ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 8, Issue, 12, pp.43666-43667, December, 2016

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

REVIEW ARTICLE

ORAL MUCOSAL PEMPHIGUS VULGARIS WITH CUTANEOUS INVOLVEMENT: RARE CASE REPORT

***,[1]Dr. Pradkhshana Vijay, [2]Dr. Nilesh Pardhe and [3]Dr. Ishank Singhal**

[1]Department of Oral Pathology and Microbiology, Faculty of Dental Sciences, KGMU Lucknow
[2]Department of Oral Pathology and Microbiology, NIMS Dental College Jaipur
[3]Department of Oral and Maxillofacial Surgery, Indraprastha Apollo Hospitals, New Delhi

ARTICLE INFO

Article History:
Received 23rd September, 2016
Received in revised form
20th October, 2016
Accepted 07th November, 2016
Published online 30th December, 2016

Key words:

Desmosomes, Immunofluorescence,
Acantholysis, Management, Chronic disease.

ABSTRACT

Pemphigus vulgaris (PV) is an autoimmune disease which is possibly life threatening affecting the skin and oral mucosa. The epithelial lesions are formed due to auto-antibodies which act in response with desmosomal glycoproteins on the keratinocyte surface causing loss of cell adhesions. This loss of intercellular adhesion leads to intraepithelial bullae formation. We report a rare case of oral pemphigus vulgaris with skin involvement in a middle aged female patient with brief on review of literature.

Citation: Dr. Pradkhshana Vijay, Dr. Nilesh Pardhe and Dr. Ishank Singhal, 2016. "Oral mucosal pemphigus vulgaris with cutaneous involvement: Rare case report", *International Journal of Current Research*, 8, (12), 43666-43667.

INTRODUCTION

Pemphigus is an autoimmune, chronic inflammatory mucocutaneous disease. The intraepithelial blister formation that occurs is due to loss of cell to cell adhesions (Shamim, 2008). The disease mostly occurs in fifth to sixth decade of life, predominantly in females (Williams, 1989). IgG anutoantibodies are directed against cell surface of keratinocytes which is responsible for bullae formation in PV, (Williams, 1989). Most commonly presented form of pemphigus is pemphigus vulgaris (PV). Most commonly buccal mucosa, labial mucosa and palate are affected (Black, 2005). We report a case of pemphigus vulgaris in a middle aged female patient who also presented with generalized skin lesions.

Case report

A 40 years old female patient presented with a chief complaint of lesions on the tongue and buccal mucosa, intermittent in presentation, with periods of remission and exacerbations since 1 year associated with burning sensation and pain on having food. Apart from the oral presentation, she also had generalized skin lesion.

****Corresponding author: Dr. Pradkhshana Vijay,***
Department of Oral Pathology and Microbiology, Faculty of Dental Sciences, KGMU Lucknow

A history of bleeding of unaffected area of skin on rubbing was also present. Intraoral examination revealed reddish and ulcerated erosive lesion on left lateral border of tongue measuring 1x0.5cm in diameter. Erosive lesion present bilaterally on buccal mucosa measuring 1x1cm in diameter. (Fig 1a, 1b). The examination of cutaneous lesion revealed reddish brown lesions on arms and trunk which appeared eroded and ulcerated measuring 0.5x1 cm and 2x2 cm respectively, (Fig 2). The eroded areas were the bullae that ruptured leaving an erythematous, denuded surface. Nikolsky's sign was positive. Routine blood investigations were within normal limits. Based upon the history and clinical findings a provisional diagnosis of bullous pemphigoid was made.

Pemphigus, mucous membrane pemphigoid and erosive lichen planus were included in differential diagnosis. Following this an incisional biopsy was performed and specimen was sent foe Histopathological evaluation. Microscopic examination revealed hyperparakeratinized stratified squamous epithelium, with a characteristic intraepithelial separation above the basal cell layer of epithelium. Superficial layer of the epithelium appears to be exposed away, leaving basal cells resembling Row of Tombstones. Few epithelial cells showed acantholysis and assumed a round shape suggestive of Tzanck cells was also noted. Mild to moderate chronic inflammatory cells were seen in the C.T stroma, (Fig 3). Based on the Histopathological

findings a final diagnosis of pemphigus vulgaris was made. Oral predinisolone 60mg/day for 4 days and multivitamins were prescribed. Diprolene 0.05% to be applied topically three times daily was advised.

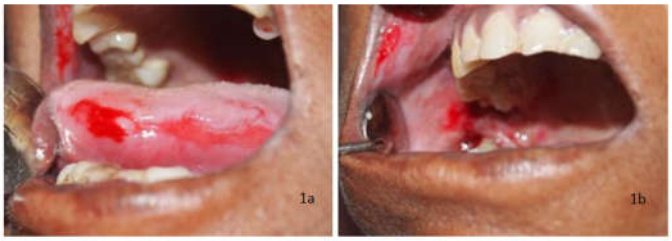

Fig. 1a. Intraoral examination of tongue showing erosive red lesion on left lateral border of tongue.
Fig. 1b. Intraoral examination of right buccal mucosa showing red erosive lesion extending upto the lip commissure on right side

