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Relato de caso

Pemphigus vulgaris: case report

Pênfigo vulgar: relato de caso

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Abstract

Objective: to report the clinical case of a patient with pemphigus vulgaris with subsequent secondary infection. **Materials and Methods**: Cross-sectional, descriptive, retrospective and documental case report study. Information was obtained through analysis of the clinical history and complementary exams present in the medical records. **Case report**: a 37-year-old woman attended the hospital service with the presence of bleeding and painful vesicobullous ulcers in the oral cavity, trunk, abdomen, armpits, back, genital region and buttocks, in addition to friable crusts on the scalp. Such lesions appeared abruptly and were preceded by aphthous-type oral lesions lasting five months. The diagnosis was based on the clinical picture and skin biopsy. She underwent treatment with methylprednisolone, prednisone and dapsone pulse therapy. Due to bacterial infection secondary to the condition and its complications, it was necessary to use antibiotic therapy. **Conclusion**: because it is a rare, serious disease, difficult to recognize in the early stages and that can evolve with secondary infections, adequate knowledge about the clinical manifestations, diagnostic methods and treatment of the disease is necessary for the good prognosis and recovery of the patient.
Keywords: Pemphigus Vulgaris. Skin Diseases. Epidermis. Vesicles.

Resumo

Objetivo: relatar o caso clínico de uma paciente com pênfigo vulgar com posterior infecção secundária. **Materiais e Métodos:** estudo transversal, descritivo, retrospectivo e documental do tipo relato de caso. As informações foram obtidas por meio de análise da história clínica e de exames complementares presentes em prontuário. **Relato de caso:** mulher, 37 anos de idade, compareceu ao serviço hospitalar com presença de úlceras vesicobolhosas sangrantes e dolorosas em cavidade oral, tronco, abdômen, axilas, dorso, região genital e nádegas, além de crostas friáveis em couro cabeludo. Tais lesões surgiram de forma abrupta e foram antecedidas por lesões orais do tipo aftosas com duração de cinco meses. O diagnóstico foi feito com base no quadro clínico e biópsia de pele. Realizou tratamento com pulsoterapia de metilprednisolona, prednisona e dapsona. Devido a infecção bacteriana secundária ao quadro e suas complicações, foi necessário o uso de antibioticoterapia. **Conclusão:** por ser uma doença rara, grave, de dificíl reconhecimento em fases precoces e que pode evoluir com infecções secundárias, é necessário um conhecimento adequado sobre as manifestações clínicas, métodos diagnósticos e tratamento da doença para o bom prognóstico e recuperação do paciente.
Palavras-chave: Pênfigo Vulgar. Dermatopatias. Epiderme. Bolhas.

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Introduction

Pemphigus vulgaris is a bullous dermatosis of autoimmune cause[1-3]. The bubbles are fragile, ephemeral and superficial and break quickly, being revealed multiple erosive and exudative areas. In addition, injuries generate a lot of pain, itching and odor [1].

It initially affects the oral mucosa (gums, inner cheeks, tongue, palate). Other mucous membranes may be involved, such as nasal mucosa, pharynx, larynx, esophagus and genital region, being able to stay a few months with blisters and wounds only in the oral mucosas[1-3], before progressing to skin involvement or other mucous membranes[1,3]. Skin lesions can be localized or disseminated, with predilection for the trunk, scalp, armpits, groin and face[1,3-5].

Patients with pemphigus vulgaris produce autoantibodies that act on desmogleins 1 (Dsg1) and 3 (Dsg3), components of desmosomes that act on intracellular adhesion of the epithelium[3]. The result is loss of adhesion between keratinocytes, which leads to the formation of bubbles. Thus, there is the classification of mucous pemphigus vulgaris, with greater dysfunction of Dsg3, and mucocutaneous, in which both Dsg1 and Dsg3 antigens are enveloped[1,3].

Pemphigus vulgaris is rare, severe, tends to occur in both sexes and, in general, affects people between the fourth and sixth decades of life[1,3,4]. In Brazil, the incidence has been increasing with higher prevalence in females[3].

Its etiology is not yet clear, being considered idiopathic[1], however it is known that genetic and immunological factors have a strong influence of susceptibility to affect[3]. A characteristic sign that can be researched is the Nikolsky sign, which appears when, when rubbing or pressing the skin close to the lesion, epidermal displacement occurs[1,3,5,6]. The lesions evolve with discromia without scarring and the disease may present periods of remission and exacerbation[3,5].

