

# Pemphigus foliaceus, seborrheic variety, case report

DOI: 10.5377/alerta.v6i2.16217

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## Abstract

**Case presentation.** a 48-year-old male with no known medical history who presented multiple lesions in the form of easily desquamative erythematous and crusted plaques, initially on the anterior thorax, which spread over the face and scalp without affecting the mucous membranes. **Treatment.** In-hospital management was mainly based on topical and systemic steroids, the management of infections superadded to the dermatologic lesions, and psychological support for the patient. A skin biopsy was taken where acantholysis was evidenced, confirming the autoimmune disease diagnosis. **Outcome.** After treatment, the multiple scaly lesions were reduced, the local infection was controlled, and the patient's skin recovered although it still had scars, its functions were restored.

## Keywords

Pemphigus, acantholysis, autoimmune disease.

## Resumen

**Presentación del caso.** Se expone el caso de un paciente masculino de 48 años de edad, sin antecedentes médicos conocidos, que presentó múltiples lesiones en forma de placas eritematocostrosas fácilmente descamativas, inicialmente en tórax anterior, que se esparcían sobre el rostro y cuero cabelludo sin afectar las mucosas. **Intervención terapéutica.** El manejo hospitalario se basó fundamentalmente en el uso de esteroides tópicos y sistémicos, así como el manejo de las infecciones sobreagregadas a las lesiones dermatológicas y el apoyo psicológico del paciente. Se tomó biopsia de piel donde se evidenció la presencia de acantólisis, confirmando el diagnóstico de esta enfermedad autoinmunitaria. **Evolución clínica.** Luego del tratamiento se logró una reducción de las múltiples lesiones descamativas, el control de la infección local y la recuperación de la piel del paciente, la cual a pesar de aún presentar cicatrices se encontraba con sus funciones restituidas.

## Palabras clave

Pénfigo, acantólisis, enfermedad autoinmune.

## Introduction

Pemphigus (from the Greek "pemphix" meaning blister) is a group of autoimmune diseases that produce blisters on the skin and mucous membranes due to the action of autoantibodies on specific proteins of the hemidesmosomes<sup>1</sup>. Likewise, it generates a loss of adhesiveness between epidermal cells, histologically characterized by the formation of intraepidermal blisters, a process

known as acantholysis<sup>2,3</sup>. The classic definition of pemphigus describes it as a severe, rare disease with a chronic and aggressive course. Continuous and systematic treatment is essential to avoid its lethal evolution<sup>4</sup>.

Three main types are distinguished: pemphigus vulgaris, paraneoplastic pemphigus, and pemphigus foliaceus, according to their clinical and histological manifestations and the type of proteins involved<sup>5,6</sup>.

## OPEN ACCESS

### Pénfigo foliáceo, variedad seborreica, reporte de caso

#### Suggested citation:

Linares Rosales CS, Brizuela Alfaro OD, Arévalo Mojica IC. Pemphigus foliaceus, seborrheic variety, case report. Alerta. 2023;6(2):113-118. DOI: 10.5377/alerta.v6i2.16217

#### Received:

May 14, 2023.

#### Accepted:

June 27, 2023.

#### Published:

July 20, 2023.

#### Authors contribution:

CSLR<sup>1</sup>: study conception, data collection, data management and analysis, writing, revising and editing. ODBA<sup>2</sup>: manuscript design. ICAM<sup>3</sup>: literature search, writing, revising and editing.

#### Conflict of interests:

The authors declare there are no conflict of interests.

The two most common types are pemphigus vulgaris and pemphigus foliaceus<sup>3</sup>. These two groups differ clinically and histologically, and by their autoantibodies. The former is the more severe form and occurs more frequently in India, south-eastern Europe, and the Middle East<sup>7</sup> and is characterized by mucosal involvement. Pemphigus foliaceus manifests mainly on the skin, in the form of well-defined erythematous or erythematous-costly plaques, with a seborrheic appearance and distribution (face, neck, and trunk), where the mucous membranes are generally not affected<sup>6</sup> and in some cases it is characterized by a burning sensation on exposure to the sun, *fogo selvagem* variant<sup>8</sup>.

The diagnosis of this type of lesion is based on a combination of clinical and histopathologic findings and is achieved through direct immunofluorescence techniques, mainly performed on perilesional biopsies of affected tissue<sup>3</sup>.

