Oral Pemphigus Vulgaris: The Importance of Patient’s Adherence

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Abstract

Background: Pemphigus vulgaris (PV) is an autoimmune mucocutaneous disease characterized by intraepithelial bulla formation involving the oral mucosa. The treatment requires long-term corticosteroid at variable doses according to the severity of the lesion.

Case report: A 41-years-old male was referred from the Dermatology Department with oral manifestations of PV. He was treated with systemic methylprednisolone. Oral examination showed severe intra oral PV lesion so that an additional of dexamethasone mouth rinse was prescribed. After 4 weeks of treatment, oral and systemic lesions were relieved. At that time the patient believed he has recovered and decided to stop the medication by himself. Several months later, his skin and oral lesion relapsed with more severe clinical condition than the previous one and he was re-treated with corticosteroid. The severity of PV was suspected by the lack of adherence in medication.

Conclusion: Discontinuation of corticosteroids without a rational reason might influence the recurrence and severity of PV. Good adherence toward corticosteroid therapy is an important factor in PV treatment.

Keywords: corticosteroid, patient’s adherence, pemphigus vulgaris

Introduction

Pemphigus vulgaris (PV) is an autoimmune mucocutaneous disease characterized with flaccid blister and erosion of the skin and mucous membranes. The etiology is still unknown but strongly associated with HLA class II genes. The worldwide incidence is 0.1–0.5 per 100,000. PV can occur in all racial and ethnic groups with the highest incidence in Ashkenazi Jews during 40–60 years old. Histological examination of PV will show acantholysis.

The main therapy of PV is long term corticosteroid which combined with steroid sparing agent. The main goals the treatment is to reduce the symptoms, maintain organ function due to long term corticosteroid or immunosuppressant agent and controls the autoimmune process by induce long period remission and or reduce the flare up. At the end, the treatment should improve the patient’s quality of life.

There is a need for patient’s adherence, in order to reach the successful treatment of long term therapy, that should be applied also in PV therapy. Adherence is more than just performed on taking medication but it also requires patient intervention to be active partner with health professionals. This case report will discuss about the importance of patient adherence during PV treatment.

Case report

A 41 years old male patient was referred to Oral Medicine clinic of International Hospital in Jakarta from Dermatoveneorology with diagnose as PV for 7 months with the biopsy result show suprabasal clefting, row of tombstone and acantholytic cell. Patient confessed the presence of oral lesion prior the skin lesion, but he took self-medication and the lesion healed. But it reappeared since 3 weeks and got worse, although with lessens pain and no swallowing complains at the moment. His oral lesion was also accompanied with blister lesions on his face, back, arm and leg. He admitted that he had delayed his follow up to Dermatology clinic for 4 months. The patient was a high school graduated entrepreneur who supports the family finance. He had no familial history of PV and the disease was suspected to be triggered by psychological stress. The medication from Dermatology department contains systemic Methylprednisolone (MP), Azathioprine, Clindamycin, Ranitidine®, with Calcium and D3 vitamin supplement. His skin lesion was treated with NaCl 0,9% and salicylic acid.

Extra oral examination showed multiple crusted and erosion area on face, chin and neck, with soft bulla presence on his leg. The submandibular lymph nodes were tender on palpation without any pain. There was multiple bleeding crusted and desquamation areas on his upper and
lower lips. Intra oral examination was revealed multiple sloughing areas, ulcers and erosion in all mucosal surfaces (Figure 1).

Oral hygiene was poor with debris, sub and supra gingival calculus and tooth staining. The oral lesion was diagnosed to be intraoral manifestation of PV.

The patient management consisted of communication, information and education (CIE) about oral manifestation of PV and treatment plan. An oral rinse to be swished and spit was prescribed that contained dexamethasone with as much as 6 mg in the morning and 3 mg in the afternoon. The lip lesions were treated with lip ointment mixture that contained antibiotic, corticosteroid, and moisturizer, to be applied thrice a day. The patient was also instructed to brush his teeth twice a day with soft toothbrush.

One week later, on the first follow up visit, the patient experienced new lesion on his fingers and painful oral mucosal lesion for 3 days. He admitted to take his medication according the doctors instruction. Extra oral examination showed black crusts on upper and lower lip. Intra orally, there were no significant difference from the first visit. Similar medication to the prior visit was prescribed to this patient for 2 weeks’ period.

On the next follow-up visit, the patient admitted that there was neither new oral nor skin lesion, and the pain was decreased. Clinical examination showed improved oral and skin lesion. The crusted-on lips were decreased in size while the ulceration and erosions seen to be improved in palate, buccal and labial mucosa. (Figure 2.) The medication prescribed was still the same with previous visit which is oral rinse to be swished and spit was prescribed that contained dexamethasone with as much as 6 mg in the morning and 3 mg in the afternoon. The lip lesion was continued with lip ointment mixture.

The patient was instructed to have a follow up visit the next week but he didn’t come. When contacted by phone, he declared that to be recovered and refused to come for follow up. The dexamethasone oral rinse had not been tapered yet and the patient claimed that he continued to all the prescribed medications as instructed.

The patient was absent from his scheduled follow up visit for about 3 months. Then, his PV lesions relapsed with skin-mouth-eye involvements that required him to be hospitalized. He complained for pain and limited mouth opening. On inward visit, clinical examination showed yellowish-black crusted and bleeding lesions on the lips and circum oral areas, while there was bulla on the right hand and erythematous eye with purulence. Intra orally, there were sloughing and erosion in buccal mucosa and the tongue (Figure 3).
swallowing. The intra oral lesion of mucosa showed less erosion (Figure 5). Skin lesion showed less erosion with squama. At that moment, the patient was discharged from the hospital. For home care, the oral treatment was still maintained as before with the intensification of oral hygiene procedure. The dermatologist prescribed the corticosteroid in tapered dose with supplements but a change of immunosuppressant agent into Mycophenolate mofetil. The patient and his family was also reeducated, then was given oral and written instruction on the use of medication, oral hygiene maintenance and the importance of follow up visit. He was planned to have a follow up visit within a week. But, again, the patient did not come and this time he could not be recalled.

