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Penfhigus Vulgaris: Report Of A Clinical Case

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ABSTRACT

The current article analyzes the topic of pemphigus vulgaris, which is described as an autoimmune disease affecting the skin and mucous membranes, characterized mainly by the formation of superficial blisters on the skin and oral mucosa. The objective was to present a case study of a patient with pemphigus vulgaris. A qualitative and descriptive investigation was carried out in this study. The 40-year-old male patient with pemphigus vulgaris had a complex but satisfactory evolution. Following an exacerbation in July 2023 and complications such as oral ulcers and ocular issues, he was hospitalized again. Aggressive treatment was initiated with medication adjustments, resulting in significa[1]nt improvement with remission of skin lesions, pain control, and no signs of infection. After multidisciplinary follow-up and treatment, he was discharged on November 8, 2023, showing a positive response and improved quality of life. This case highlights the importance of a comprehensive approach in pemphigus vulgaris treatment and the need for ongoing monitoring to prevent relapses and future complications.

Keywords*: pemphigus vulgaris, skin disease, autoimmunity, skin lesions, treatment.*

INTRODUCTION

The current article discusses the topic of Pemphigus vulgaris or pemphigus vulgaris, which is described as an autoimmune disease that affects the skin and mucous membranes, with an incidence ranging from 0.5 to 3.2 cases per 100,000 inhabitants per year. Several types of this disease are identified, with pemphigus vulgaris being the most severe in terms of progression. However, in clinical terms, pemphigus vulgaris is mainly characterized by the formation of superficial blisters on the skin and oral mucosa.(1)(2,3)

Among the most prominent clinical features are the presence of soft blisters that rupture easily, leaving painful erosions. According to a study, it was observed that this disease initially affects the mucous membranes in 41.7% of cases, involving both the mucous membranes and the skin in 37.4%, and manifests itself only with skin involvement in 20.4%. This is due to IgG autoantibodies against desmoglein type 1 and 3 related to mucocutaneous pemphigus vulgaris.(3,4)

The most distinctive histological alteration consists of acantholysis, which leads to the formation of intraepidermal phlyctenae. Diagnosis is based on clinical, pathological, and immunological criteria, where the results of biopsy of the affected skin and immunofluorescence or ELISA tests reveal the presence of intraepidermal deposits of immunoglobulins (IgG) types 1 and 4.(4)

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Diagnosis is based on skin biopsy for microscopic examination with hematoxylin-eosin and direct immunofluorescence (5). Without treatment, this disease can be life-threatening. Prior to the introduction of systemic glucocorticoids (GS), mortality ranged from 60% to 90%. At present, with the use of GS in combination with GS-sparing drugs, the mortality rate has been reduced to 10 . The initial dose of GS is determined empirically, based on clinical experience, and is usually adjusted according to the severity of the case, with a dose range between 0.5 and 2 mg/kg/day. Sparing drugs, such as methotrexate, azathioprine, mycophenolate (MM) mofetil, and rituximab, improve treatment efficacy, reduce the need for glucocorticoids, and prevent potential adverse effects of these medications (6)(7).
The study developed by Villalba et al. through a case report of a 56-year-old patient from the city of Quito, in which a patient with lesions on the face and scalp is described, who was diagnosed with this disease both clinically and through histological examination. Treatment included the use of oral corticosteroids and immunosuppressants, which resulted in a remarkable clinical improvement for the patient.(9)
It was also established in a bibliographic study carried out by Barrera et al. Early treatment, accurate diagnosis of mild forms of the disease, and use of corticosteroids were found to be key factors in improving the prognosis of patients with pemphigus vulgaris. Thanks to the widespread use of corticosteroid therapy, the mortality rate has decreased significantly. Despite this, the main cause of mortality in this disease continues to be of infectious origin, in many cases associated with complications of treatment with corticosteroids and immunosuppressants.(10)
Finally, the study developed by Álvarez et al. We present a case of a 9-month-old male patient who was admitted to the Pediatrics Department. In this case, pruritic erythematous plaques with yellowish discharge were observed, which was consistent with a diagnosis of bullous pemphigoid. The patient received treatment and presented a favorable clinical evolution.(11)

Therefore, the present study focused on conducting a clinical study of a 40-year-old patient diagnosed with pemphigus vulgaris in the Mexican context. The only available studies focus on clinical cases and one cohort case, which shows the need to develop updated research on this type of disease in our region.