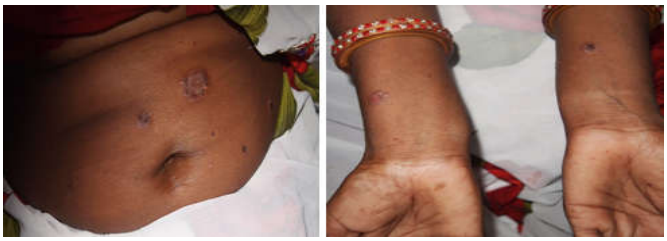

Fig. 2. Cutaneous involvement of lesion showing ruptured bullae with erosive areas on trunk and arms

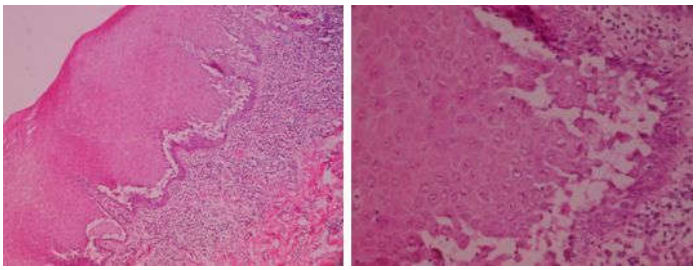

Fig. 3. Photomicrograph showing hyper parakeratinized stratified squamous epithelium with intraepithelial split and areas of acantholysis and tzanck cells with subepithelial inflammatory cell infiltrate chiefly lymphocytes

The predinisolone dose was tapered at regular intervals. Patient was called for review after 15 days and showed relief in the symptoms.

DISCUSSION

Pemphigus is derived from greek word Pemphix meaning "blister or bubble", (Hashimoto, 2003). It was named at first by Wickham in 1971. "Vulgaris" in latin means common. PV is the most common disease among the pemphigus family of lesions (Endo, 2008). It is an autoimmune disease showing acantholysis of epithelium. IgG autoantibodies produced; target two structured desmosomes, Desmoglein 1 and 3 (Endo, 2008). Female patients are affected more commonly and are mostly in the fourth and fifth decades of life. Initial involvement is of oral mucosa followed by skin in about 50% cases (Iamaroon, 2006). The present case also showed oral lesions prior to the skin lesions. The oral bullae rupture easily and appear as erosion in the area. The present case also showed bullae which ruptured and presented as erosions. Clinical differential diagnosis includes aphthous ulcers, pemphigoid, erythema multiforme, Bullous lichen planus (Iamaroon, 2006). Chronic ulcers appear mostly on the lower lip. The pemphigoid group of lesions don't show oral lesions preceding the skin lesion and histopathologically show a sub-epithelial split. Erythema multiforme mostly is associated with a medical or drug history with a triad of lesions involving eye, genitals and oral cavity. In Lichen planus, there is no intraepithelial split and it shows dense juxtaepithelial lymphocytic infiltrate (Kancoar, 2011).

The classical lesion of pemphigus is bulla arising in the normal oral or skin mucosa. The characteristic sign of disease is the positive Nikolysky's phenomenon which is produced by application of pressure to the adjacent normal area which results in new lesion (Iamaroon, 2006). The present case also showed positive nikolysky's sign. The diagnosis can be made provisionally on clinical findings, but confirmation is done histopathologically by the presence of an intraepithelial split with presence of acantholysis and tzanck cells in the epithelium. Direct immunofluorescence detects the presence of IgG or IgM in intercellular spaces giving a fish net pattern. The present case revealed intraepithelial split with acantholysis and tzanck cells (Kancoar, 2011). The aim of the treatment is to reduce the remissions. Corticosteroids are the preliminary drugs in the management of PV. The mild cases of PV localized to oral mucosa can be managed with topical corticosteroids like clobetasole propionate. For resistant cases Inj. Triamcinolone can be administered (Iamaroon, 2006 and Kancoar, 2011).

Conclusion

PV is a fatal disease with oral cavity being the first manifestation. The diagnosis is based on clinical, Histopathological and immunofluorescent findings. Early treatment should be felicitated to benefit the patient recovery. Long term follow up is needed to identify possible remission of disease.

REFERENCES

Black, M., Mignogna, M. D. and Scully, C. 2005. "Number II. Pemphigus vulgaris," Oral Diseases, 11(3):119–130.

Endo, H., Rees, T.D., Hallmon, W.W. 2008. Disease progression from mucosal to mucocutaneous involvement in a patient with desquamatous gingivitis area with Pemphigus Vulgaris. *Journal of Periodontology.,* 99 (2): 368-375.

Hashimoto, T. 2003. "Recent advances in the study of the pathophysiology of pemphigus," *Archives of Dermatological Research,* 295(1): 2–11.

Iamaroon, A., Boonyawong, P., Klanrit, P., Prasongtunskul, S. and Thongprasom, K. 2006. "Characterization of oral pemphigus vulgaris inThai patients," *Journal of Oral Science*, 48(1):43–46.

Kancoar, A.J., Deo. 2011. Pemphigus in India. *Indian J Dermatol Venereai leprol*, 77: 439-49.

Shamim, T., Varghese, V.Z., Shameena, P.M., Suddha, S. 2008. Pemphigus Vulgaris in oral cavity. Clinical analysesof 71 cases. *Med Oral Pathol Buccal*, 2008; 13:2622-6.

Williams, D.M. 1989. Vesiculobullous Mucocutaneous Disease: Pemphigus Vulgaris. *J Oral Pathol Med*., 18:544.

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