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JOIS

CASE REPORT

The role of the dental surgeon in the diagnosis and treatment of pemphigus vulgaris: case report

RELATO DE CASO

O papel cirurgião dentista no diagnóstico e tratamento de Pênfigo vulgar: relato de caso

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Abstract

Pemphigus vulgaris is a low-prevalence autoimmune disorder characterized by vesiculobullous lesions affecting the skin and mucous membranes. Lesions in the oral mucosa can be the initial manifestations of the disease and highlight the importance of the dentist in early diagnosis. The objective of this study is to report a clinical case of pemphigus vulgaris with oral lesions in a 44-year-old female patient with no comorbidities. Intraoral physical examination revealed ulcerated lesions throughout the oral cavity, accompanied by bullous lesions, ulcerations, and crusts on the skin. The patient reported having had the lesions for four months, with no conclusive diagnosis. These lesions made eating difficult and resulted in marked weight loss and the need for hospitalization. During hospitalization, the diagnosis was established by the medical and dental team based on clinical characteristics and medical history, followed by histopathological confirmation. Oral lesions were treated with 0.12% chlorhexidine digluconate mouthwash and laser photobiomodulation, combined with systemic corticosteroid therapy, resulting in significant remission. Therefore, dentists' knowledge of the clinical and histopathological characteristics of pemphigus vulgaris is essential for early diagnosis and effective treatment, promoting patient health and quality of life. Further studies should be conducted to optimize management strategies for oral lesions in patients with pemphigus vulgaris.

Keywords: Pemphigus; Hospital Dental Service; Autoimmune Diseases

Resumo

O pênfigo vulgar é uma desordem autoimune de baixa prevalência, caracterizada por lesões vesículo-bolhosas que acometem pele e membranas mucosas. Alterações na mucosa bucal podem ser as manifestações iniciais da doença, evidenciando a importância do cirurgião-dentista no diagnóstico precoce. O objetivo deste estudo é relatar um caso clínico de pênfigo vulgar com lesões bucais em uma paciente de 44 anos, sem comorbidades. Durante o exame físico intrabucal, foram observadas lesões ulceradas em toda a extensão da cavidade bucal, acompanhadas por lesões bolhosas, ulcerações e crostas na pele. As lesões apresentavam evolução de 4 meses, sem diagnóstico conclusivo. A presença de lesões bucais extensas dificultava a alimentação e culminaram em acentuada perda de peso e necessidade de internamento. Durante o período de internamento, o diagnóstico foi estabelecido em conjunto com a equipe de Odontologia, baseada nas características clínicas das lesões e na história médica, seguido pela confirmação histopatológicas. O tratamento das lesões bucais foi baseado em bochecho com digluconato de clorexidina 0,12% e fotobiomodulação com laser, associado a terapia sistêmica com corticosteroides, e resultou em remissão significativa das lesões. Portanto, o conhecimento dos cirurgiões-dentistas sobre as características clínicas e histopatológicas do pênfigo vulgar é essencial para o diagnóstico precoce e tratamento efetivo, promovendo a saúde e qualidade de vida dos pacientes. Outros estudos devem ser conduzidos visando otimizar as estratégias de manejo das lesões bucais de pacientes acometidos por pênfigo vulgar.

Palavras- chave: Pênfigo; Unidade Hospitalar de Odontologia; Doenças Autoimunes.

Introduction

Pemphigus vulgaris is an autoimmune disease with an estimated incidence of 14 cases per million individuals per year[1]. This condition is predominantly observed in adults between 40 and 60 years of age, with a slight predilection for females[2-7]. Despite its low prevalence, pemphigus vulgaris has historically been associated with high mortality rates, which have decreased significantly with the use of appropriate drug therapies[8].

The etiology of this autoimmune disorder is characterized by a type II hypersensitivity reaction, in which autoantibodies specific for desmogleins 1 and 3 (Dsg 1 and Dsg 3), adhesion proteins present in the skin epithelium (Dsg 1) and oral mucosa (Dsg 3), are involved. The interaction of autoantibodies with these molecules results in acantholysis and the formation of intraepithelial clefts. Clinically, acantholysis results in the formation of blisters that rupture and develop into painful erosions and ulcers. In addition to severe pain, the most common symptoms include fever, dysphagia, and weight loss.

The diagnosis of pemphigus vulgaris is carried out through clinical examination, associated with complementary exams such as histopathological analyses or direct immunofluorescence[1-4,9,10]. Another procedure that contributes to the diagnosis of pemphigus vulgaris is the positive Nikolsky sign, characterized by the application of sliding pressure on the affected mucosa. This pressure causes the rupture of intracellular adhesion, leading to the formation of blisters that can easily rupture and generate painful ulcerations[1,4,11,12]. Although not a pathognomonic sign, Nikolsky's sign can contribute to clinical diagnosis of this condition[13].

