Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Internal Medicine

Pemphigus Vulgaris about a Case and Review of the Literature

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DOI: 10.36347/sjmcr.2023.v11i11.017 | **Received:** 10.10.2023 | **Accepted:** 06.11.2023 | **Published:** 19.11.2023

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Abstract **Case Report**

***Introduction*:** Pemphigus vulgaris (PV) is a serious autoimmune disorder of the skin, which accounts for 80% of all pemphigus cases. ***Observation*:** This article describes the case of a 27-year-old woman with multiple skin wounds who was diagnosed with PV. The article also reviews the existing literature on the diagnosis and treatment of the disease. ***Conclusion*:** PV is a serious pathology with complex diagnosis and therapy, it requires multidisciplinary care associated with therapeutic education.

Keywords: Pemphigus vulgaris, Immune disorder, Corticosteroid therapy, Internal Medicine.

INTRODUCTION

Pemphigus vulgaris (PV) is a rare autoimmune bullous dermatosis (AIBD), characterized by the production of autoantibodies directed against the interkeratinocyte junction system. It results clinically as a bubble and histologically as acantholysis. It is a chronic condition, common in adults. Several clinical forms have been described. Treatment is based on immunosuppressants; nevertheless, it is corticosteroid therapy that is widely used in our context. Difficulties in accessing specialized care delay diagnosis and adequate treatment. The prognosis is poor, with mortality of 70 to 90% at two years; especially if there are associated comorbidities. We thus describe the case of a patient suffering from PV.

OBSERVATION

We report the case of a 27-year-old patient of Malian nationality; household. She was received on July 17, 2023 in the Internal Medicine department of the Point G Hospital and University Center in Bamako for multiple skin wounds.

The start of the symptoms dates back around a year, marked by the progressive appearance of multiple oral erosions located on the hard palate, on the inner side of the two cheeks, itchy on contact, with uncharacterized limits, painful on contact and estimated at 8/10 according to the simple verbal scale (EVS); without triggering factors or sedative, non-bleeding, non-purulent and without fever. No therapeutic attitude had been undertaken. Three months later, the oral erosions spontaneously decreased and permanent itching gradually set in on the scalp, without scratching lesions and which then spread to the rest of the body and without lesions. After around three months, bubbles of different sizes and poorly characterized, painful (8/10 according to the EVS), of cloudy content, rupture spontaneously or under pressure leaving room for erosions; first appeared on the scalp and then spread to the rest of the body. Faced with this symptomatology, she consulted in a medical center in the city of Bamako, where after a clinical examination, a certain number of paraclinical assessments and unspecified treatments were administered but without success. Faced with the persistence of the aforementioned symptoms, she consulted us for treatment.

In addition to the history, she reports significant unquantified weight loss. As antecedents; she has been hypertensive for around 10 years under irregular care, she is 4th procedure, 2nd parent, 2 living children and 2 abortions (the first at 4 months of pregnancy and the second at 6 months of pregnancy of unknown etiologies); married in a monogamous regime. She reportedly took cotrimoxazole and unspecified anti-hypertensives. She consumes coffee and her diet is mainly based on cereals.

The examination of the general condition revealed a conscious, bedridden, emaciated patient; athletic and afebrile to the touch; his Karnofsky index was 50%. Blood pressure in a seated position in the left arm at 130/90 mmHg; a heart rate of 88 bpm; a respiratory rate of 18 cycles per minute; a left axillary temperature of 36.9°C; capillary blood glucose at entry at 0.87g/L. His measurements at entry were a weight of 60kg, height of 164cm for a body mass index (BMI) of 22.30kg/m².

The dermatological examination noted:

- Multiple bubbles of variable size, with cloudy contents, oval shapes, with clear borders, flaccid, not painful, scattered throughout the integument, with spaces of healthy skin; associated with post-bullous erosions, pigmentary scars in places, detached and removable skin surface estimated according to Wallace at 15% (the head 4% - the front thorax 4.5% - the right upper limb 1% - the left upper limb 1.5% - the back 3% - the left and right lower limbs 1 and 1%). Positive Nicholsky sign, onychopathy of the middle finger of the left hand, the index and ring fingers of the right hand associated with spontaneous pain on palpation (5/10 according to EVS). White, peelable, non-bleeding crusty lesions scattered all over the scalp
- At the locoregional level: no peri-lesional edema, no peri-lesional heat, no satellite lymphadenopathy.

