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Case Report

Oral Limited Pemphigus Vulgaris-A Case Report

Maryam Jalili Sadrabad¹ and Azra Mohiti²

¹Assistant Professor, Oral Medicine Department, Dental School, Semnan University of Medical Science, Semnan, Iran
²Assistant Professor, Oral Medicine Department, Dental School, Shahid Sadoughi University of Medical Science, Yazd, Iran

*Corresponding Author: Azra Mohiti, Assistant Professor, Oral Medicine Department, Dental School, Shahid Sadoughi University of Medical Science, Yazd, Iran.

Received: March 02, 2020; Published: April 14, 2020

Abstract

Background: Pemphigus vulgaris (PV) is a chronic inflammatory mucocutaneous disease that occurs mostly in the 5 - 6 decade of life and Ashkenazi Jews racial groups. In most cases the disease begins with oral lesions and progresses to other mucosa and skin. Diagnosis of PV includes histopathologic examination of the affected area and confirmation with direct immunofluorescence test (DIF). Now days pemphigus Vulgaris is treated with corticosteroids in combination to other immune suppressor drugs, although consumption of them for long periods of time has its own challenging side effects.

Case Presentation: The patient was a 20-year-old woman who referred to the Oral Medicine Department of Semnan University of Medical Sciences by a dermatologist. Intraoral examinations revealed widespread wound throughout the mouth. The histopathologic report indicates Pemphigus Vulgaris. The patient was instructed to take oral corticosteroid. One month later, the dose of the drug was tapered and no recurrence was observed at the constant maintenance dose for 4 months.

Discussion and Conclusion: As stated PV is an autoimmune disease which are is common in middle age but this patient was only 20 years old which is an uncommon age for PV. It can be debated that in the modern era because of higher stress levels in the society especially women, the incidence of stress related diseases could increase and also the occurrence age could decrease significantly as seen in this case. In our case because there was only oral cavity involvement is seen, low dose of corticosteroids was initiated as treatment in despite of standard high doses. This report showed that low doses of corticosteroids could be used in cases of isolated PV in the oral cavity.

Keywords: Pemphigus Vulgaris; Oral Medicine; Stress

Introduction

Pemphigus vulgaris (PV) is a chronic, autoimmune disease. It has a mucocutaneous origin and defined as a vesiculo bulus disease [1]. The word of pemphigus comes from the greek word “pemphix” with the meaning of blister [2]. Pemphigus is rare and the incidence rate in the world is 0.1 - 0.5 per 100,000 person each year. It occurs in all racial groups but Ashkenazi Jews have a higher prevalence. It is commonly in the fifth and sixth decade of life [3]. It is important to know that in most cases the disease begins with oral lesions and progresses to other mucosa and skin. The initial lesions are blisters which are ruptured quickly and result in mucosal ulcers and erosions [4].

The pathogenesis of pemphigus includes autoantibodies secretion against keratinocyte cell surface desmosomal molecules desmoglein 3 (Dsg3) and desmoglein 1 (Dsg1) which are cell to cell adhesion proteins from cadherin family. The presence of these antibodies

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causes the loss of cell adhesion resulting in vesicle formation in the supra basilar layer of the epithelium [3]. Diagnosis of PV includes mucosal or skin biopsy of the affected area and histopathologic examination in addition confirmation with direct immunofluorescence test (DIF) [4]. Under the microscope we expect to see supralaminar acantholysis and inter epithelial blisters. In the DIF test IgG deposits are seen in intercellular spaces in the epithelium [4]. Nowadays PV is treated with corticosteroids in combination with other immune suppressing drugs [3]. Mortality rates are not high yet because of appropriate response to drugs, even though consumption of corticosteroids for long periods of time has its own challenging side effects.

Case Presentation

The patient was a 20-year-old female who was referred to the Oral Medicine Department of Semnan University of Medical Sciences by a dermatologist. In the review of systems, no related problem was found but the patient had occupational stress because she was a college student. Head and neck examination revealed no pathology and skin was intact. Intraoral examinations revealed a widespread wound throughout the oral cavity that had developed one month ago. The lesion was a diffused map like ulcer located in the left and right buccal mucosa, and ventral surface of the tongue and tissue tags could be seen around it. Desquamative gingivitis was present in both the maxillary and mandibular gingiva. After obtaining informed consent, incisional biopsy under local anesthesia was done and the sample was referred to the oral pathologist. After receiving the pathology report PV was the final diagnosis. The treatment plane was to start with low doses of corticosteroids and increase the dose as required. The patient was instructed to take 20 mg prednisolone per day which included three 5 mg prednisolone tablets (sobhane daru) early in the morning and one tablet late in the evening. After one month the dose of the drug was tapered gradually to 5 mg prednisolone per day. The patient was observed carefully and no recurrence was observed in the maintenance dose of 5 mg for 4 months.

**Figure 1:** Patients oropharyngeal area in the first visit.

**Figure 2:** Patients cheeks showed map like lesions in the first visit.

Discussion

Pemphigus Vulgaris contains a group of rare life-threatening autoimmune organ-specific diseases which causes blisters and erosions in the skin and mucous membrane as vesiculobullous disease [1,5,6]. It affects the quality of life of patients and their families [7].