Treatment is based on corticosteroid therapy and is initiated with high doses, which are subsequently adjusted according to the clinical condition[9,10]. In addition to corticosteroids, treatment of pemphigus vulgaris may include immunosuppressive agents, which are often used in combination to improve therapeutic response[7]. These drugs help reduce immune system activity and control autoantibody production, contributing to effective disease management[4,8,12,14].

Lesions are observed on the skin and mucous membranes, with oral lesions commonly preceding skin lesions. Thus, the dentist plays an important role in early diagnosis and treatment. In the oral cavity, lesions can affect all regions of the mucosa. However, they are more common in the palate, buccal mucosa, labial mucosa, and tongue[11]. Therefore, the objective of this study is to report the case of a patient diagnosed with pemphigus vulgaris presenting manifestations in the oral mucosa and skin. In addition, this study aims to describe the clinical approach and emphasize the role of the dentist in diagnosis and treatment.

Case Report

A 44-year-old female patient with leukoderma was admitted to the hospital complaining of multiple skin lesions. The patient reported that the lesions had been present for four months and that the first lesions were located on the oral mucosa. These lesions then spread to the skin on her upper and lower limbs, chest, and back. The patient reported significant weight loss (20 kg) in recent months due to painful symptoms that made eating difficult. The patient had previously undergone dental and medical evaluation with a prescription for intravenous antibiotics, oral antifungals, and antiretroviral therapy, with no improvement in her condition. At this time, the patient reported using itraconazole to treat skin lesions due to suspected fungal infection.

The intraoral examination revealed the absence of all teeth. The patient reported using a full upper denture, but she was unable to use it due to the painful symptoms of the oral lesions. In fact, it was observed multiple and extensive ulcerated and erosive lesions on the bilateral jugal mucosa, dorsum and ventral surface of the tongue, and upper and lower labial mucosa, with bloody exudate and painful symptoms (Figure 1). The extraoral physical examination also revealed ulcerated lesions with overlapping blood crusts in the perioral region, labial commissures, and vesicles near the labial filter (Figure 1).

Figure 1. Multiple ulcerated lesions with exposed connective tissue on the dorsum of the tongue (A), ventral surface of the tongue (B), lips (C), and labial commissure (C). Lesions with hemorrhagic crusts in the perioral region (D).

The patient presented ulcerated and crusty lesions on the upper and lower limbs, chest, neck, and dorsal region. In addition, bullous lesions were also identified on the lower limbs (Figure 2). Like the oral lesions, these lesions began with the formation of blisters, which later ruptured, resulting in ulcers. Considering the clinical characteristics of the lesions, the possibility of fungal infections was excluded, and antifungal medication was discontinued. Subsequently, a positive Nikolsky sign was observed, suggesting pemphigus vulgaris as a diagnostic hypothesis.

Figure 2. Clinical appearance of the ulcerated and crusted lesions in the chest and neck region (A). Clinical appearance of the vesiculobullous lesions in the lower limbs(B).

In this context, an exfoliative cytology evaluation of the oral lesions was performed to exclude the possibility of fungal lesions. Thus, superficial scraping of the oral lesions was performed with a swab, and the material obtained was rubbed onto a glass slide, stored in 95% alcohol solution, and subjected to histological analysis. In conjunction with this, an incisional biopsy of the skin lesions was performed, since the oral lesions presented more intense painful symptoms. Exfoliative cytology revealed negative results for fungal infection. Thus, Itraconazole (200mg) was suspended and empirical therapy with Prednisone 80mg every 24 hours was initiated, according to the diagnostic hypothesis of autoimmune disease. The histopathological report revealed suprabasal bullous dermatosis with the presence of acantholytic cells. Based on these data, the diagnosis of pemphigus vulgaris was confirmed.

Mouthwash with 0.12% chlorhexidine digluconate was also prescribed twice a day to control biofilm and prevent secondary infections. In addition, antimicrobial photodynamic therapy sessions were performed with the application of 0.01% methylene blue gel on the lesions for five minutes, followed by the application of a red laser (660 nm | 100 mW) with an energy of 9 J for 90 seconds (Laser DUO - MMO®) (Figure 3). Topical use of triamcinolone 1mg/g twice a day was also prescribed.

