Case Report

Oral mucosal changes in pemphigus vulgaris and its treatment: A case report

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Abstract

Pemphigus Vulgaris is an autoimmune disease that causes severe blistering of the skin and mucous membrane which are fragile and breaks away leaving erosions that are extremely painful. Here we report a case of a patient suffering from pemphigus vulgaris with typical oral manifestations. We have reported the effect of treatment after two consecutive recalls at an interval of 1 week where the patient got 100% relief from her previous signs and symptoms.

Keywords: Autoimmune disease, blistering of mucosa, oral ulcers, pemphigus vulgaris, stomatognathic manifestation

Introduction

Pemphigus Vulgaris (PV) is a chronic auto-immune life threatening disease characterized by mucosal and cutaneous blistering. Pemphigus can be classified into six types: pemphigus vulgaris, pemphigus vegetans, pemphigus erythematosus, pemphigus foliaceus, paraneoplastic pemphigus, and IgA pemphigus. Oral mucosal changes are one of the first manifestations. PV affects 1–5 patients per million population per year. The age predilection ranges between fourth and sixth decades of life with a female predilection.[1] Previously mortality rate was around 75% which is presently about 10% except the paraneoplastic pemphigus with a mortality rate of 75%. PV has strong genetic and environmental association and more prevalent in certain ethnic groups such as Ashkenazi Jews, Japanese, and populations from the Mediterranean ancestry.[2] Quality of life is an important subjective tool for evaluating effectiveness of patient care. A study using generic health-related quality-of-life instrument (the SF-36) has shown that pemphigus patients have dramatically reduced quality of life compared to the general population. Depression was present in over 50% of the study population. Patients with depressive traits had worse health status where 70% of the patients expressed enormous shame about their appearance.[3]

Pathogenesis

In PV, the auto-antibodies are produced against the desmoglein protein. It has an adhesive property which helps epithelial cells to be together. Serum antibody responsible for pemphigus vulgaris is IgG. Antibodies to DSG 1 and DSG 3 are mostly responsible for pemphigus disease.[4] PV is caused by antibodies to DSG 3 which is mostly found in the oral cavity while DSG 1 is found over the skin which forms "bullae" a characteristic sign of PV.[5]

Case Report

A 33-year-old female patient reported to the department of oral medicine and radiology with a chief complain of multiple ulceration in the oral cavity for 1 month. On asking she gave history of similar kind of ulcerations 3 years back but did not take

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any treatment during that period and that healed by itself. Past medical history was not contributory. On general examinations, all the vital signs were within normal limits. On extraoral examination, face was bilaterally symmetrical with no TMJ disorder. On intraoral examination there were diffuse ulcerations seen on the right and left buccal mucosa [Figure 1a], [Figure 1b] and on the ventral surface of the tongue [Figure 1c] on the left side. Size of each ulcer was around 1 * 2 cm approximately, shape roughly linear, surface covered by yellowish slough surrounded by erythematous halo. Margins well defined. Blisters were seen on the right and left corners of the lower lip [Figure 1d]. Other intra oral findings included missing with 36, stain and calculus present. Routine laboratory investigation included complete hemogram with SGOT and SGPT were advised. Treatment advised were tablet Livozit 70 mg at a dose of twice daily for 7 days, local application of hexigel three times daily for 7 days and local application of Tess buccal paste four times daily for 3 days followed by three times daily for 2 days, and then two times daily for the next 2 days. Patient was recalled after 1 week. After 1 week, patient reported with the blood investigation reports wherein all the values were within normal limits. Patient got complete relief pain and ulcerations. On examination it was observed that there were no ulcers seen on the right and left buccal mucosa [Figure 2a and b], and on ventral surface of tongue [Figure 2c]. There were also no blisters observed on the lower lip [Figure 2d]. Patient was advised to take tablet livozit 70 mg at a dose of two times daily for 7 days. Patient recalled after 7 days. Patient came for second recall visit after 7 days and patient got 100% relief. There were no evidence of any ulcer on the buccal mucosa [Figure 3a and b], and on the ventral surface of tongue [Figure 3c] and no blister were visible on the lower lip [Figure 3d].

