ISSN: 2754-4893

Journal of Dental Science Research Reviews & Reports

SCIENTIFIC Research and Community

Review Article

Open Access

Oral Pemphigus Vulgaris: Case Report and Literature Review

Aicha Ibourk[1]*, Ihsane Ben Yahya[2] and Haitami S[2]

[1]Faculty of Dentistry of Casablanca, Resident, Department of Oral Surgery and Pathology, Rue Abou Alâa Zahr, Casablanca 21100, Morocco

[2]Faculty of Dentistry of Casablanca, Professor and Head of the Department of Oral Surgery and Pathology, Department of Oral Surgery and Pathology, Rue Abou, Alâa Zahr, Casablanca 21100, Morocco

ABSTRACT

Pemphigus is a potentially life-threatening autoimmune disease that causes blisters and erosions of the skin and the mucous membrane. The oral cavity could be involved. These lesions are sometimes isolated and may precede the skin manifestations by several months. This work is aimed at reviewing oral pemphigus vulgaris and its management in oral medicine.

***Corresponding author**

Aicha Ibourk, Faculty of Dentistry of Casablanca, Resident, Department of Oral Surgery and Pathology, Rue Abou Alâa Zahr, Casablanca 21100, Morocco.

Received: July 01, 2024; **Accepted:** July 11, 2024; **Published:** July 18, 2024

Introduction

The pemphigus vulgaris is a disease that causes potential blisters and erosions of the skin and mucous membranes. The autoantibodies play a role in these epithelial lesions. They primarily affect the mucous membranes of patients over the age of 50 years, resulting in the formation of intraepithelial bullae and mucosal ulceration [1].

Pemphigus vulgaris, a deep form of Pemphigus, represents 80% of clinical forms and is characterized by the frequency of associated oral manifestations which are often revealing. These lesions are sometimes isolated and may precede the skin manifestations by several months. Hence the role of the dentist in the diagnosis and treatment of the disease [2]

The objective of this work is to review oral pemphigus vulgaris and its management in oral medicine.

Discussion

Pemphigus is a rare autoimmune condition with elementary lesions of intra-epidermal bullae that mainly affect the oral, genital or ocular mucous membranes and the epidermis [3].

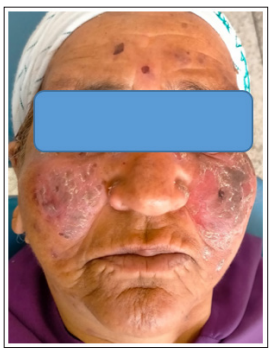

Figure 1: Intra-Epidermal Bullae Lesions of the Epiderme

There are six types of pemphigus: pemphigus vulgaris, pemphigus vegetans, pemphigus erythematosus, pemphigus foliaceus, paraneoplastic pemphigus, and IgA pemphigus. Oral mucosal changes are one of the first manifestations [4].

Pemphigus vulgaris is the most common form of pemphigus, representing almost 80% of cases [5].

The prevalence of this disease is 0.1-0.5 cases per 100,000 inhabitants per year. It touches especially women with male to female ratio of about 1:2 and the fourth to sixth decade of life [6].

The etiology is uncertain. The pemphigus group of diseases are classified as autoimmune diseases since they are manifested by the development of autoantibodies against intercellular substances. Viral infection may also act as predisposing factor in the production of autoantibody. The environmental factors such as foods (garlic), infections, neoplasms, and drugs could be the cause [7].

It is manifested by erosive lesions without a spontaneous tendency to heal. Oral lesions most often precede the appearance of skin lesions. They are manifested by ubiquitous erosions of the oral mucosa.

The bright red color which indicates the erosion is characteristic [8].

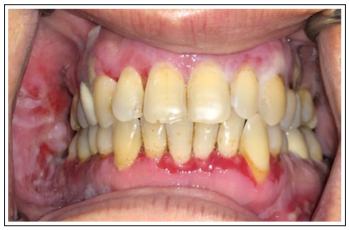

Figure 2: Bright Red Color Erosion in the Mucosa

Cutaneous involvement occurs several weeks or months after the appearance of mucosal lesions. It is characterized by the appearance of flabby bubbles with clear contents. The fragile bubbles, quickly ruptured, give way to erosions surrounded by an epidermal collar. The pressure of the healthy skin induces either a bubble or an erosion: this is the Nikolsky sign [9-11].

Figure 3: Erosions Surrounded by an Epidermal Collar: The Nikolsky Sign

Histopathologically PV is characterized by intraepidermal acantholysis, with basal keratinocytes still attached to the basement membrane zone assuming a characteristic tombstone-like morphology [12].

A blood test is systematically requested, supplemented by an indirect immunofluorescence to search for anti-basal membrane antibodies.

