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Steroids as a boon in oral pemphigus vulgaris: A case report and treatment overview

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ABSTRACT

Pemphigus vulgaris (PV) is an autoimmune blistering disease characterized by circulating pathogenic IgG autoantibodies against desmoglein 3 and desmoglein 1. Oral lesions readily rupture, new bullae developing as the older ones rupture and ulcerate. Oral lesions may precede cutaneous lesions in 53.52% cases. Prompt recognition and diagnosis is critical since it may prevent cutaneous involvement. Systemic steroids remain the gold standard treatment of PV. Their use has transformed what was almost invariably a fatal illness into one whose mortality is now below 10%. We report a case of patient with oral ulcers since 4 months, who is diagnosed with PV without cutaneous involvement.

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1. Introduction

Pemphigus encompasses a group of life-threatening autoimmune mucocutaneous bullous diseases characterized by flaccid blisters and erosions of the mucous membrane and/or skin caused by IgG autoantibodies against the desmosomal adhesion proteins, desmoglein 3 and/or desmoglein 1, on epidermal keratinocytes.[1]

Accompanying body catabolism with loss of body fluids and proteins and secondary bacterial and viral infections may lead to sepsis and cardiac failure.[2] Before the advent of systemic corticosteroids, the prognosis of pemphigus was almost fatal within 2 years after making the diagnosis.

Pemphigus vulgaris (PV) is the most common form and frequently involves the oral cavity. Intraoral involvement is considered to be the initial manifestation in 53.52% patients while 23.94% had onset on skin and oral mucosa simultaneously with peak incidence in fifth and sixth decade of life.[3]

We report a case of intraoral PV without cutaneous involvement in a 40-year-old male patient treated using corticosteroids and discuss various treatment modalities currently available.

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2. Case report

A 40-year-old male patient reported to the outpatient dental department with complaint of recurrent intraoral ulcers since 4 months which were very painful and affected his normal oral functions. Past medical and dental history was not contributory. Patient was a chronic tobacco chewer. Intraoral examination revealed widespread erosions involving hard palate, buccal mucosa and lower labial mucosa. Lesions over lower lip showed bloody crustations (Figs. 1 and 2). The lesions were tender on palpation and exhibit bleeding on touch. No skin lesions were observed. Incisional biopsy was performed and tissue sent for histopathology.

2.1. *Histopathological examination*

H&E stained sections exhibit suprabasal split with acantholysis, intercellular odema and loss of intraepithelial bridges in lower third of epithelium. Underlying connective tissue shows moderate inflammatory cell infiltrate (Fig. 3).

2.2. *Treatment*

Patient was given oral prednisolone 40 mg/day for 1 week along with topical application of triamcinolone acetonide 0.1% (w/w) thrice daily for 1 week. Oral rinse of benzydamine hydrochloride was prescribed for control of oral symptoms. Also, Nystatin mouthpaint was given to the patient to prevent secondary candidal infection. Calcium supplements and B complex vitamin tablets were also prescribed concomitantly. Significant improvement of symptoms and improvement in lesions was visible by the end of 1 week. Prednisolone dose was tapered to 20 mg/day for next 7 days, 10 mg/day for next 7 days, 5 mg/day for next 7 days followed by stoppage of prednisolone. Patient was symptom free with compete resolution of lesions on his follow up visit after 1 month (Fig. 4). Patient is free of lesions during his 6-month follow up visit.



Fig. 1 – Patient with ulcer and crustation over lower lip and buccal mucosa.



Fig. 2 – Patient with ulcers over palate.

3. Discussion

Pemphigus is primarily considered to be a dermatologic disease. The fact that PV commonly and initially affects the oral mucosa and then the skin gives dentists a great opportunity to detect the disease at an early stage. Oral mucous membrane is the first site of involvement in many cases of PV and may remain confined or extend to involve the skin with an average lag of 4 months.[4]

The initial goal of treatment is control and healing of lesions, induces disease remission, improve quality of life of the patients as well as attempting to minimize, as much as possible serious side effects of treatment.

1. *Systemic corticosteroids* are the best-established therapy for the management of PV attributed to their strong anti-inflammatory and immunosuppressive effects. Their introduction in the early 1950s resulted in a dramatic fall in mortality to an average of 30% with complete remission rates of 13–20%.[6] Systemic corticosteroids can be combined with an immunosuppressive agent, particularly when complications due to expected prolonged use (4 months) such as hypertension, diabetes mellitus, and osteoporosis are expected.[2,4]
2. *Immunosuppressive adjuvants*: Based on the current evidence, adjuvants have only a steroid-sparing effect, and may lead to steroid-free remission.

Azathioprine is a well-established choice as an adjuvant drug for the management of pemphigus.[4]

Mycophenolate mofetil should be considered in recalcitrant cases or when azathioprine and cyclophosphamide cannot be used.[4]

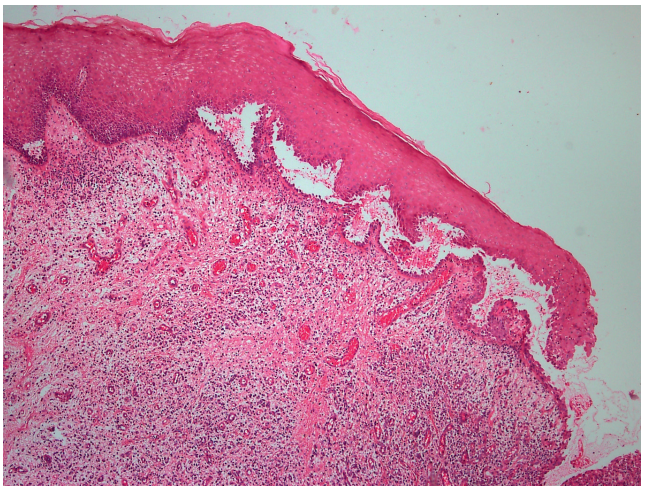


Fig. 3 – H&E stained photomicrograph showing suprabasilar split and Tzank cells with juxtaepithelial moderate inflammatory infiltrate (original magnification 4×).



Fig. 4 – Resolution of lesions after 30 days treatment with systemic corticosteroids.

Rituximab (anti-CD20 monoclonal antibody) has the potential to reduce desmoglein autoantibodies and selectively deplete B cells. It is indicated in patients who remain dependent on more than 10 mg prednisolone combined with an immunosuppressive adjuvant according to the EDF.[5]

Methotrexate could be considered as an adjuvant drug if more established drugs cannot be used because of high mortality and morbidity associated with it.[4]

Other less commonly used adjuvants include intravenous immunoglobulin, plasmapheresis, immunoadsorption, gold salts and cyclophosphamide.[5]

Topical treatment with potent corticosteroids like clobetasol propionate and oral typical corticosteroids like triamcinolone acetonide gel or calcineurin inhibitors in combination with systemic therapy may be beneficial. Intralesional injections of triamcinolone acetonide are beneficial for isolated oral lesions.[6]

4. Conclusion

We emphasize the importance of early diagnosis of PV and early initiation of treatment before the disease becomes too severe or widespread since prompt treatment has been associated with improved prognosis.

Conflicts of interest

The authors have none to declare.

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