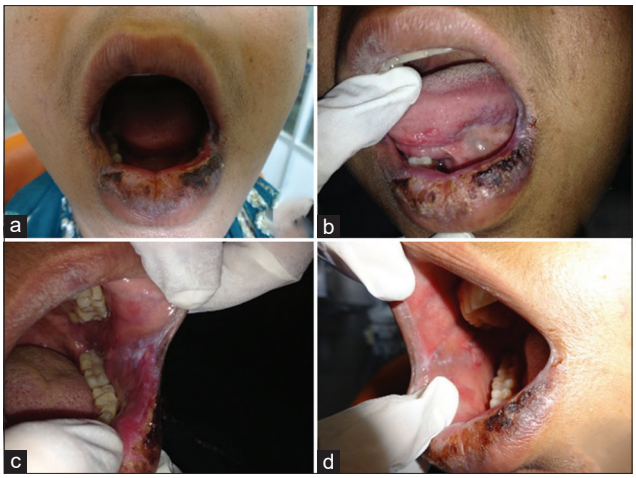

Figure 1: Diffuse ulceration seen on the right buccal mucosa on intraoral vision

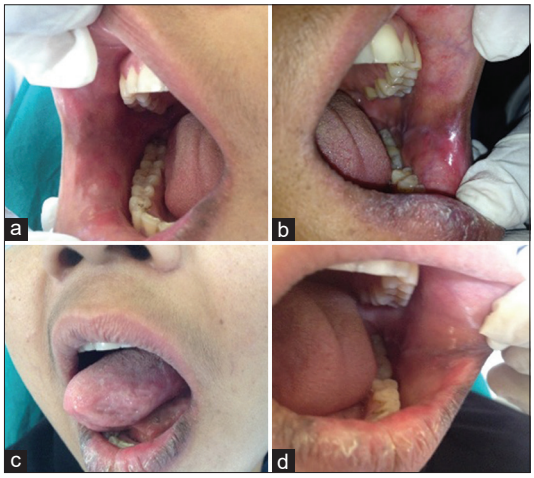

Figure 2: Diffuse ulceration seen on left buccal mucosa on intraoral vision

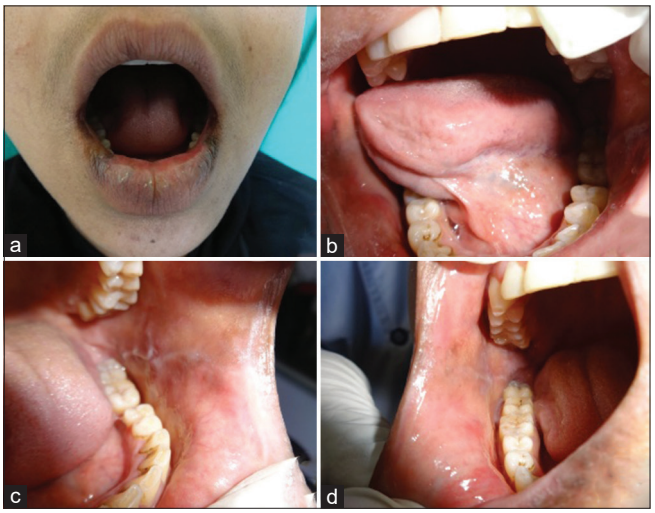

Figure 3: Diffuse ulceration seen on the ventral surface of the tongue on the left side

Discussion

Proper diagnosis leads to proper line of treatment. According to Japanese diagnostic criteria, pemphigus is diagnosed when at least 1 item from every three findings or two items from clinical findings are satisfied. The three groups of findings are: 1. Clinical findings: multiple, easily rupturing flaccid blisters of the skin, subsequent progressive refractory erosions, or crusts after blisters, noninfectious blisters, or erosions of visible mucosa including oral mucosa. 2. Histologic findings: intra-epidermal blisters caused by acantholysis. 3. Immunologic findings: IgG or complement deposition in the intercellular spaces of skin and mucosa detected by immunofluorescence-antibody-assay.[6]

Treatment is administered in two phases: A loading phase, to induce disease remission and a maintenance phase, which is further divided into consolidation and treatment tapering.[7] Moderate doses of oral glucocorticoids without immunosuppressive agents are effective for controlling oral pemphigus.[7] There are studies which states methotrexate can be considered as the first line of drug. Use of pentoxifyline, sulfasalazine, Dapson as adjuvant treatmen.[8] Intra-venous IgG can be administered in the maintenance phase with refractory disease un-responsive to other adjuvant drugs.[9] Pulsed therapy, such as intravenous dexamethasone–cyclophosphamide pulse therapy, has been used in the management of recalcitrant lesions.[10]

The clinical features in our case were suggestive of PV with characteristic mucosal ulcers followed by blistering. The patient was prescribed tablet Livozit at a dose of 70 mg twice daily, local application of hexigel at a frequency of 3 times daily and Tess Buccal Paste at a frequency of four times daily with gradually decreasing the dose. Based on this treatment patient got complete relief from all her previous signs and symptoms.

Conclusion

PV is a debilitating dermatologic condition which has the oral mucosal disease as the first presentation. Proper diagnosis is of prime importance for the proper treatment. The treatment of this disease is a complex procedure and it needs to be more streamlined and advanced. There are various options of treatment which may decrease the likelihood of exacerbating new medical condition secondary to PV.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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