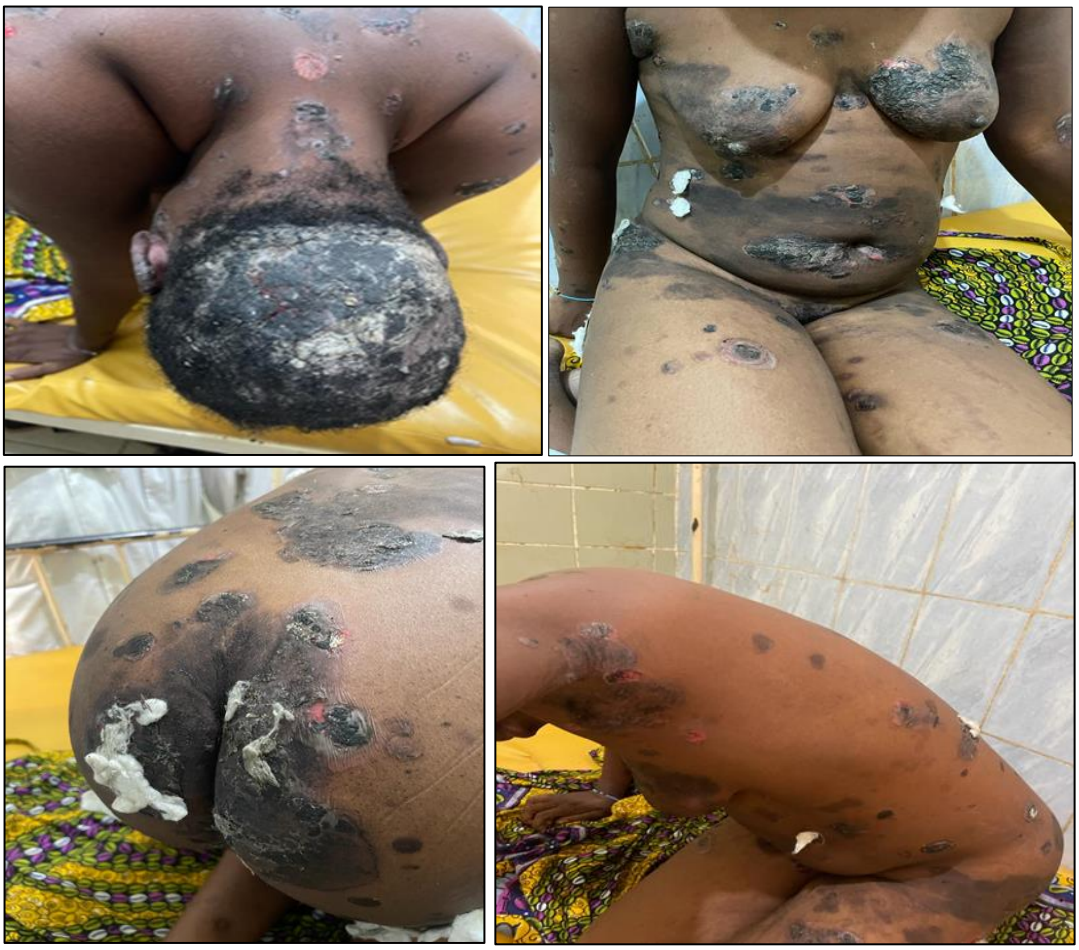
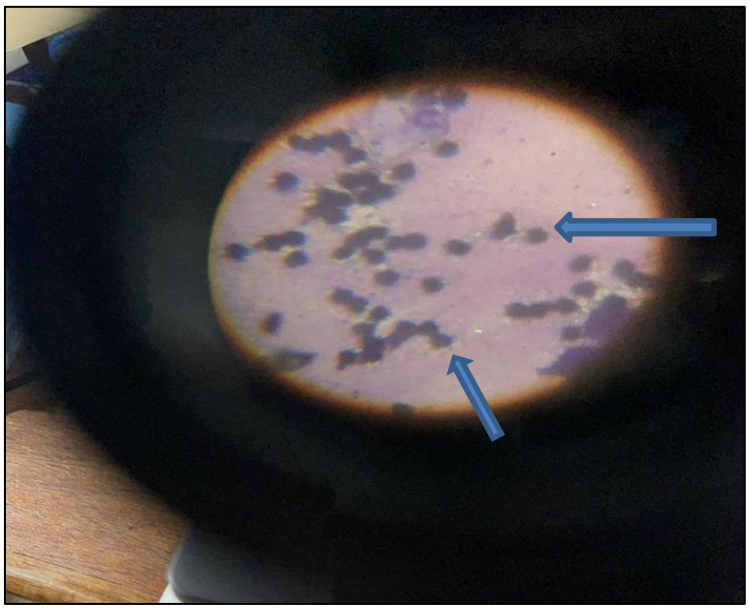


