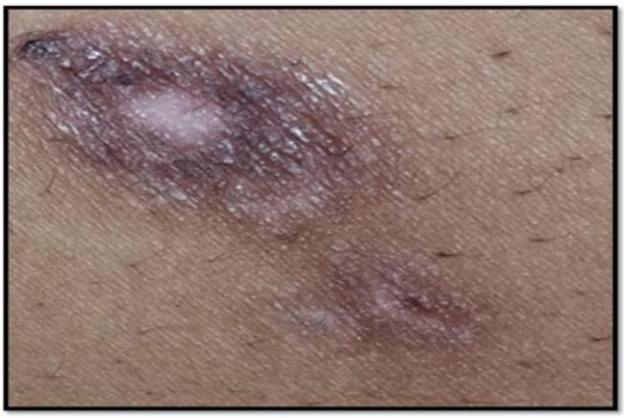


Fig 32 Dry scaly papules on abdomen

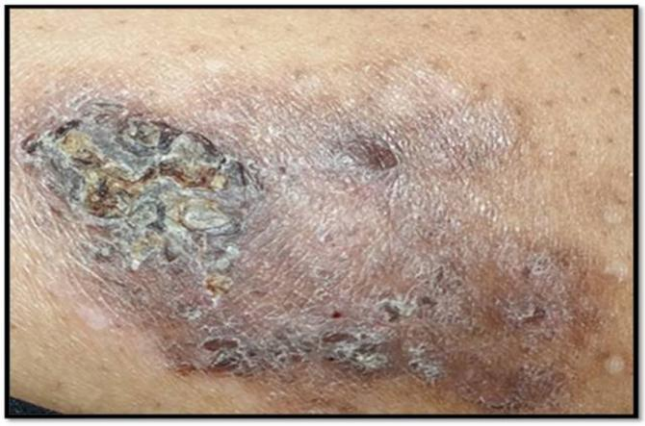


Fig 33 Erosive lesions on buccal mucosa

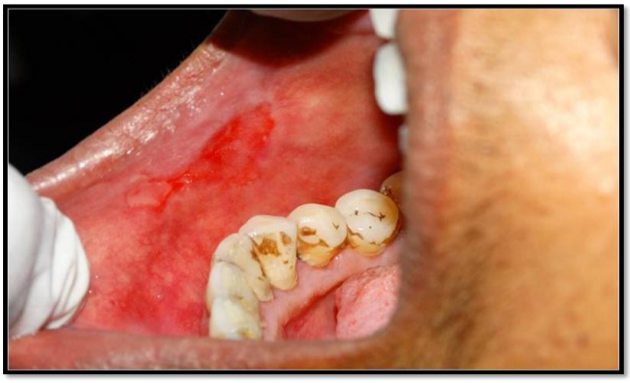


Fig 34 Erosive lesions on dorsum of tongue

Fig 35 Erosive lesions on palate

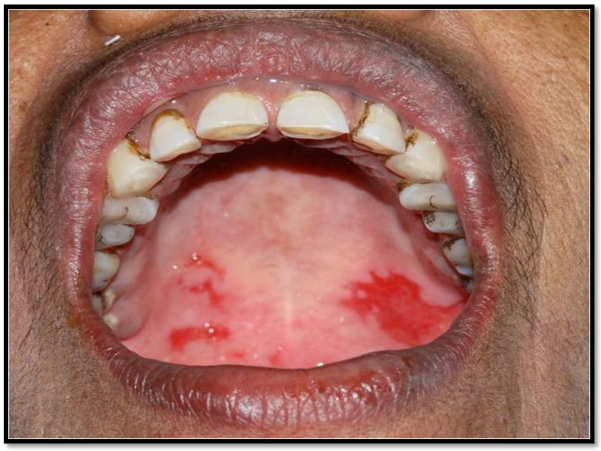
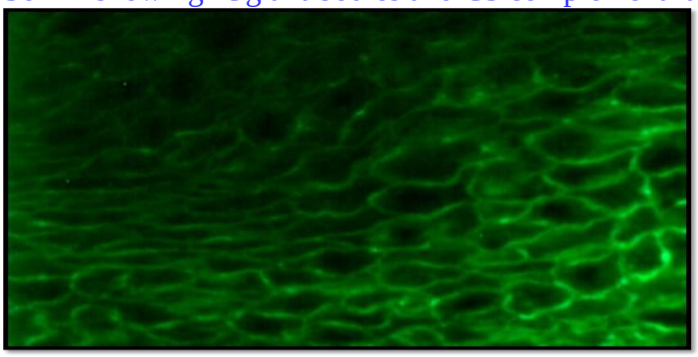


Fig 36 Dif showing IGg antibodies and C3 complement factor



Corticosteroids are the cornerstone of the treatment. Corticosteroids can be prescribed to pemphigus patients with systemic low dosages of 45-60 mg/day or high dosages of 120-150 mg/day depending upon the severity of the case and then gradually tapered in 6-10 weeks. Before the patients tapers to less than 2.5 mg daily, cortisol level before the daily dose of prednisolone is checked to evaluate adrenal insufficiency

and accordingly side effects should be monitored simultaneously (Solankhi *et al.*, 2012; Santoro *et al.,* 2013). In patients with non progressing oral lesions moderate to high potency topical corticosteroids are recommended, applied 2-3 times a day such as 0.05% clobeatsole proprionate. Topical anesthetic rinses should also be prescribed for symptomatic relief of oral lesions along with antihistaminics to provide relief from severe itching with skin lesions. In order to reduce the corticosteroid dose such treatment is combined with immunosuppressive drugs such as cyclophosphamide 100 mg per day, azathioprine 1-2 mg/kg/day, other options includes cyclosporine 5-8 mg/kg/day, mycophenolate mofetil 35-45 mg/kg/day, dapsone, gold, minocycline, prostaglandin E_2. As the corticosteroids therapy are tapered in refractory cases rituximab are initiated which is a safer alternative to steroids. Other emerging treatments includes intravenous immunoglobulins, topical tacrolimus, Dexamethasone - Cyclophosphamide Pulse therapy (DCP) and plasmapheresis (Scully, 2008 a; Santoro *et al.*, 2013; Darling, 2006). Non pharmacological strategies includes cleansing with antibacterial soap twice daily with topical application of antibiotics on skin lesions followed by bandaging with non stick gauze such as petroleum gauzes to prevent sepsis. Patient should avoid aggressive oral hygiene practice including flossing due to increase risk of pain and bleeding (Santoro *et al.,* 2013). The above mentioned treatment plan was given to all 3 patients in case series with successful results.

Conclusion

The pathogenesis of PV is rapidly being unravaled and search for etiological factors may soon bear fruit. Sometimes the mouth may be the only site of involvement in PV for a year which may lead to delayed diagnosis and inappropriate treatment as seen in above three cases. Thus newer and more effective, specific, safer treatments are needed to reduce the morbidity and mortality rates. Our present 3 cases supported the literature of PV in all aspects and proved us to be a challenging job in diagnosing and successfully treating these patients. Hence, oral physicians plays equally crucial role along with dermatologists to diagnose and treat such potentially life threatening disorders reducing morbidity and mortality and improving the overall quality of life of patient.

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