Worldwide, the annual incidence is estimated at 21.7 cases per million population. Although it is a rare disease, it is the most frequent form of autoimmune blistering disease. It has been observed in all ages, with a predominance in older adults<sup>1</sup>. Furthermore, although it occurs in both sexes, it has a slight predominance in women. Mortality has been reported at 17.7 %, mainly related to sepsis<sup>9</sup>.

Pemphigus has an estimated incidence of two patients per million inhabitants per year in central Europe. In the Americas region, pemphigus is endemic in Colombia and Brazil. However, pemphigus has been reported in young adult patients and children, mainly in El Salvador and other Latin American countries such as Brazil, Colombia, Paraguay, and Peru; thus, the importance and relevance of case documentation<sup>10</sup>.

## Case presentation

A 48 year old male patient with no known medical history, with a history of one month of observing the appearance of small blisters in the anterior region of the thorax with a progressive increase in number and size until rupture, with serous discharge, moderate pain and crusting, subsequently, the lesions spread to the face and scalp, due to this, he consulted a private physician who indicated treatment with topical neomycin bacitracin.

The patient did not notice any clinical improvement, so he consulted at the emergency unit of San Juan de Dios National Hospital Santa Ana. He denied symptoms such as fever and pruritus. Neither relevant

pathological history was recorded nor contact with people with similar symptoms. He denied having medications, drugs or other substances prior to the clinical record. On physical examination, the patient was alert, oriented and multiple erythematous plaques with irregular, well-defined borders, with melic crusts were observed on the anterior thorax, scalp and mandibular region, without mucosal involvement (Figure 1).

Blood pressure of 120/70 mmHg, heart rate of 76 beats per minute, respiratory rate of 16 per minute, oxygen saturation of 97 %, temperature of 37.1 degrees Celsius.

Laboratory tests showed leukocytosis with a predominance of neutrophils (Table 1). The presumptive diagnosis was over-infected pemphigus vulgaris; therefore, hospital admission was decided, and treatment was started with ciprofloxacin 0.2 grams intravenous every twelve hours and ampicillin plus sulbactam 3 grams intravenous every six hours. After five days, hydrotherapy was started, which after 48 hours, presented sphacelation of the lesions with increased pain and discomfort of the patient (Figure 2). So these signs of clinical deterioration led to the decision to refer the patient to the regional hospital, where they described the patient with multiple dark brown lesions, crust-like with serosanguinolent discharge, erythematous skin-base, easily scaly, on the anterior and posterior thorax, face (beard and eyebrow area) and scalp. The mucous membranes were intact, with no other abnormal findings on physical examination.

Laboratory tests revealed mild leukocytosis, predominantly neutrophils; the rest of the tests were within normal values (Table 1). Chest X-ray and electrocardiogram showed no abnormalities (Figure 3). The patient was admitted to the hospital for suspected pemphigus and started treatment with the following medications: intravenous methylprednisolone, 60 mg every six hours for five days; betamethasone 0.1 % combined with neomycin 0.5 % and bacitracin 0.5 % every day. The patient had completed seven days of double antibiotics in the hospital; therefore, no antibiotics were prescribed.

## Treatment

During the patient's hospital stay, a multidisciplinary approach was carried out, including psychological, dermatological, and surgical care; since the presence of dermatological lesions was accompanied by depressive symptoms and anhedonia, it was decided to manage the patient by psychology with non-directive verbal intervention therapies

every 48 hours. A presumptive diagnosis of pemphigus foliaceus was established by dermatology evaluation to rule out seborrheic dermatitis; in addition, daily healing of the lesions was indicated. After three days, the lesions were debrided, and samples were taken for skin biopsy and culture of the secretions (Figure 4). After five days of intra-

venous steroids, prednisone 50 mg orally was started for one month.

The culture isolated *Klebsiella pneumoniae ssp. pneumoniae*; for this reason, ten days of antibiotic therapy were completed with meropenem 1 g intravenously every eight hours, according to the result of the antibiogram.