Figure 3. Antimicrobial photodynamic therapy with application of 0.01% methylene blue gel on the ulcerated lesions in the oral cavity and perioral region (A). Application of red laser (660nm | 100 mW) with a dose of 9J for 90 seconds (B).

Antimicrobial photodynamic therapy was alternated with photobiomodulation therapy (application of red laser – 660 nm, 100 mW, 2J, 20 seconds, spot technique), and the proposed treatment was performed daily throughout the entire period of hospitalization. After 10 days of treatment, the patient reported significant improvement in painful symptoms and remission of oral lesions (Figure 4).

Figure 4. Clinical aspect of the lesions in the perioral region (A) and dorsum of the tongue (B) after 14 days of follow-up

After 10 days of hospitalization, the patient was discharged due to a significant reduction in symptoms and partial remission of the lesions. Treatment with systemic corticosteroids was maintained with a gradual reduction in doses until complete suspension. The patient was kept under weekly dental follow-up for laser photobiomodulation for 2 months until complete remission of the lesions. The patient then continued to be monitored for one year. No recurrence was observed during this period (Figure 5).

Figure 5. Clinical appearance of the perioral region (A), ventral (B) and dorsal (C) aspects of the tongue, and buccal mucosa (D) after one year of treatment.

Discussion

Pemphigus vulgaris is a mucocutaneous disease characterized by vesicular eruptions, blisters, erosions, and ulcerations affecting the mucous membranes and skin. In about 80% of cases, oral lesions precede skin lesions and are commonly located on the palate, buccal mucosa, lips, gums, and tongue. Ulcerated lesions spread rapidly, with intense painful symptoms intense[7,9,10,15]. The clinical manifestations detailed in the case mentioned correspond to these characteristics.

In addition to clinical features, histopathological evaluation is essential for accurate diagnosis, as pathological conditions such as epidermolysis bullosa and Stevens-Johnson syndrome may share some clinical characteristics[16].

The purpose of therapies for pemphigus vulgaris is to relieve painful symptoms by reducing serum autoantibody levels, either directly or through immunosuppression[8]. In the present case, treatment was based on empirical drug therapy with topical and systemic corticosteroids, in addition to photobiomodulation and antibacterial photodynamic therapy for oral lesions, even before confirmation of the diagnosis. This preliminary approach resulted in significant improvement in painful symptoms until confirmation of the diagnosis was obtained.

Among the therapeutic options, oral or intravenous corticosteroids are the first line of treatment[8,9]. In the clinical case mentioned, prednisone was chosen due to its clinical efficacy. The drug administration protocol varies according to the severity of the disease, usually starting with doses of 0.5 to 2.0 mg/kg/day, with

adjustments according to clinical response[6,15]. Treatment with systemic corticosteroids is often started with high doses, which are then gradually reduced[7,15,17].

In oral lesions of pemphigus vulgaris, topical corticosteroid therapy is often used to reduce inflammation and relieve symptoms[10]. Topical corticosteroids, such as triamcinolone, are used to treat these lesions locally [8,15,17]. In fact, this treatment strategy has supported tissue repair of oral lesions and minimized the inflammatory response[1,14,17].

Several therapeutic alternatives are described for the treatment of pemphigus vulgaris, such as anti-CD-20 agents, anti-CD-25 agents, TNF-α inhibition, FAS Ligand Inhibition, FcRn inhibition, BAFF inhibition, Bruton's tyrosine kinase (BTK) inhibition, CAAR T Cells, JAK inhibition, mTOR inhibition, abatacept, IL-4 inhibition, IL-17 inhibition, IL-6 inhibition, polyclonal Regulatory T Cells, and autologous hematopoietic stem cell transplantation[18].

Low-power laser photobiomodulation is also a promising strategy in the management of oral lesions associated with systemic disorders. This therapy promotes cell proliferation and favors the recovery of affected tissues, contributing to a significant reduction in symptoms[7,17]. In the case presented, the proposed treatment resulted in improved oral hygiene and eating ability in the patient due to pain control[19]. It is important to note that, although this approach is beneficial, it does not replace the use of systemic corticosteroids, which are essential for disease control[17].

Conclusion

The reported case shows that the dental surgeon's knowledge of the clinical characteristics of pemphigus vulgaris is essential for early diagnosis of the disease since oral manifestations may be the first evidence of the disease. In the case presented, the patient had had oral lesions for 4 months without a definitive diagnosis, requiring hospitalization to obtain a diagnosis and effective treatment. Thus, the intervention of the dental surgeon in a hospital setting is extremely important for the management of oral manifestations of systemic diseases. In this case, medical interventions associated with integrated dental treatment optimized the treatment.

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