

Oral pemphigoid - a case report and a brief review of the literature.

Ioanna Polichroniadou¹, Assya Krasteva²

1. Undergraduate student, Faculty of Dental Medicine, Medical University of Sofia, Bulgaria.
2. D.M.D, Ph.D, Professor of the Department of Diagnostic Imaging and Oral Diagnostic, Faculty of Dental Medicine, Medical University of Sofia, Bulgaria.

Abstract

Aim: The present analysis focuses on examining a case report of a patient diagnosed with a pemphigoid located in the oral cavity. Materials and Methods: A thorough study of the English literature of the electronic databases "Pubmed", "Scopus" and "Google Scholar" was held in order to accomplish our review properly. Case Description: The patient's clinical state was thoroughly studied, along with the histopathological examination findings. Patient's treatment and postoperative course are also within the scope of this analysis. Conclusion: Oral pemphigoid is an autoimmune disease usually located on the gingival. The use of corticosteroids with or without immunosuppressive agents comprises the treatment of choice. In any case, histological examination confirms both the clinical diagnosis and the differential diagnosis between other disorders.

Keywords: pemphigoid, oral lesion, autoimmune disorder

Background

Pemphigoid is defined as an autoantibody-mediated, bullous disorder which may occur in mucosa and skin, leading in two main types of the lesion, the mucosal membrane pemphigoid (MMP) and bullous pemphigoid (BP) respectively(1,2). Oral pemphigoid constitutes a subcategory of MMP and it occurs exclusively in the oral mucosa(3).

A female prevalence has been detected with a ratio 2:1 while most cases occur from 55 to 70 years of life (1,3,4,5,6). As regards MMP, it has been estimated that 85% to 90% of these patients have also lesions in oral mucosa (1,7,8).

The most common intraoral location of the lesion is gingival followed by buccal mucosa, soft and hard palate, and, less commonly, alveolar ridges, tongue, floor of the mouth and lips (1,3,6,7,9).

Although pemphigoid is characterized as a painful disease with healing including scarring, oral pemphigoid comprises no scars or organ disorders (4,6). Albeit, in some cases oral pain, gingival erythema and hyperemia, ulceration, atrophy of buccal mucosa and tongue, halitosis and desquamation may be present, leading in brushing difficulty and low level of oral hygiene (1,6,7,10). Consequently, all periodontal parameters (bleeding on probing, clinical attachment level, probing depth, plaque index, gingival index, recession) appear to be aggravated (11). Periodontal treatment in such cases may be followed by delayed healing whereas secondary management of the disease is needed due to its antibody-mediated nature (7,10). As regards blisters and bullae of the oral cavity these may occur, even though they are rare clinical symptoms of this type of pemphigoid (6). These blisters usually are disrupted quite quickly rather than remaining untouched (7,9).

Case description

A female 57-year-old patient came to the Clinic of Oral Pathology of the Faculty of Dental Medicine of Sofia due to a painful ulceration in the oral cavity. The patient was feeling pain and discomfort for two months before proceeding to examination by an expert. The damage was located at the gingiva, in the region between left upper first premolar and left upper first molar (Fig.1).

Initially, there was a bulla which erupted rapidly in the first two hours and left irregular ulceration covered by pseudomembrane. A thorough examination by a dermatologist reported no skin lesions. Her medical history revealed hypertension which was medically controlled with the use of B-blockers (one tablet per day). Initial diagnosis included pemphigus vulgaris, pemphigoid and oral lichen planus.

Figure1. Initial clinical appearance of oral pemphigoid located at the gingiva, in the region between left upper first premolar and left upper first molar.



After three weeks of local application of clobetasol propionate with commercial label “dermovate”, the oral manifestation disappeared and the gingiva appeared normal. However, during the second oral examination a small bulla was detected, located at the lingual gingival between the right lower first incisor and the left lower first incisor. Biopsy was conducted as well as a histopathological exam with the use of hematoxylin and eosin stain.

The treatment of choice included prescription of interleukin 10 for six months combined with interleukin 1B for one month with commercial label “Guna”. There was no recurrence after treatment while complete rehabilitation of mastication and swallowing was reported.

The final diagnosis established with the use of histopathological exam the presence of pemphigoid.

Discussion

A case of oral pemphigoid is described. The lesion was painful, located at the gingiva, in the region between left upper first premolar and left upper first molar. Histological examination of the damage confirmed the diagnosis of oral pemphigoid.

Histologically, oral pemphigoid is characterized by subepidermal disruption of adherence through discontinuance of hemidesmosome junctions or autoantibody reaction to type VII collagen at the basal membrane (8,12). A number of antibodies reacted to immunoglobulin G (IgG) and complement fraction have been detected in presence of oral pemphigoid (Table1) (1,6). Moreover, a relation between the expression of interleukin-4 (IL-4) and the lack of scarring of oral pemphigoid has been proposed (3).