**Table 1.** Historial de exámenes de laboratorio

	ER LNH*	ER SJDDNHSA**	Within 14 days of entry	Within 28 days of discharge
Leukocytes	12.53 10 <sup>3</sup>	12.75 10 <sup>3</sup>	12.34 10 <sup>3</sup>	10.65 10 <sup>3</sup>
Neutrophils	69.1 %	83.2 %	68.6 %	85.1 %
Lymphocytes	20.6 %	13.2 %	16.8 %	10.0 %
Platelets	502 10 <sup>3</sup>	223 10 <sup>3</sup>	480 10 <sup>3</sup>	333 10 <sup>3</sup>
Hemoglobin	13.0 g/dL	13.4 g/dL	12.9 g/dL	12.5 g/dL
Hematocrit	37.3 %	39.7 %	36.0 %	37.2 %
Mean corpuscular volume (MCV)	82.7 fL	87.3 fL	82.9 fL	86.9 fL
Mean corpuscular hemoglobin (MCH)	28.8 pg	29.5 pg	29.7 pg	29.2 pg
C-reactive protein	116.05 mg/dL	-	156.9	-
Albumin	2.8 g/dL	2.8 g/dL	3.1 g/dL	3.3 g/dL
Urea Nitrogen	17 mg/dL	22 mg/dL	22 mg/dL	30 mg/dL
Creatinine	1.26 mg/dL	0.85 mg/dL	0.98 mg/dL	0.84 mg/dL
Corrected calcium	8.3 mg/dL	8.3 mg/dL	8.2 mg/dL	8.9 mg/dL
Phosphorus	2.55 mg/dL	3.33 mg/dL	4.28 mg/dL	2.43 mg/dL
Magnesium	1.7 mg/dL	1.5 mg/dL	2.0 mg/dL	1.9 mg/dL
Sodium	137 mEq/L	136 mEq/L	132 mEq/L	137 mEq/L
Potassium	4.3 mEq/L	4.1 mEq/L	4.3 mEq/L	4.2 mEq/L
Chlorine	98 mEq/L	92 mEq/L	88 mEq/L	100 mEq/L
Prothrombin time	11.1 s	-	-	-
Partial thromboplastin time	28.5 s	-	-	-
Fibrinogen	454.2 mg/dL	-	-	-
International Normalized Ratio (INR)	1.03	-	-	-
HIV	-	-	Non-reactive	-

\*ER LNH: Emergency Room Local National Hospital

\*\* ER SJDDNHSA: Emergency Room San Juan de Dios National Hospital Santa Ana



**Figure 1.** Patient admission status

## Outcome

After 28 days of hospital stay, there was a notable improvement in the dermatological lesions (Figure 5), as well as in the patient's mood; consequently, it was decided to discharge him from the hospital with the plan to continue with betamethasone 0.1 % daily and prednisone 5 mg orally every day for three months.

## Clinical diagnosis

The diagnosis of pemphigus was confirmed through the result of the skin biopsy, which revealed that the epidermis presented a superficial epidermal blister with inflammatory cells corresponding to neutrophils, acantholysis in the superficial layer and

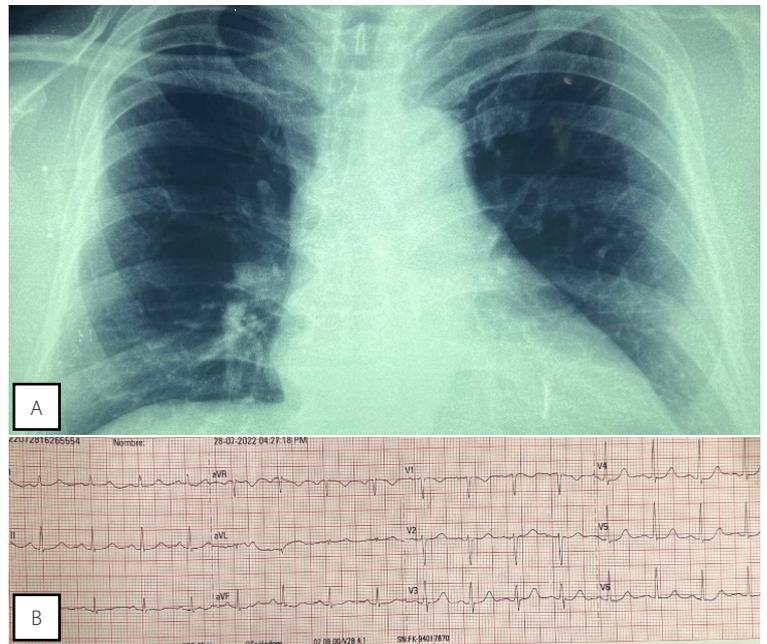
presence of keratinocyte, and due to the clinical features, it was classified as seborrheic pemphigus. In addition to acute stress, diagnosed by psychology.