Image 1: Pictures of the patient – Day 1 (Dr Ibrahima A Dembélé – Dr Achta Ahmat a Hassane)

The rest of the physical examination revealed at the genital level, upon inspection of the vulvar, vestibular and perineal region, an erythematous vulva, without scratching lesions, without vesicles or ulcerations. Vaginal examination reveals leucorrhoea with a brown appearance, thick, brittle and fetid; without pruritus, metrorrhagia and associated pelvic pain. The other devices were carefully examined and found no anomalies.

The paraclinical assessment showed:

- Hemogram: hypochromic normocytic anemia with a hemoglobin level of 11.9 g/dl, the mean corpuscular volume (MCV) of 86.6 fl; a CCMH at 37.1 g/dl. Erythropenia at 3,710,000/mm3
- Inflammatory assessment: an inflammatory syndrome with a CRP of 19.8 mg/l
- Infectious assessment: negative HbsAg, anti-Hbc Ab not done, anti-Hbs Ac not done, HIV1-2 and HCV serologies are negative. Analysis of the vaginal sample revealed Enterobacter claocae sensitive to cefotaximen, ceftazidime, amikacin, colistin and nalidixic acid.
- Biochemical assessment: serum creatinine at 97 µmol/l with clearance at 79.92 ml/min. Azotaemia at 5.8 mmol/L. Hyperphosphatemia at 108mmol/L, hypocalcemia at 1.40 mmol/L, hypomagnesemia at 0.52 mmol/L.
- Immunological assessment: anti-nuclear antibodies (ANA-Screen) at 0.20 (negative); anti-SSA antibodies at 6.00 AU/mL (negative); anti-SSB antibodies at 3.00 AU/mL (negative), complements c3 and c4 are at 1.42 g/L and 0.33 g/L respectively. Anti-desmoglein antibodies types 1 and 3 were requested, but were not performed.
- Anatomo-cytopathological assessment: Tzanck's cytodiagnosis revealed acantholytic cells. A skin biopsy was performed, but the result was not contributory due to poor sampling technique.



Images 2: Tzanck cytodiagnosis: acantholytic cells (Dr. Ibrahima A Dembélé)

In view of all the clinical and paraclinical elements, the diagnoses of pemphigus vulgaris associated with Enterobacter claocae vaginosis were retained.

An initial treatment with white vaseline, a body application twice a day, salicylated Vaseline, an application to the scalp twice a day, Tetracycline ointment 2%, an application to erosions twice a day, Cytéal solution, a bath twice a day, Betamethasone 0.05% a body application twice a day. Paracetamol 1g every 8 hours, Cefotaxime injection 1g every 8 hours, Enoxaparin (0.2 ml) 1 injection SC/24 hours. Prednisone 1mg/kg (60mg/day for 15 days then reduced in steps of 10mg), Calcium tablet 1000mg per day, Potassium tablet 600mg per day, Albendazole tablet 400mg for 3 days and Serum salty 0.9% one liter per 24 hours. Daily supportive psychotherapy and therapeutic education for the patient and those around her.

The evolution after 3 weeks was marked by an improvement in his general condition; a resumption of mobilization. Dermatologically, a disappearance of skin bubbles (without reappearance of new bubbles) and generalized pruritus, loss of scalp crusts associated with hair regrowth; the regression of post-bullous erosions; but the persistence of hyperchromic macules of irregular shapes, of variable sizes, scattered all over the body. The vulva was non-erythematous and leukorrhea was normal on gynecological examination. The therapeutic approach was the cessation of salicylated Vaseline and Paracetamol; the addition of Diprolene 0.05% (4 tubes) mixed with Vaseline with 5% urea, one application per day on hyperchromic macules; continue with Prednisone 30 mg per day orally associated with adjuvant measures.