![Image](94x456 to 251x603)

**Figure 5. Last follow up visit, February 9, 2016**

### Discussion

This case report describes a Pemphigus Vulgaris patient who had oral and skin involvement. The oral lesion erupts in the first place but he did not come to dentist at the first eruption, since he did not feel any discomfort at that time. The patient finally came to the dentist after had some oral discomfort and referred from Dermatovenerology department. In most cases, PV lesion appears initially on the oral mucosa, and can be located anywhere within the oral cavity with the most common found in buccal mucosa. The occurrence of oral lesion is related to the circulating auto antibodies against desmosome molecules of Desmoglein 1 (Dsg1) and Desmoglein 3 (Dsg3) which directed majoring Dsg3. Oral mucosa prominently express Dsg3. Auto antibodies circulating induce loss of adhesion cell in epithelial suprabasal and forming of intraepithelial vesicles or bulla.

The patient was treated with systemic corticosteroid for his skin lesion, while the oral lesion was treated with topical corticosteroid. Systemic and topical corticosteroids are the first line therapy of PV. The corticosteroids therapy has important function as anti-inflammatory and immunosuppressant. Although the patient was already given with systemic corticosteroid therapy, but he was still given the topical ones since oral lesions often unaffected by systemic medication. Oral lesion can be managed with topical corticosteroid rinse or cream.

Topical treatments of corticosteroid can speed up re-epithelialization of pemphigus lesions include epidermal growth and decrease disease progression and persistently decreased desmoglein-reactive antibody titer.

The treatment of PV patient requires long term use of immunosuppressant agents. Here in our case, the patient was treated for almost a year. Without any treatment, PV lesions can spread more extensive and life-threatening fatal within 1 to 3 years, because of loss of fluid and protein or opportunistic infection. The wide variety of therapies can be used but no one can guarantee a permanent remission and sometimes recurrence may still happen. The patient was also treated with steroid sparing agents such as azathioprine and mycophenolate mofetil. These are immunosuppressive non-steroidal drugs steroid sparing agents, used to reduce adverse effect of long term corticosteroid without eliminate the beneficial effect of corticosteroid. The long-term use of corticosteroid and its sparing agent can cause various side effects.

In this case, the patient frequently stopped his medication without health professional advice, including his corticosteroid treatment, every time he felt any improvement of his condition. When abruptly discontinued, long term corticosteroid will cause adrenal insufficiency. The patients stopped the medication, and for several times, happened when the corticosteroid doses were not tapered down yet. The dose of corticosteroid can be tapered by 25% if remission is induced and maintained with healing of the majority of lesion.

The relapses of PV lesions happened in this case. The etiology of PV is unknown but strongly associated with genetic some HLA class II genus. Some inducing factors such as drug intake, viral infections, ultra violet radiation, nutrition, chemicals, and emotional stress. The patient in our report was never in complete remission, since the lesion of PV can be declared to have complete remission without treatment when no new lesions appear without therapy for two months. The relapses of this case might happen due to the frequent discontinuation of medication or the presence of inducing factors that had not been identified yet.

The frequent medical treatment discontinuation showed the patient’s poor adherence toward PV medication. Poor adherence may associate with confusion, forgetfulness, language difficulty and feeling good enough to abstain from medication. World Health Organization (WHO) classifies the barrier for medication adherence into 5 categories. These are barriers in socioeconomic factors, healthcare organizations, disease related, therapy-related factors and patient-related factors. In this case, the patient adherence might be related to barrier of patient-related factors. There is also a possibility of economic factor that took a role in this case, since the patient admitted himself as the financial backbone of his family and still had to work even when he wasn’t recovered yet.

During PV treatment, the patient was given oral and written medical information and instruction. That was also to the family during inward treatment period. The content of information includes life-threatening diseases, the recurrences, and the purpose treatment using
corticosteroids and long-term side effects that might occur without medication. The patient was also persuaded to have good adherence and self-care during the medication period but the patient failed to perform good adherence towards dentist instruction. Patient’s health attitude and literacy suspected to be the main factor of non-adherence to complete the medication. Medical adherence also can be influenced by the level of awareness of the health and socioeconomic status. The ability of patients to bond with the doctor’s information may be influenced by the level of knowledge and patient’s expectations. In adherence, patient plays active role as partner and collaborate to dentist about the medical treatment.

On this case report, non-adherence is strongly suspected as a factor that worsened the severity and recurrences of PV. Lack of adherence will have an impact toward the increased of diseases complication and reduced quality of life which resulting higher health care cost. Non-adherence increases the risk of morbidity, hospitalization and mortality with the result on decreasing health. To increase patients’ adherence, dentist’s communication skills and good manners also plays important role. One of communication technique that can be applied by dentist is teach-back method, where the patient or family are asked to show or do the dentist’s information or instructions to ensure their acceptance. The availability of good information, communication and support to patients can increase the engagement and involvement of patients in medical treatment.

**Conclusions**

The long-term use of corticosteroid in Pemphigus Vulgaris treatment requires a good patient adherence. There is a need of good communication between the medical professional and patients/family to gain optimal adherence in order to achieve the best treatment result.

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