The diagnosis should be given as soon as possible through the analysis of the clinical picture and complementary tests, such as histopathological, cytological, immunofluorescence and ELISA[5]. Differential diagnosis should be made with other forms of pemphigus and other bullous diseases, such as acquired bullous epidermolysis, bullous pemphigoid and mucous membrane pemphigoid[3,5].

Pemphigus vulgaris is potentially fatal if there is no adequate treatment, as an extensive area of the skin may lose its epidermal barrier function and lead to secondary infections[3]. The treatment is based on systemic administration of corticosteroids in high doses, and usually indicated prednisone, which has a rapid response. In addition, pulsotherapy with methylprednisolone may be used, with a lower chance of adverse effects. If there is no significant improvement or

contraindication to the use of corticosteroids, the use of adjuvant drugs such as azathioprine and mycophenolate mofetil is indicated[3,5].

The aim of this study was to report the case of a young adult patient with pemphigus vulgaris with secondary complication.

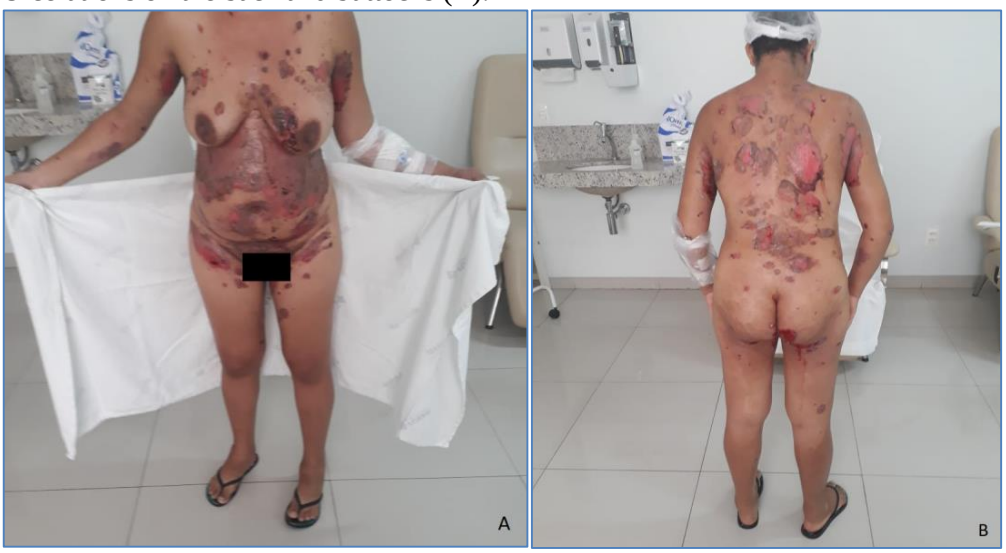
Case report

A 37-year-old female patient attended the hospital service, referred from another service, reporting the presence of very painful ulcerations spread throughout the body, including the oral cavity, which hindered her eating and prevented her from sleeping.

He said that the lesions began to appear in the oral cavity, about five months, with the appearance of canker sores and that, three days ago, there was the abrupt appearance of blisters around the body, which evolved to bleeding and painful ulcers when she was hospitalized. Patient claimed to be previously healthy, without comorbidities and denied family history of similar disease. He reported allergy to dipyrone and denied smoking, alcohol and previous hospitalizations.

On clinical examination, vesicobullous ulcers were found in the oral cavity, trunk, abdomen, armpits, back, genital region and buttocks (Figures 1A and 1B) with the presence of Nikolsky's sign, as well as friable crusts adhered to the scalp. He also had characteristic oral lesions of candidiasis, which had already been treated with the use of nystatin.

Figure 1 - Ulcerations on the trunk, abdomen, arms, genital region and groin (A). Ulcerations on the back and buttocks (B).



Source: images gave by the patient

She underwent biopsy of the arm injury, presenting bullous acantolytic dermatosis with suprabasal cleavage, compatible with pemphigus vulgaris. The indirect immunofluorescence test in a sample of one of the lesions verified the presence of anti-epidermis antibodies in the intracellular substance, confirming the diagnosis.

The admissions tests showed: hemoglobin = 11.2 g/dL; hematocrit = 34.5%; leukocytes = 6,250/mm³; platelets = 407,000/mm³; positive PCR; creatinine = 0.5 mg/dl; potassium = 3.9 mEq/L and sodium = 138/L. Pulsotolonisone was started with methyl (1) and then maintained prednisone (60 mg/day). In addition, a liquid diet was prescribed orally without salt and without acids, nystatin (100,000 U of 8/8h), tenoxicam (20 mg of 12/12h), dexpanthenol (5% topical of 2/2h) and tramadol for pain relief.