Clinical features, histopathology, and immunological tests confirm the diagnosis of PV [13]. The differential diagnosis of PV includes certain chronic mucodermatoses that present as bullous, ulcerative, or erosive lesions. In rare cases, pemphigus lesions can be confused with ulcerative lesions of Crohn's disease or ulcerative colitis or ulcerative lesions related to dietary deficiencies, such as iron deficiencies (hypochromic iron deficiency anemia), zinc (acrodermatitis enteropathica), folic acid or vitamin B12 (Biermer's anemia) [14].

The purpose of the treatment is to induce disease remission. PV is generally managed with topical, oral, and intralesional corticosteroids. Systemic corticosteroids have been used as the cornerstone of management for PV since the time of their approval in the 1950s [15].

European Dermatology Forum and European Academy of Dermatology and Venereology recommended administration of a higher prednisolone dose (up to 2 mg/kg) if the control of the disease is not achieved with an initial dose of prednisolone of 0.5 mg-1.5 mg/kg/d within two weeks [16].

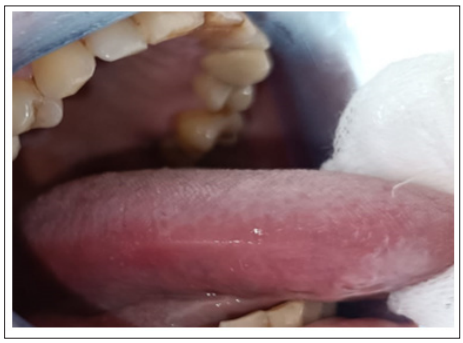

Figure 4: Ulceration of the Tongue

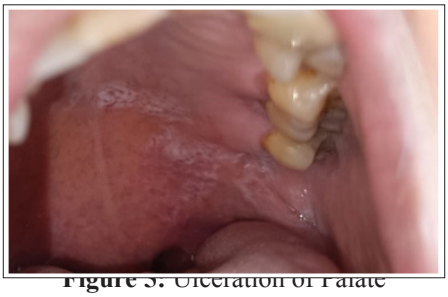

Figure 5: Ulceration of Palate

References

1. Greenberg M, Glick M (2003) Burket's OralMedicine, 10th ed. Philadelphia: JB Lippincott.https://rlmc.edu.pk/themes/images/gallery/library/books/dental/%5BLester_William_Burket,_Martin_S._Greenberg,_Micha(BookFi).pdf.
2. Gharibi A,Khlil N, Chemlali S,Kissa J (2014) Pemphigus vulgaris and oral involvement: Role of the dentist (Case study and literature review). https://www.lecourrierdudentiste.com/cas-clinique/pemphigus-vulgaire-et-atteinte-buccale-role-du-dentiste-etude-dun-cas-et-revue-de-la-litterature.html#google_vignette.
3. Fellner MJ, Sapadin AN (2001) Current therapy of pemphigus vulgaris. Mt Sinai J Med 68: 268-278.
4. Hina H, Pooja K, Vijayta S (2017) A comprehensive review on pemphigus vulgaris. Biomed J Sci and Tech Res 1: 1-4.
5. Endo H, Rees TD, Hallmon WW (2008) Disease progression from mucosal to mucocutaneous involvement in a patient with desquamatous gingivitis area with Pemphigus Vulgaris. Journal of Periodontology 99: 368-375.
6. Ariyawardana A, Tilakaratne WM, Dissanayake M, Vitanaarachchi N, Basnayake LK, et al. (2005) Oral pemphigus vulgaris in children and adolescents: a review of the literature and a case report. Int J Paediatr Dent 15: 287-293.
7. Subadra K, SS, Warrier SA (2021) Oral Pemphigus Vulgaris. Cureus 13: e18005.
8. Vaillant L (1999) Maladie bulleuse autoimmune de la muqueuse buccale. Rev Stomatol Chir Maxillofac 100: 230-239.
9. Bickle K, Roark TR, Hsu S (2002) Autoimmune bullous dermatoses: a review. Am Fam Physician 65: 1861-1870.
10. Cotell S, Robinson ND, Chan LS (2000) Autoimmune blistering skin diseases. Am J Emerg Med 18: 288-299.
11. Kneisel A, Hertl M (2011) Autoimmune bullous skin diseases. Part 2: diagnosis and therapy. J Dtsch Dermatol Ges 9: 927-947.
12. Bystryn JC, Rudolph JL (2005) Pemphigus. Lancet 366: 61-73.
13. Nadia B, Vincent P, Jean-Claude R (2005) Pemphigus vulgaris: update from a clinical case. JADC 71: 667-672.
14. Leung DY, Bloom JW (2003) Update on glucocorticoid action and resistance . J Allergy Clin Immunol 111: 3-22.
15. Hertl M, Jedlickova H, Karpati S (2015) Pemphigus. S2 Guideline for diagnosis and treatment--guided by the European Dermatology Forum (EDF) in cooperation with the European Academy of Dermatology and Venereology (EADV). J Eur Acad Dermatol Venereol 29: 405-414.
16. Aya A, Karim K, Ankur N (2021) Oral pemphigus vulgaris: dentists take-home message. Clin Case Rep 9: e04494.