## Discussion

Pemphigus foliaceus, also called superficial pemphigus, is a rare disease of chronic course, characterized by lesions only on the skin<sup>3</sup> and by the presence of anti-desmoglein 1 antibody in the subcorneal area<sup>11</sup>. Pemphigus foliaceus is classified as endemic (*fogo selvagem*) and non-endemic or sporadic<sup>7</sup>. The latter includes a variety known as seborrheic pemphigus, which is identified as localized and is also known as pemphigus erythematous or Senear-Usher syndrome<sup>8</sup>.



**Figure 2.** Status of Post-hydrotherapy patient



**Figure 3.** A. Chest X-ray of patient on admission. B. Electrocardiogram from patient on admission



**Figure 4.** Biopsy site from thoracic skin



**Figure 5.** Patient status at discharge.

Its etiology is due to the presence of anti-IgG antibodies against the extracellular amino-terminal domain of desmoglein 1 of the keratinocytes of the granular layer<sup>6</sup>. It is important to note that desmoglein 1 is found throughout the epidermis, predominantly in the skin, and is almost absent in the mucosa. However, it is expressed in greater proportion in the more superficial layers<sup>12</sup>. Clinically, the primary lesion is a flaccid vesicle or blister, but due to the location of the epidermal separation, lesions tend to rupture, so intact blisters or vesicles are rarely seen on physical examination<sup>13</sup>. On the contrary, scattered, crusted, well-demarcated, erythematous lesions on the face, scalp and upper trunk are common<sup>6</sup>. Mucosal involvement is infrequent, as presented in the described case, where the initial lesion was in the anterior region of the thorax and spread to the characteristic crusted areas. Skin lesions may cause burning and pain, but the patient does not present severity<sup>3</sup>.

On histopathology, pemphigus foliaceus blisters form in the superficial layers of the epidermis, as presented in the biopsy result, while pemphigus vulgaris blisters can form at any epidermal level, but typically, in the lower layers of the epidermis<sup>12</sup>. Diagnosis is based on clinical findings with confirmation of the characteristic histopathological lesion<sup>12</sup>.

The main objective of treatment is the control and healing of cutaneous blistering lesions. Pharmacological management aims to cure the blistering eruption, eliminate the functional deterioration associated with the disease, prevent recurrences, reduce the common side effects associated with long-term corticosteroid treatment, and improve the quality of life of patients<sup>4,14</sup>.

Depending on the severity, Pemphigus is classified as mild, moderate, or severe to guide treatment<sup>4</sup>. Then, Mild pemphigus foliaceus will be defined as less than 5 % body surface area involvement or a pemphigus disease area index (PDAI) score  $\leq 15$ <sup>14</sup>.

For the mild variety, the first-line treatment scheme consists of the use of topical corticosteroids<sup>4</sup>; other authors mention the combination of these with the use of dapsone at starting doses between 50 and 100 mg daily, with an increase of up to 1.5 mg/kg of body weight<sup>4</sup>.

As second-line management, treatment with rituximab (two infusions of 1 g two weeks apart) alone or associated with topical corticosteroids or oral corticosteroids is mentioned. Azathioprine and mycophenolate mofetil or mycophenolate sodium have also been described<sup>4,14</sup>.

On the other hand, moderate and severe pemphigus foliaceus is that which meets the following clinical criteria: significant pain and an affected body surface area above 5 % or according to the PDAI score (between 15-45 points for moderate, and above 45 points for severe)<sup>4,14</sup>. Its different treatment schemes are based on Rituximab associated with systemic corticosteroids; systemic corticotherapy alone or associated with an immunosuppressive drug as a corticosteroid-sparing agent (azathioprine, mycophenolate mofetil or mycophenolate sodium)<sup>4,14,15</sup>.

## Ethical aspects

The preparation of this case was based on the Helsinki principles, in which the confidentiality of the patient, who authorized the publication of the clinical case and the images through informed consent, was guaranteed.

## Funding

No external funds were received for this work.

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