Image 3: Pictures of the patient – Day 21 (Dr Ibrahima A Dembélé - Dr Achta Ahmat A Hassane)

After 1 month and 12 days of treatment, the patient's progress was favorable, marked by an absence of bubbles, hyperchromic scars and some post-bullous erosions; the presence of a ruptured boil of approximately 2cm, painful, giving way to an oozing ulceration. Therapeutic use of Diprolene 0.05% (4 tubes) mixed with vaseline with 5% urea, one application per day on hyperchromic macules; tetracycline ointment 2% one application on the ulceration, and continue with prednisone 20 mg per day orally associated with adjuvant measures. The biochemical control assessments did not find any abnormalities.

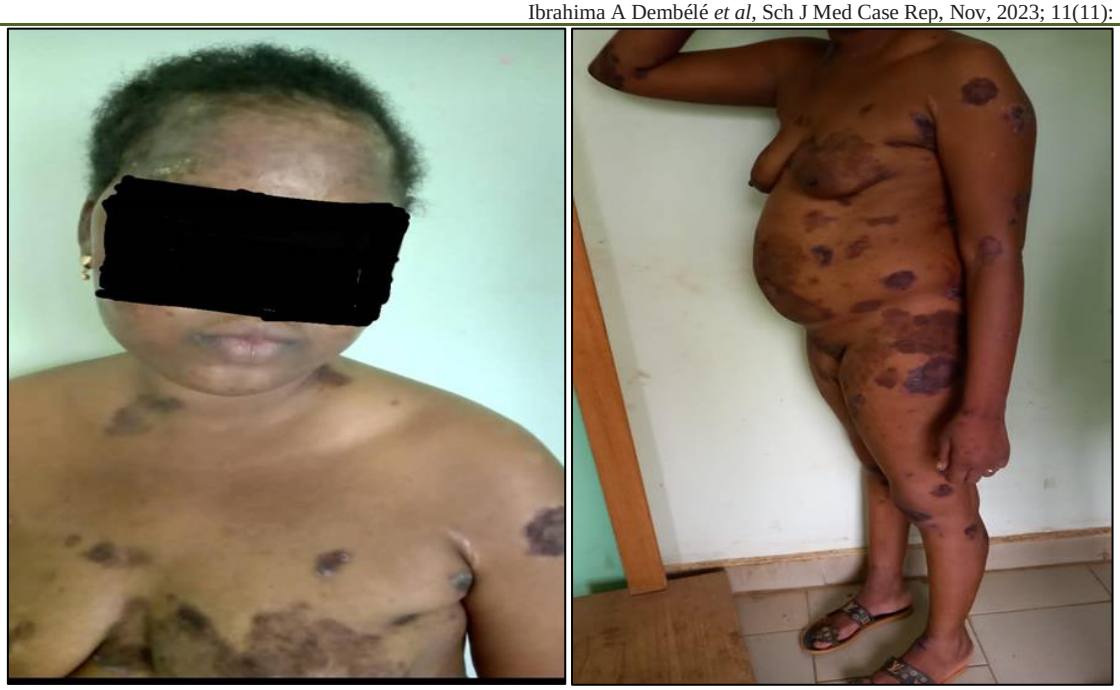


Image 4: Pictures of the patient – Day 41 (Dr Ibrahima A Dembélé - Dr Achta Ahmat A Hassane)

DISCUSSION

PV is a rare organ-specific autoimmune disease, characterized by the production of pathogenic autoantibodies directed against proteins, desmosomes and is most frequently encountered between 45 and 70 years of age without difference in sex or origin ethnic [1]. It is the most common autoimmune pemphigus [2]. Bamba found during his study a hospital frequency of 0.13% (38 cases of autoimmune pemphigus out of 26,000 patients) [8], among which pemphigus vulgaris was the most represented with 50% of cases, followed by pemphigus foliaceus with 44% of cases, pemphigus erythematosus 3% [8].

The etiopathogenesis is not fully elucidated, but humoral and cellular immune mechanisms are suggested. Autoantibodies are of the IgG class and can activate complement (classical pathway) with production of the membrane attack complex capable of altering the viability and functions of keratinocytes [4, 5]. This is valid for IgG1 class autoantibodies, but cannot apply to anti-desmoglein 1 and 3 IgG4, always found in situ on desmosomes [4, 5]. The IgG would first bind to the antigen on the surface of the keratinocytes which would activate plasminogen, releasing plasmin into the intercellular space. Plasmin allows the dissociation of the intercellular cement by cleavage of desmosomal proteins, leading to the acantholytic process [4].

