Bullous Pemphigoid A Rare Autoimmune Disease: A Case Report

Sharma Manek*, SP Saravanan, Rai Rahul
Department of Periodontology, Army Dental Centre, New Delhi, India
*Corresponding author: maneksharma130@gmail.com
Received January 15, 2022; Revised February 18, 2022; Accepted February 27, 2022

Abstract
Bullous pemphigoid [BP] is an autoimmune blistering disorder characterized by the formation of autoantibodies causing blister formation potentially triggered by trauma, antimicrobials and non-steroidal anti-inflammatory drugs. A 31-year-old female patient reported with complaint of fluid filled blisters which appear intermittently for past 2 years, leaving painful ulcerations. Incisional biopsy was performed, along with adjunctive use of Low Level Laser Therapy, report of which showed bullous pemphigoid and better healing due to LLLT respectively. Thus, based on our clinical experience and result the use of laser as an adjunctive therapy in symptomatic management of desquamative lesions is indeed a food for thought.

Keywords: autoimmune, bullous pemphigoid, low level laser therapy


1. Introduction

Bullous pemphigoid [BP] is an autoimmune blistering disorder characterized by the formation of autoantibodies directed against components of the basement membrane. There is experimental and clinical evidence that these autoantibodies are responsible for causing the blisters [1]. The potential triggers that have been identified include trauma, antimicrobials and non-steroidal anti-inflammatory drugs [2]. There is also a biologically plausible relationship between BP and certain neurological conditions such as multiple sclerosis and parkinsonism [3]. Treatment depends on severity of disease, site of involvement and its progression. Till date, the mainstay of treatment remains symptomatic management of the condition with topical and systemic corticosteroids [4]. However, long term steroid therapy is proven to result in unwanted side effects, thus other modalities to provide symptomatic relief includes adjunctive use of laser in form of low level laser therapy [LLLT] and photo dynamic therapy [PDT]. LLLT is known to provide direct biostimulative light energy to body cells and it is proven to stimulate faster wound healing in chronic non-healing lesions of autoimmune origin [5].

2. Case Report

A 31-year-old female patient reported to Department of Periodontology, Army Dental Centre, Delhi Cantt, with chief complaint of fluid filled blisters which appear intermittently for past 2 years, leaving painful ulcerations. Chewing and brushing induced blister formation, which resolved within 6–7 days. Patient also presented history of pruritis followed by appearance of extraoral bullae which ruptured eventually leaving discoloration on the skin of face and neck. She has past history of multiple dental visits and was on topical steroids for more than one year.

2.1. Oral Examination Findings

On intraoral soft tissue examination, gingiva exhibited generalised diffuse erythematous zones with localized areas of desquamation and ulceration [Figure 1].

Figure 1. Baseline picture of gingiva

The surface was shiny with loss of stippling and blunt interdental papillae with spontaneous bleeding tendency. Nikolsky’s sign was positive on unaffected labial gingiva. In addition, whitish striae were present on the upper labial mucosa. The ventral mucosa of the tongue appeared ulcerated with irregular margins. On palpation, gingiva was tender, soft in consistency and easy bleeding tendency was evident.
2.2. Diagnosis and Treatment

A provisional diagnosis of bullous lesion of oral cavity was made with a differential diagnosis of erythema multiforme, epidermolysis bullosa, cicatricial pemphigoid, bullous pemphigoid and bullous lichen planus.

Initial therapy included supragingival plaque removal, medications benzocaine 10% gel twice daily, candid 1% w/v mouth paint twice daily for five days and systemic antioxidants for 30 days (Cap Lycopene 1 OD).

Under aseptic conditions, biopsy was performed from labial gingiva of 23 and 33-34 region using the stab and roll technique [Figure 2]. The sample was subjected to histopathological analysis and direct immunofluorescence [DIF].

In phase two therapy, laser ablation was done using 980 nm diode laser at 0.8 watt in continuous wave mode. Post laser therapy, the patient was on Benzydamine HCl mouthwash (10 ml thrice daily prior to meals) for 1 week.

2.3. Biopsy and Histopathologic Findings

Histopathological evaluation revealed hyperparakeratinized stratified squamous epithelium with underlying connective tissue stroma with characteristic subepithelial split along with mononuclear inflammatory cell infiltrate [Figure 3].

DIF revealed linear deposition of IgG and C3 along the dermal–epidermal junction [Figure 4].