After approximately 20 days, reported worsening of the lesions, referring increased pain and emergence of new ulcers. Still claimed general malaise and sweating. On examination, he presented friable ulcers with a fetid odor in the oral cavity, trunk, abdomen, armpits, genital region, back and buttocks, in addition to the presence of hyaline secretion in regions of folds. Secondary infection was suspected.

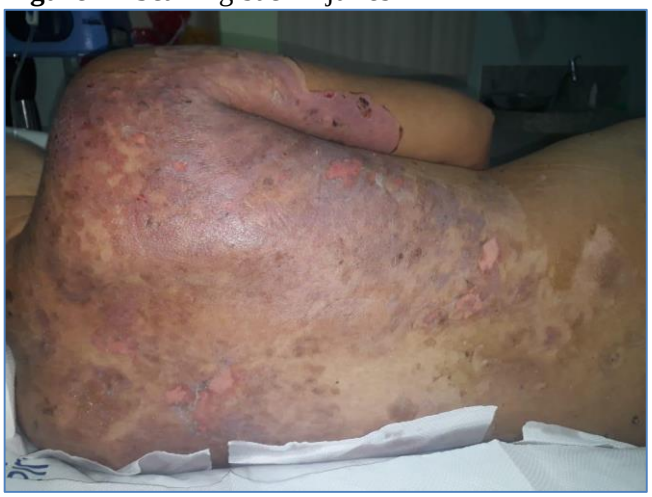
New pulsotherapy was started with methylpredinisolone for three days, maintained prednisone (100 mg/day), started dapsone (100 mg/day) and antibiotic therapy with ceftriaxone and oxacillin. A week later, she presented with slowed speech, tachycardia, tachypnea and 79% desaturation. She reported asthenia and palpitations. New blood count showed hemoglobin = 10.2 g/dL; leukocytes = 16,680/mm³ and platelets = 273,000/mm³. The high flow mask was placed and the antibiotics were replaced by vancomycin and meropenem. Potassium of 2.2 was also obtained and then replaced.

The patient obtained progressive improvement of the lesions, which evolved to scar character (Figure 2), and of the clinical and laboratory changes, receiving hospital discharge. After release, she was referred for follow-up in the dermatology outpatient clinic in a longitudinal manner, when dapsone and predinisone were maintained, with reduced doses.

Ethical care

The project of this work was evaluated and approved by the Research Ethics Committee of the Funorte University Center with approval opinion number: 4,215,751.

Figure 2 - Scarring back injuries



Source: Image gave by the patient

Discussion

Lesions of pemphigus vulgaris usually begin with painful oral ulcers that do not heal quickly and usually precede the appearance of skin vesicles[1,3]. The patient under study obeys this pattern of development, starting the picture with oral lesions with the appearance of aphthous stomatitis, five months before the spread of lesions throughout the body.

The skin of the individual with pemphigus vulgaris undergoes a process of acantholysis, loss of adhesion between keratinocytes. This is due to the production of IgG autoantibodies against structures present in the dermosomes, elements responsible for anchoring and adhesion between the cells of the epidermis. This fact leads to intraepithelial spaces that allow the accumulation of fluid and the formation of fragile bubbles that rupture and form painful ulcers1,3, as presented by the patient in question.

In addition to the clinical analysis, the diagnosis is also made through complementary tests, such as: the histopathological examination, which demonstrates the level of epidermal cleavage of the blister; the cytological, which presents acantholitic keratinocytes which exhibits anti-IgG autoantibodies and the ELISA examination that detects circulating autoantibodies through recombinant Dsg1 and Dsg3[3,5]. In the case described, the diagnosis was made through histopathological examination and immunofluorescence and presented expected results for diagnostic confirmation.

Systemic corticotherapy is the basis of the treatment of pemphigus vulgaris, because it is presented as an excellent anti-inflammatory and immunosuppressive, causing a decrease in the

production of pathogenic autoantibodies. The introduction of this drug as a therapeutic key reduces mortality from 75% to 30%, demonstrating its strong impact[3].

Doses of 1 to 2 mg/kg/day (maximum dose of 100 mg/day) should be given and re-evaluated in 7 to 10 days. Severe patients who do not respond to the treatment described above can be treated with pulsotherapy with methylprednisolone 1g/day intravenously for 3 consecutive days[5,7]. Due to the severity of the patient's condition, the treatment was started with pulsotherapy with methylprednisolone.

The most common side effects of using methylprednisole and prednisolone are: osteoporosis, diabetes mellitus, infections, gastric ulcer, hypertension and subcapsular cataract. These effects directly influence the morbidity and mortality of the disease, requiring monitoring of vital signs and laboratory tests of the patient[3,8]. The aforementioned side effects were not observed in the reported case, but the patient presented immunosuppression, which was expected due to corticosteroid therapy.

