

Periodontal and Systemic Treatment Approach on Pemphigus Vulgaris: A Case Report

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Abstract

Objective: In this case report, both the diagnosis of pemphigus vulgaris and the periodontal treatment approach including the use of local/systemic medications are presented.

Case Presentation: 36-year-old female patient applied to the periodontology clinic with complaints of burning mouth and pain. Physical examination revealed cutaneous blisters on nose, hand and fingers while intra-oral examination showed widespread desquamation and ulcers depending on PV and severe gingival inflammation due to the lack of oral hygiene and oral PV. Initial periodontal treatment (IPT) was implemented to the patient along with local and systemic medications. Periodontal parameters including plaque Index (PI), gingival Index (GI), probing depth (PD) and clinical attachment level (CAL) were recorded before and six weeks after IPT. Periodontal treatment procedures did not cause any negative effect on the lesions. Six weeks following IPT and the use systemic medications, all clinical parameters improved significantly. Furthermore, lesions including mucosal blisters and desquamations partially recovered, the patient started to perform oral hygiene more effectively.

It was concluded that atraumatic and non-invasive periodontal treatment supported by the use of local/systemic corticosteroid and immunosuppressive medications was efficient on controlling of widespread desquamations and gingival inflammation of PV patients.

Key Words: Corticosteroids, immunosuppressive, nonsurgical periodontal debridement, pemphigus vulgaris.

Introduction

Pemphigus, originated from Greek word ‘pemphix’ (bubble or swelling), is the name of a potentially life threatening autoimmune mucocutaneous disease. The incidence of the disease was reported as 0.1-0.5/100.000 per year in world population.(1) Pemphigus vulgaris (PV) manifests particularly during middle age with an age peak between 4th and 6th decade of life (1). Higher prevalence is reported in Jewish population and Mediterranean countries, particularly (1). The classification of pemphigus is based on the anatomic features of lesions, associated antibody and target antigens (Table 1) (2, 3). The pathogenesis of PV involves immunoglobulin G (IgG) antibodies against desmosome proteins such as desmoglein 3, separating keratinocytes from the basal layer of epidermis (2). PV most commonly affects stratified squamous epithelium (4). Clinically, patients with PV have blister formation and vesiculobullous disintegration of involved areas of skin and mucous membrane (5). In most of the cases oral lesions precede skin lesions. Most frequently affected sites in oral mucosa are reported as labial and buccal mucosa, gingiva and lips (5). Pressing or rubbing on normal looking mucosa

can trigger bullae formation or erosion which is called Nikolsky phenomenon. Nikolsky sign is important in differential clinical diagnosis between PV and other immunopathogenic blistering diseases (5). Diagnosis of PV is based on characteristic clinical symptoms confirmed by histopathological and/or direct immunofluorescence microscopic analysis (6). Mostly, oral lesions heal slowly thus patients with PV have discomfort with eating, drinking swallowing and speaking (6). In treatment of patients with PV, moderate to higher dose of systemic corticosteroids play a crucial role. In some circumstances, immunosuppressive agents such as azathioprine, mycophenolate mofetil, cyclophosphamide and methotrexate as well as cyclosporine and chlorambucil can be added to the treatment (6). Moreover, topical and intra-lesional glucocorticoid applications may be useful for resistant mouth lesions (6). In this case report, periodontal treatment approach including local and systemic medications on histopathologically and immunohistochemically diagnosed PV patient was presented.



Case Presentation

36-year-old female patient was applied to our clinic with intraoral burning and pain complaints. In addition to cutaneous blisters on nose, finger and hand (Figure 1a) intraoral examination revealed severe gingival inflammation, widespread desquamation and ulcers (Figure 1b). PV diagnosis was made according to the histopathological and immunofluorescence evaluation conducted on the biopsy samples taken from the cutaneous and oral lesions. Histological examination with hematoxylin-eosin dye revealed suprabasal acantholysis, neutrophil infiltration in basal membrane and intraepithelial bullae (Figure 1c). Direct immunofluorescence microscopy revealed a fraction of C3 accumulation in basal membrane cell and IgG deposits on epithelial cell surface (Figure 1d).

Patient received IPT including oral hygiene instructions together with mechanical removal of all deposits. During the IPT sessions, local corticosteroid (Nasonex® Aqueous Nasal Spray, Merck Sharp and Dohme, USA) was prescribed for desquamative lesions 4 times per day. She was also prescribed systemic corticosteroid (Prednol®, Mustafa Nevzat, Istanbul) and immunosuppressive (Imuran®, Glaxo Smith Kline, England) by her dermatologist. Before and 6 weeks after IPT, all periodontal parameters; plaque index (PI), gingival index (GI), probing depth (PD), clinical attachment level (CAL) were assessed (Table 2).