Based on clinical, histopathological and immunological findings, a final diagnosis of BP was arrived.

The patient was referred to Dermatologist for management of skin lesions. Dermatologic treatment included systemic prednisolone 30 mg and 0.1% tacrolimus topical gel for 15 days. The post-op review after 1 month revealed that the incidence of bullae had reduced significantly following which systemic steroid was tapered and patient placed under maintenance phase [Figure 5].

3. Discussion

BP is one of the most common autoimmune blistering disorders affecting the oral cavity [6]. It usually affects individuals during the 3rd and 4th decades of life, with female gender predilection and only rare cases of BP were reported in children and young adults [7,8]. BP is characterized by autoantibodies that recognize self-antigens at the basement membrane zone [BMZ] known as BP180 [180kDa] or BPAG2, and BP230 [230kDa] or BPAG1. Both the antigens are key components of hemidesmosome, which is responsible for union of epidermis and dermis [7].

Lesions of BP usually appear on the skin with mucous membrane involvement in approximately one third of patients. The skin lesions are distributed mainly in trunk and limbs. Although vesicles and bullae are present, they are often preceded by or associated with erythematous papular eruptions. Oral mucosal lesions of BP can’t be differentiated from other bullous disorders including Mucous Membrane Pemphigoid. Bullae and erosions usually present on the attached gingiva being a commonly affected site and other sites include buccal mucosa, soft palate and floor of the mouth [9]. Clinical presentation of chronic desquamative gingivitis is usually evident in almost all such cases of autoimmune blistering oral disorders. Severe itching and painful blister/ ulcer formation are the most commonly encountered symptoms in these patients [10].

Diagnosis depends on the careful correlation of clinical features along with histopathological examination and immunological confirmation. Histopathological analysis of non-bullous lesions usually demonstrates the presence of cosinophilic spongiosis with a mixed dermal inflammatory infiltrate. Bullous lesions are usually present with subepidermal split with eosinophils, neutrophils and fibrin within the blister and dermal inflammatory infiltrate [11].

Apart from patient history and clinical presentation, histopathological analysis along with DIF remains the...
gold standard for confirmation of the condition. Direct and indirect immunofluorescence plays a central role in the diagnosis of all such conditions. In direct immunofluorescence the biopsy reveals deposits of IgG, C3, Ig A and IgM along the BMZ [12].

Treatment is based on the degree of cutaneous and oral involvement. Topical steroids usually give satisfactory result in case of smaller area of skin involvement, whereas in larger areas recurrent cases are treated with systemic steroids and immunosuppressive agents. The relapse rate of BP ranges from 27.87% to 53% after disease remission, while the majority of relapses occur as early as 6 months during remission [13].

Topical/ systemic steroids are aimed at reducing disease activity with the minimal possible dose to reduce systemic adverse effects. The recommended initial dose for prednisolone is 0.3 mg/kg/day in case of localised disease without any systemic involvement. Once the disease activity is under control dose can be tapered gradually. Only in exceptional cases a prolonged course of moderate to high dose oral steroids/ immunosuppressants may be indicated. The immunosuppressive agents advocated include adjunctive use of azathioprine or mycophenolate mofetil. [14]

Wound healing and tissue repair are complex processes that involve dynamic events including clotting, inflammation, granulation tissue formation, epithelialization, collagen synthesis and tissue remodeling. It is studied that LLLT can lead to increased mitochondrial activity with a consequent rise in adenosine triphosphate, vasodilation and protein synthesis. It also leads to decrease in prostaglandin levels, increased cellular mitosis, migration and proliferation of keratinocytes and neo-angiogenesis. LLLT is a non-invasive, non-thermal and painless procedure with no thermal damage at tissue or cellular level. It also has potential biostimulatory effects on tissues and cells with an increase in systemic microcirculation and tissue oxygenation, cell metabolism, tissue regeneration and thereby leading to promotion of tissue healing [6].

The ultimate goal of treatment includes improved oral health quality of life by reducing new blister formation and promoting healing of existing lesions.

4. Conclusion

Our results were consistent with our expectations. Post lasering, an excellent effect was observed in zones of high inflammation and friable tissue. Also, the patient experienced decreased post operative pain associated with improved VAS scores from an initial baseline value of 6 to 3, three weeks post laser therapy with less bleed and inflammation, rapid hemostasis and healing. Thus, based on our clinical experience and result the use of laser as an adjunctive therapy in symptomatic management of desquamative lesions is indeed a food for thought.

References