Pemphigus has 6 different forms consist of vulgaris, foliace, erythromatosis, paraneoplastic, medicamentosa, vegetance, with vulgaris (PV) is one of the most common and important type with 80% prevalence [3].

Recent reports have revealed a very high prevalence of pemphigus in Iran. Several hereditary and acquired factors are supposed relevant to the pathogenesis of the disease [5]. It established that autoreactive IgG antibodies secreted by the different stages of B cell such as centroblasts, plasmablasts and plasma cells attack extracellular adhesions of keratinocyte proteins which possibly influenced by the expression of human endogenous retroviruses (HERVs) and caused the initiation and progression of blister formation. So, measuring of HERVs expression could be used as a probable diagnostic means for recognition of PV and monitoring of the treatment progression [5,6].

In addition, recently it suggested that oxidative stress is responsible for many human diseases and is one of several factors participated in the etiopathogenesis of it [1,8].

Sajedianfard, et al. expressed that PV significantly impressed the "Family Dermatology Life Quality Index". They concluded that education and instructions for family and care providers by several support groups for example "Pemphigus Family Associations" could be effective in improving the quality of life of them. In current study we suggest education of family of the patient can be important for reducing stress and increased quality of life of patients and their family [7].

PV is more common in women in the world and in Iran too [9,10]. The mean age of 42.5 years and predisposition to 4 - 6 decades is established [3]. Oral involvement is common and is seen in 90% [4]. The buccal mucosa, palate, tongue, gums are the most common sites of oral involvement, respectively [3].

PV Clinical diagnosis is done with Nikolsky sign and confirmation with DIF and IIF tests [11].

In the treatment of PV corticosteroids, Azathioprine, Cyclophosphamide, Cyclosporine, Methotrexate, Dapsone and Gold have been used [2].

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Systemic corticosteroids are fast-acting and appropriate for disease control in comparison with azathioprine and mycophenolate mofetil, which have a slower onset [12].

Consist of this study, Gheisari M., et al. stated that steroid pulse therapy has satisfactory effect in treating PV and caused to shorter periods of hospitalization and reduce the long-term high-dose therapy [13].

Although, PV patients with Severe oral involvement may be non-responders, have several side effects or recurrences. In such cases, Rituximab as a new targeted therapy (monoclonal antibody) is suggested to use because of a valid and safe properties to adjuvant therapy in earlier phases of the disease which could decrease the adverse effects of high-dose systemic corticosteroids [14].

Side effects of high dose corticosteroids includes: headaches, insomnia, fluid retention, mood changes, increased appetite and weight, blood pressure, dementia, diabetes, moon face, susceptibility to infection, osteoporosis and suppression of HPA axis [15].

Pemphigus is not a common disease and the possibility of high-level clinical trial studies regarding its treatment regimens is poor and it's regimens usually determined based on small series studies [1,8].

Nevertheless, first-line treatment by corticosteroids is the first line of treatment and commonly used and regularly in the form of oral prednisolone. The evidences for choosing dose is poor; but, 1 mg/kg/day along with antifungal drugs is commonly used (0.5 - 1 mg/kg/day) is suitable for mild and moderate disease [16]. In this study we followed this instruction at initiation and then tapered the dose. It is noted that since the introduction of systemic steroids for treatment, the prognosis of patients with PV has improved significantly [17].

Older age, sex, more than six months between diagnosis and treatment initiation, and more than 10 skin lesions at admission time are associated with worse prognosis in patients with pemphigus vulgaris [17].

PV has a clinical importance because it can lead to high morbidity and mortality if left untreated [18].

Sirois DA claimed that PV diagnosis is often delayed, on average, five doctors and 10 months takes to make an accurate diagnosis [19].

PV has usually initial presentation as oral mucosal ulcers and it is followed by skin involvement with delay. At early stage, patients are more likely to see dentists, oral surgeons, ENT specialists and overall they could not recognize the disease timely because the limited awareness of vesiculobullous diseases due to their rarity but if they referred to oral medicine specialists, it is diagnosed sooner because they are more experienced about oral manifestation of systemic disease.

As stated previously, Pemphigus Vulgaris is an autoimmune disease which is common in middle age but this patient was only 20 years old which is an uncommon age for this group of diseases that commonly occur later in life. It can be debated that in the modern era, because of higher stress levels in the society especially women the incidence of stress related diseases could increase and also the occurrence age could decrease significantly as seen in this case. In our case because there was only oral cavity involvement is seen, low dose of corticosteroids was initiated, although high doses of corticosteroids are the standard treatment. This report showed that low doses of corticosteroids could be used in cases of isolated PV in the oral cavity.

Conclusion

In the modern era the pattern of many diseases are changing and are influenced by stress which could occur in different setting and the treatment protocol should be modified in these cases.

Acknowledgement

We would like to express our special thanks to Dr. Ramin Taheri, Dermatology Specialist, for timely and accurate patient referral.

Conflict of Interest

There is no conflict of interest in this study.

Funding/Support

No funding/support was received.

Bibliography