Periodontal procedures did not have any negative effect on the lesions. Six weeks after the treatment, all periodontal parameters were improved. Enhancement of the quality of life and patient comfort was provided (Figure 2).

Table 1. Classification of Pemphigus (2, 3).

Type	Anatomic Features	Associated Antibody	Target Antigens
Pemfigus Vulgaris (PV)	Persistent, painful oral lesions;	IgG	Desmoglein 3
Mucosal PV	skin folds are effected;	IgG	Desmoglein 1 and 3
Cutaneous-mucosal PV	vegetans-like; fetid, reddish plaques	IgG	Desmoglein 1 and 3
Pemphigus vegetans			
Superficial pemphigus	Characterized by mainly cutaneous lesion	IgG	Desmoglein 1
Pemphigus foliaceus		IgG	Desmoglein 1
Pemphigus erythematosus			
Endemic pemphigus			
Brazil		IgG	Desmoglein1, desmocollin 1
Tunisia		IgG	Desmogleins 1 and 3
Colombia		IgG	Desmoglein 1
Paraneoplastic pemphigus	Characterized by proliferation of various types of tumors, particularly lymphoid hemopathies	IgG	Desmoplakin I/II, Desmoglein 1 and 3, Envoplakin, periplakin, Antigen 170 and 230 kilodalton
Ig A pemphigus	Exudative lesions with vesicopustules	IgA	Desmocollin 1 and another unidentified antigen
Herpetiform pemphigus	Rosette-like lesions	IgG	Desmoglein 1 and 3
Drug-induced pemphigus	Mainly cutaneous lesions	IgG	Heterogeneous

Table 2. Changes in periodontal parameters before and after IPT.

	Before IPT	After IPT	Changes
Plaque Index (PI)	2.78 ± 0.41	1.25±0.51	-1.52±0.57
Gingival Index (GI)	2.64±0.48	0.45±0.42	-2.18±0.67
Probing Depth (mm)	2.21±0.27	1.84±0.18	-0.36±0.24
Clinical Attachment Level (CAL) (mm)	2.89±0.66	2.68±0.82	+0.20±0.35



Figure 1: **a.** Cutaneous blisters on nose, finger and hand **b.** Intraoral clinical view shows lack of oral hygiene because of pain related to desquamative lesions. **c.** Histological examination; → Suprabasal acantholysis → Neutrophil infiltration in basal membrane → Intraepithelial bullae (Hematoxylin-eosin stain X100) **d.** Direct immunofluorescence showed fraction of C3 in basal membrane cells and IgG deposits on epithelial cell surface (X 200 magnification).



Figure 2: At 6th week follow up intraoral clinical view presents reduced gingival inflammation and healing of PV desquamative lesions

Discussion

In this case report, successfully periodontal treatment of a PV patient was presented. Certain diagnosis of the PV was based on the histopathological and immunofluorescence evaluation in accordance with the literature (1, 7).

The relationship between periodontitis and oral PV is challenged by conflicting results. The lack of correlation between severity of oral lesions and periodontal parameters were reported in studies (8). Thorat et al. showed that in PV patients periodontal parameters such as plaque score, PD and CAL were higher than control group (9). The study by Akman et al. using Community Periodontal Index of Treatment Needs also revealed impaired oral health in PV patients (10). In our case, periodontal status was evaluated before and after the treatment. Initially, the patient was unable to provide adequate oral hygiene due to the painful oral lesions. The severity of the lesions was decreased together with the enhanced clinical parameters.

In the treatment of PV, glucocorticoids have been cornerstone since 1950's (1). Due to the side effects of steroids, some other steroid sparing agents (azathioprine and cyclophosphamide) and intravenous human Ig applications were successfully used for this purpose (6). In our case, in addition to IPT, patient was prescribed for local steroid application to the oral lesions 4 times per day together with systemic corticosteroid and immunosuppressive medication. The improvement in periodontal inflammation revealed by the IPT led to a decrease in the doses of systemically used drugs.

Consequently, successful results can be achieved by reducing inflammation with performing IPT in an atraumatic manner which is supported by corticosteroid or immunosuppressive agents regarding the severity of PV lesions.

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ÖBA: Revisions,

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