MATERIALS AND METHODS

In the present study, a qualitative descriptive research was carried out, focusing on a clinical case study of pemphigus vulgaris. The methodology was based on data collection from two main sources. First, a comprehensive review of the scientific literature related to pemphigus vulgaris was conducted, using reliable medical and scientific databases such as Scielo, Pubmed, Scopus, Proquest, and Redalyc. (12) (13) (14)

Specific keywords such as pemphigus vulgaris, skin disease, autoimmunity, skin lesions, treatment, among others, were used to identify and review scientific articles, previous case studies, and any relevant information about this disease. The second phase of the study consisted of the detailed collection of information from the selected patient with pemphigus vulgaris, including their complete medical history, test results, diagnosis, previous and current treatments, as well as any relevant information about their clinical evolution. All data was obtained directly from the patient or their legal representative, ensuring confidentiality and respect for their privacy at all times.

In this case report study, pertinent ethical considerations have been strictly followed, including obtaining informed consent from the patient and anonymizing information to protect patient privacy. The confidentiality of the data has been guaranteed and the case has been presented honestly and objectively, with a clear educational and scientific purpose. In addition, the corresponding ethical approval has been obtained, ensuring that the study complies with the established ethical standards. These ethical measures have been implemented to preserve the integrity and respect for the patient, as well as to contribute ethically to the advancement of medical knowledge. (15)

CLINICAL CASE PRESENTATION

A 40-year-old male patient, with no history of vaccinations or recent infections, taking medications or dietary supplements. He began his condition in February 2023, with dermatosis that affected the head in the left parietal region consisting of multiple vesicles, blisters and erosions of different sizes, with Nikolsky positive, which spread to the face in the right zygomatic region and anterior thorax, accompanied by pain and later with impetiginization of the same, for which he required hospitalization.

Management with steroids and steroid sparing was performed, and after clinical improvement, medical discharge was indicated one month later with azathioprine 50mg every 12 hours, prednisone 50mg every 12 hours, leflunomide 20mg every 8 hours and losartan 50mg every 12 hours due to hypertension secondary to corticosteroid use. Surveillance was continued by dermatology outpatient clinic, with a gradual decrease in systemic steroids, reaching a prednisone dose of 25 mg every 24 hours over a period of two months. In July 2023, after the reduction of corticosteroids, the patient presented an attack on the general condition and exacerbation of the skin lesions with myelic crusts at the level of the left temporal region, approximately 10cm, pruritic and scaly, he started a regimen with ceftriaxone 1 g every 12 hours, without improvement of the condition. The lesions extend to the scalp, face and neck, with the presence of macerated areas in folds and chills, so the infectious disease service was consulted and trimetropin was added to treatment with sulfamethoxazole 80mg/400mg every 12 hours for 10 days. On August 2, the lesions extended to the anterior aspect of the thorax, hands, groin, buttocks including the intergluteal line, knees and dorsum of the feet, with subsequent loss of liquid content and detachment of the epidermis, he presented lancinating pain with intensity of 8/10 on the analog scale of pain, exacerbated with movement and touch, decreased with analgesics and absolute rest was eradicated. On August 15, ulcers in the oral cavity, conjunctival hyperemia, chemosis, purulent discharge from both eyes, corneal ulcer of the right eye, photophobia, increased pain in dermal lesions and areas of alopecia that were decided to be hospitalized again were added (Figures 1 and 2).

Laboratory studies showed the presence of leukocytosis at the expense of neutrophilia, preserved renal function, mild isoosmotic euvolemic hyponatremia, hypoalbuminemia, hypertransaminasemia, and elevated GGT. Skin biopsy of the back of the right hand reported lichenoid reaction. Due to the extent of the lesions, it was thought to be a paraneoplastic syndrome, so CT scans of the skull, thorax, abdomen and pelvis were performed with a report in normal limits.

