Oral Pemphigus Vulgaris: A Case Report

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Pemphigus vulgaris is an autoimmune mucocutaneous disorder that affects the skin and other mucous membranes of which the intra-oral lesions are first to appear. The occurrence of this disease is very few in the general population. Nevertheless, pemphigus vulgaris is a critical condition because if untreated, it often results in patient's death. This disease is characterized by the production of autoantibodies against intercellular bridges. Most of the patients are misdiagnosed. Hence, it is necessary for the dental professionals to be aware with the clinical manifestations of this disease to ensure early diagnosis and treatment. Here, we present a case of 57-year-old female patient, which was diagnosed as pemphigus vulgaris.

Keywords: Direct immunofluorescence, Mucocutaneous, Pemphigus vulgaris, Tzanck cells

INTRODUCTION

Pemphigus is a severe chronic disease characterized by the presence of vesicles and bullae, small or large fluid-filled blisters that develop in cycles. Pemphigus is derived from Greek word Pemphix meaning bubble or blister. Pemphigus was originally named by Wichman in 1791. The disease commonly affects the skin and mucous membranes. Blistering is due to production of auto-antibodies against desmoglein 1 and 3. Histologically it is characterized by the intraepithelial cleft, tzanck cells and immunologically by circulating immunoglobulin G (IgG) antibody directed against cell surface of keratinocytes.

CASE REPORT

A 57-year-old female patient reported to the outpatient department with the chief complaint of painful oral ulcers since 5 months. The patient complained that the lesions caused discomfort and pain during normal oral function. She gave a history of hyperthyroidism for which she was under medication for 4 years. Personal and family histories were uneventful. On intra-oral examination, multiple ulcers were seen on the cheek and palatal mucosa (Figures 1 and 2) and labial vestibule (Figure 3). No extra oral findings were observed. Cytological smear was taken from the base of the ulcer, and incision biopsy was done. Papanicolaou stained smear showed the presence of acantholytic Tzanck cells (Figure 4). Histological findings revealed the presence of para keratinized stratified squamous epithelium showing intraepithelial separation just above the basal layer. The intraepithelial cleft showed the presence of acantholytic rounded Tzanck cells. A mild to moderate chronic inflammatory cell infiltrate was seen in the underlying connective tissue (Figure 5). Based on the above findings the diagnosis of pemphigus vulgaris was made.

DISCUSSION

In pemphigus vulgaris, lesions at first comprise small, asymptomatic blisters. These are very thin-walled and easily rupture giving rise to painful and hemorrhagic erosions. In most cases, the first signs of disease appear on the oral mucosa. The lesions can occur anywhere within the oral cavity, but mostly found in the areas subjected to frictional trauma, such as the cheek mucosa, tongue, palate, and lower lip. The ulcerations may affect other mucous membranes such as conjunctiva, nasal mucosa, pharynx and genital mucosa. In the present case, the oral lesions were seen mainly in the palate, cheek mucosa and labial vestibule. The diagnosis is generally based on the oral manifestations, while confirmation is done by
reveals IgG or IgM and complement fragments in the intercellular space. In our case, a biopsy of the intra-oral lesions and smear were obtained. The sections were stained with hematoxylin-eosin, and the principal histological characteristics were evaluated.

The blistering that typifies this disease is due to an abnormal production, for unknown reasons, of autoantibodies that are directed against the epidermal cell surface glycoproteins, desmoglein 3 and desmoglein 1.

In a study by Suliman et al. of 588 patients, majority of the patients with pemphigus vulgaris presented with oral lesions. In 2012 Rath and Reenesh reported a case of gingival pemphigus vulgaris with cutaneous lesions.

According to Anuradha et al. histopathology remains the gold standard for the diagnosis and immunofluorescence can provide a valuable additional criterion in the diagnosis.
Oral lesions of pemphigus vulgaris respond partially to topical or intralesional corticosteroids or often immunosuppressants. The use of specific therapies for the underlying disease where available, and often local immunosuppressive treatment, but systemic immunosuppressive therapy notably corticosteroids, is almost inevitably required in pemphigus.\(^7\)

In 2014 Nguyen et al. reported a case of pemphigus on the lateral border of the tongue and showed the importance of local therapy and its potential to induce long-term remission.\(^8\) According to study on 155 patients on immunosuppressive therapy 94 patients developed secondary infections.\(^9\) According to Pavlic et al. low-level laser therapy can provide immediate and significant analgesia in patients with pemphigus vulgaris.\(^10\)

**CONCLUSION**

Pemphigus vulgaris is an immune-mediated mucocutaneous lesion, with oral vesiculo bullous manifestations at the initial presentation. Detailed clinical, histopathological and immunohistochemical examinations are essential for confirmatory diagnosis.

**REFERENCES**


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