Table 1. Antigens detected in presence of oral pemphigoid (1,6).

	Antigen
1.	120-kDa antigen
2.	BP antigen 1 (230 kDa)
3.	BP antigen 2 (180 kDa)
4.	168-kDa antigen
5.	Alpha-6 integrin subunit
6.	Epiligrin (laminin-5)
7.	Laminin-6

Diagnosis of oral pemphigoid is usually based on histopathological exam either alone or combined with the direct immunofluorescence (1,7,13). Positive Nikolsky sign constitutes a helpful diagnostic tool; according to this examination, clinician presses with his finger the lesion and segregates normal epithelium from the underlying lamina propria, formatting at once a vesicle or erosion[9]. Differential diagnosis of oral pemphigoid appears to be crucial among other disorders, including lichen planus and pemphigus vulgaris (10).

The treatment of choice of patients with oral pemphigoid is corticosteroid therapy (avoiding high-dose prolonged regimen) either alone or combined with immunosuppressive agents (6). Patients with oral pemphigoid appear to have generally excellent prognosis after treatment (3).

Conclusion

The oral pemphigoid constitutes an autoantibody-mediated disorder, usually growing on the gingival. Treatment of choice includes corticosteroid therapy with or without immunosuppressive agents. The histological examination confirms the diagnosis and it should be conducted carefully, in order to avoid misdiagnosing the oral pemphigoid as other disorder.

References

1. Challacombe S. J., Setterfield J., Shirlaw P., et al. Immunodiagnosis of pemphigus and mucous membrane pemphigoid *Acta Odontol. Scand.*, vol. 59, no. 4, pp. 226–234, 2001.
2. Batts A. F., Jalalat S. Z., Hunter-Ellul L., et al. Exacerbation of bullous pemphigoid after hand, foot, and mouth disease treated with rituximab *JAAD Case Reports*, vol. 2, no. 1, pp. 7–9, 2016.
3. M. Carrozzo et al., "Interleukin-4RA gene polymorphism is associated with oral mucous membrane pemphigoid *Oral Dis.*, vol. 20, no. 3, pp. 275–280, 2014.
4. Maturana-Ramírez A., Adorno-Farías D, Farías-Vergara M., et al. Retrospective Analysis of Reactive Hyperplastic Lesions of the Oral Cavity : Study of 1149 Cases Diagnosed Between 2000 and 2011 , Chile," pp. 103–107, 2015.
5. Brennan P. A., Ammar M, Matharu J. Contemporary management of benign parotid tumours – the increasing evidence for extracapsular dissection *Oral Dis.*, vol. 23, no. 1, pp. 18–21, 2017.

6. Van Dis M.. Oral surgery , oral medicine , oral pathology , oral medicine Editors : Jed Jacobson and Margot Van Dis Periodontal treatment and its effects on glycemc control A review of the evidence vol. 87, no. 3, pp. 311–316, 1999.
7. Sollecito T. P., Parisi E. Mucous membrane pemphigoid *Dent. Clin. North Am.*, vol. 49, no. 1 SPEC.ISS., pp. 91–106, 2005.
8. Hansen M. S., Klefter O. N, Julian H. O., et al. Management of patients with ocular manifestations in vesiculobullous disorders affecting the mouth *Oral Dis.*, vol. 23, no. 7, pp. 849–853, 2017.
9. Schifter M., Yeoh S.-C., Coleman H., et al. Oral mucosal diseases: the inflammatory dermatoses *Aust. Dent. J.*, vol. 55, pp. 23–38, 2010.
10. Hayes E, Challacombe S. J. An Overview of Vesicobullous Conditions Affecting the Oral Mucosa *Prim. Dent. J.*, vol. 5, no. 1, pp. 46–50, 2016.
11. Jascholt I., Lai O., Zillikens D, et al. Periodontitis in oral pemphigus and pemphigoid: A systematic review of published studies *J. Am. Acad. Dermatol.*, vol. 76, no. 5, p. 975–978.e3, 2017.
12. Eversole L. R. Immunopathology of oral mucosal ulcerative, desquamative, and bullous diseases. Selective review of the literature *Oral Surgery, Oral Med. Oral Pathol.*, vol. 77, no. 6, pp. 555–571, 1994.
13. Hayakawa T. et al., Diagnosis of oral mucous membrane pemphigoid by means of combined serologic testing *Oral Surg. Oral Med. Oral Pathol. Oral Radiol.*, vol. 117, no. 4, pp. 483–496, 2014.

Corresponding author:

Assya Krasteva
Faculty of Dental Medicine,
Department of Diagnostic Imaging
and Oral Diagnostic,
Medical University of Sofia