The clinic is marked by an often insidious onset of erosive mucosal lesions: oral; painful, dragging and recurrent erosion that can interfere with eating (dysphagia) and lead to weight loss; genitals (Vulva, Penis) less common than oral involvement; esophageal, intravaginal, anal and ocular, sometimes [2, 3]. Janumpally *et al*., found during their observation ulcerations and labial encrustations associated with diffuse ulcerations as the main signs associated with a positive Nikolysky sign [10]. The observation of almost similar symptoms was made by De-Sousa *et al*., [7]. Skin involvement is generally secondary, several weeks or months after mucosal erosions, with bubbles: flaccid areas with clear contents, located in healthy skin; fragile, quickly giving way to post-bullous erosions surrounded by an epidermal collar, readily located in the flexion folds and the scalp, but other locations are possible; and a Nikolsky sign in peri-lesional skin, and sometimes in healthy skin [3].

Cytological examination using Tzanck cytodiagnostic analysis of the scraping product from the bottom of an erosion reveals the presence of suggestive acantholytic cells (non-specific); histological examination of a recent bleb shows a bleb with supra-basal intra-epidermal cleavage [4]. Ahankare *et al*., found the presence of acantholytic cells in their patient with PV [12], the Tzanck cytodiagnosis was carried out in our patient and gave a similar result. Direct immunofluorescence of the biopsy specimen of skin or peri-lesional mucosa shows deposits of IgG and C 3 around the keraticocytes, taking on a fishnet or mesh appearance [4, 5]. The skin biopsy performed on our patient was not contributory. Examination of serum by standard indirect immunofluorescence shows circulating anti-SIC antibodies of the IgG class whose titer is correlated with disease activity [4, 5]. Immunoblotting and ELISA determine the antigens recognized by circulating autoantibodies (desmoglein 3 plus or less desmoglein 1 in pemphigus vulgaris, desmoglein 1 in superficial pemphigus) [4, 5]. Janumpally *et al*., found autoantibodies against desmoglein 1 and 3 in indirect immunofluorescence [10], we did not perform indirect immunofluorescence in our patient.

Pemphigus vulgaris is a serious illness. Before the use of corticosteroid therapy, 70% of patients died from the disease itself, its metabolic consequences, secondary infections, or the treatments used [2]. Since corticosteroid therapy and immunosuppressants, mortality has decreased considerably, from 21% when corticosteroids were introduced to 4 - 10% when adjuvant treatments were used [7].

The goal of treatment is to achieve both remission of the rash; to avoid complications and recurrences and to cure the patient [3]. Therapeutic education of the patient and his family must be essential because it is a chronic pathology evolving in outbreaks; as well as the application of hygienic-dietary measures strict salt-free diet, sugar-free and low- potassium diet [4]. Our patient and her family benefited during her hospitalization and during the various check-ups from therapeutic education on pemphigus, its complications and the goal of therapy; as well as supportive psychotherapy. Systemic glucocorticoids often represent the first line of treatment, combined with immunosuppressants and adjuvant measures. Cyclophosphamide, mycophenolate mofetil, azathioprine, rituximab and cyclosporine have sometimes shown beneficial effects, as well as intravenous immunoglobulin (IVIG), plasmapheresis and more recently alemtuzumab, daclizumab and ibrutinib [5]. Topical steroids can also be used to speed healing of persistent skin lesions [3]. De Sousa *et al*., used in their observation high initial doses of corticosteroids (Prednisolone, 90 mg per day) combined with an immunosuppressant (Azathioprine 150 mg/day), which was later replaced by (Azathioprine 100 mg/day) [7].

In our case, the treatment was mainly based on systemic and topical corticosteroid therapy associated with adjuvant measures (calcium-potassium supplementation, prevention of gastritis); Immunosuppressants were not deemed necessary for use here.

CONCLUSION

Pemphigus vulgaris is a rare, life-threatening autoimmune disease characterized by extensive intraepidermal blisters and erosions on apparently healthy skin and mucous membranes. Diagnosis is based on skin biopsy with direct and indirect immunofluorescence and ELISA (Enzyme-Linked Immunosorbent Assay) test. Treatment is multidisciplinary and is based on corticosteroids and sometimes other immunosuppressants.

Consent

Written informed consent was obtained from the patient to publish this report in accordance with patient consent policies.

Competing Interests: The authors declare no conflict of interest.

Authors' Contributions

All authors participated in the evaluation and follow-up of the patient, in the writing and correction of the case report. All the authors of the manuscript have read and agreed to its content

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