Symptomatic, such as tenoxicam (non-steroidal anti-inflammatory drugs) and dexpanthenol (skin emollient), is prescribed for the relief of discomfort and acceleration of the healing process, as well as antifungals, to avoid associated fungal infections[9], and these were used in the case under analysis with good response.

Pemphigus vulgaris a serious disease because, by breaking the protective barrier of the epidermis, it facilitates the entry of pathogens and the emergence of secondary infections that present the risk of developing severely into sepsis[3]. The risk of infection is also associated with the hospital environment and the low immunity provided by high doses of systemic corticosteroids[1,5]. The patient under analysis developed probable secondary bacterial infection after 20 days of treatment initiation. In these situations, it is important to intensify the basic treatment, justifying new pulsotherapy with methylprednisolone and association with antibiotic therapy.

Oxacillin is the antibiotic of choice in severe staphylococcal infections, being part of the group of penicillins resistant to the action of penicillinase produced by community staphylococcus. It is indicated in intense skin conditions, such as bullous impetigo and scalded skin syndrome, as well as cases of osteomyelitis, sepsis, septic arthritis, among others[9].

Oxacillin is active against gram-positive cocci and bacilli, aerobic and anaerobic. However, it does not act against enterococci, and it is not active against gram-negative cocci[9]. Ceftriaxone, in turn, is a cephalosporin of the third generation with the main indication in the treatment of sepsis by

gram-negative bacilli and high antimicrobial potency against enterobacteria[8,9]. Thus, the association of ceftriaxone with oxacillin in the case complemented in the treatment of secondary infection.

The patient presented a drop in general condition and laboratory worsening, requiring antibiotic therapy to be scheduled to cover germs of a hospital nature. Staphylococci, both Staphylococcus aureus and *Staphylococcus epidermidis*, resistant to oxacillin reach 30% to 60% of isolated strains, being called oxacilinarreentes (ORSA - *Oxacillin Resistant Staphylococcus aureus*). These are sensitive to the action of vancomycin, teicoplanin and clindamycin, justifying the substitution of oxacillin for vancomycin.

Meropenem belongs to the class of carbapenems and has activity against gram-negative and anaerobic bacilli, being indicated for serious infections acquired in hospital or immunocompromised patients, reserving itself to situations of greater severity, such as a possible case of sepsis, main cause of death in patients with pemphigus vulgaris [9]. Its clinical indication complements the performance of vancomycin in this new scheme.

Antibiotic therapy should only be indicated in the case of secondary infection, and should preferably be guided by hemoculturalty[3]. Topical antibiotic therapy has no proven efficacy in treating lesions and increases the risk of allergies, sensitivity and bacterial resistance related to its indiscriminate use[1]. Dapsone, prescribed in treatment, is an anti-inflammatory and anti-TNF (Tumor Necrosis Factor) drug that can be used as an adjuvant drug at a dose of 50 to 200 mg/day, oral. Side effects are dependent and reversible doses[3].

Other drugs have demonstrated effectiveness in the treatment of pemphigus vulgaris, such as metrotrexate, cyclophosphamide and plasmapheresis, by acting in the control of circulating autoantibodies and reducing the side effects of corticosteroid[3]. It was not necessary to associate these drugs to the case under analysis due to the good therapeutic response to corticosteroids.

The aim of treatment is to induce and maintain remission. In this first phase, we seek to control the present condition, prevent the emergence of new lesions and promote the re-epithelialization of the injured sites. The maintenance of remission, on the other hand, seeks to minimize the side effects of treatment, such as immunosuppression resulting from the use of systemic corticosteroid[3].

Lesions from pemphigus vulgaris are resistant to treatment and can persist for years, significantly affecting the life of patients[3]. Therefore, periodic follow-up with dermatologist was recommended, even with the control of the condition, since recurrence is frequent.

Conclusion

Pemphigus vulgaris is a rare, severe disease, difficult to recognize at an early stage and may develop with secondary infections. The present case report showed that attention to the initial symptoms of the disease, especially to oral lesions, which may remain for a long period, was necessary. Having knowledge about the diagnosis and proper conduct, especially in the face of secondary infections, is also necessary for the patient to have a good prognosis and recovery.

Contributions

The authors participated in the conception and design of the study, analysis and interpretation of the data, writing or relevant critical review of the intellectual content of the manuscript, final approval of the version to be published, and are responsible for all aspects of the work, including ensuring its accuracy and integrity.

Conflicts of interest

The authors declare that there is no conflict of interest.

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