Figure 1. Dermatosis in the left parietal region

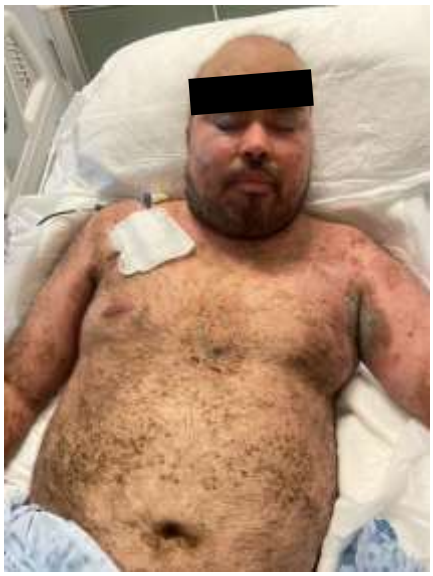
Note. The figure shows a male patient with pemphigus vulgaris.

Figure 2. Spread to cervical, dorsal and anterior chest

Note. The figure shows the spread of the lesions to the back of the body.

Throughout his stay in Internal Medicine, the antibiotic therapy scheme was scaled to vancomycin for 15 days, then with ceftacidim for 7 days, without superinfection of the lesions, he was managed with an initial dose of prednisone 100mg every 12 hours, azatriopine 50mg every 8 hours, methotrexate 7.5mg every 7 days, folic acid 5mg every 24 hours, In addition, he presented hyperglycemia secondary to corticosteroids and was managed with NPH insulin, as for ophthalmic lesions he was managed with 2% hypromellose, ophthalmic ciprofloxacin in the right eye on October 25, 2023, right eye surgery was performed for the perforated corneal ulcer and scleral patch plus conjunctival coating was placed. Patients with significant improvement of the lesions, a gradual decrease of the corticosteroid was initiated until reaching 50mg every 24 hours without presenting new lesions or exacerbation of existing ones. In November 2023, he received 1g of rituximab without adverse reactions (Figure 3), and was discharged on November 8, 2023.

Figure 3. Dermatological lesions in remission



Note. An improvement in clinical characteristics can be seen in the figure after 9 months of treatment.

DISCUSSION

The case report by Villalba et al. (9) and the bibliographic analysis by Barrera et al. (10) highlight the relevance of early diagnosis and appropriate treatment in the prognosis of patients with Pemphigus vulgaris.

The clinical case of the 40-year-old male patient coincides with the clinical characteristics described in the literature, including the presence of blisters on the skin and mucous membranes, as well as the exacerbation of the disease with involvement of the oral cavity and other systemic symptoms. These findings are consistent with the study by Villalba et al. (9), where a patient with lesions on the face and scalp was described.

Regarding therapeutic management, the use of steroids and steroid-sparing drugs in the 40-year-old patient reflects the treatment strategy recommended in the current literature, as evidenced in the study by Barrera et al. (10). The initial positive response to treatment and the subsequent exacerbation of the disease are common aspects in the evolution of patients with Pemphigus vulgaris, as observed in the case presented.

The hospitalization of the patient and the escalation of the therapeutic scheme with the addition of antibiotics and other medications, as described in the patient's study, highlight the complexity and need for a multidisciplinary approach in cases of Pemphigus vulgaris, in line with the recommendations of Barrera et al. (10) for the management of complications and possible secondary infections.

CONCLUSIONS

The evolution of the 40-year-old male patient with pemphigus vulgaris has been complex but satisfactory so far. After presenting an exacerbation of the dermatosis in July 2023 and subsequently developing complications such as corneal ulcer, development of systemic arterial hypertension and hyperglycemia secondary to the use of corticosteroids, the patient was hospitalized again. Laboratory studies revealed alterations compatible with the clinical picture observed, and aggressive management was initiated that included adjustments in antibiotic therapy, immunosuppressants, and symptomatic treatment. Throughout his stay in Internal Medicine, a remarkable clinical improvement was observed, with remission of skin lesions, pain control and absence of signs of superinfection. After careful follow-up

and multidisciplinary treatment, the patient was discharged on November 8, 2023, showing a positive response to treatment and a better quality of life.

The lack of specific studies on pemphigus vulgaris represents a significant limitation in the understanding and management of this autoimmune disease in the local population. The scarcity of research and accurate epidemiological data makes it difficult to identify risk factors, implement prevention strategies, and develop treatment protocols tailored to the needs of these patients. Therefore, it is imperative to foster clinical and epidemiological research in the country, as well as to promote interdisciplinary collaboration between health, academic and government institutions to comprehensively address pemphigus vulgaris and improve the quality of life of affected patients.

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