Pemphigus vulgaris: a rare auto-immune skin disease

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European Journal of Medical Case Reports
Volume 4(12):449-452
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https://www.discoverpublish.com/
https://doi.org/10.24911/ejmcr/173-1601973044

ABSTRACT

Background: Pemphigus vulgaris (PV) is a rare autoimmune chronic blistering skin disease of type II hypersensitivity reaction.

Case Presentation: The authors present a case of a 42-year-old male with the chief complaint of crusted plaques on face, chest, and scalp for 3 months associated with itching. On investigation, direct immunofluorescence showed immunoglobulin G antibodies. The patient was prescribed antibiotics, anti-inflammatory agents etc., but they did not show any effect. So, the physicians opted for pulse therapy.

Conclusion: PV is a rare chronic ulceration of the mucosa where crusted plaques and blisters are seen. This condition can be effectively resolved using glucocorticoids and immunosuppressant. If untreated, this may lead to the death of the patient.

Keywords: Case report, autoimmune, blistering, hypersensitivity, steroids, acanthosis.

Received: 7 October 2020 **Accepted:** 15 November 2020

Type of Article: CASE REPORT **Specialty:** Dermatology

Funding: None.

Declaration of conflicting interests: The authors declare that there is no conflict of interests regarding the publication of this case report.

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Background

The term "pemphigus" is derived from the Greek word pemphix (blister) [1-3] and "vulgaris" is a Latin word meaning common [4]. Pemphigus refers to a group of autoantibody-mediated intraepidermal blistering disease characterized by loss of cohesion between epidermal cells (acanthosis) [5]. Pemphigus vulgaris (PV) has an incidence rate ranging from 0.5 to 3.2 per 10,000 per year [6]. The peak incidence of PV occurs between the fourth and sixth decades of life with a male-to-female ratio of 1:2 [7]. PV typically begins on mucosal surfaces and often progresses to involve the skin. This disease is characterized by fragile, flaccid blisters that rupture to produce extensive denudation of mucous membranes and skin. The mainstay of treatment is systemic glucocorticoids with immunosuppressant (pulse therapy).

The dexamethasone cyclophosphamide pulse (DCP) therapy is divided into four phases. Phase I consists of dexamethasone 100 mg in 5% dextrose as a slow intravenous (IV) infusion over 2 hours for three consecutive days along with cyclophosphamide 500 mg infusion on one of the days. This constitutes one DCP. Such DCPs are repeated every 28 days until no new lesion appears between two pulse therapies. Cyclophosphamide 50 mg/day is given orally. Phase II consists of the DCP therapy given for a fixed duration of 9 months. In Phase III, only oral cyclophosphamide 50 mg/day is given for 1 year, and in Phase IV, all the drugs are withdrawn and the patient is followed-up [8].

Case Presentation

A 42-year-old male was admitted to the dermatology ward with chief complaints of crusted plaques on face (Figures 1 and 2), chest region (Figure 3), and on scalp (Figure 4) for 3 months, associated with itching. The patient had a



***Figure 1**. Patient with rusted plaques all over body.*



Figure 2. *Crusted plaques on patient's face.*



Figure 3. *Crusted plaques on chest region.*

history of femur and joint pain for 2 weeks. The patient had a past medical history of hypertension for 8 years and was on medication with cilnidipine. The patient was on multi-bacillary leprosy multi-drug treatment for 2 months.



Figure 4. *Crusted plaques on scalp.*

Investigations

On physical examination, the evaluations of blood pressure and pulse rate were 130/80 mmHg and 88 bpm, respectively. On local examination, the face, chest, and scalp were found to have crusted plaques with several erosions. Oral mucosa showed a lacy pattern. The patient had an increased neutrophil count: 88%, erythrocyte sedimentation rate: 48 mm/hour, and pus cell: 6-8 cells/h.p.f. In skin biopsy, gross examination showed single partly skin covered with soft tissue (0.5*0.4*0.3 cm), which showed a stratified squamous epithelium keratinizing type; the epidermis showed acanthosis in the lower half of the epithelium; dermis showed mild edema and few lymphocytic collections; and immunofluorescence showed immunoglobulin G (IgG) antibodies.

Treatment

T. ATM (azithromycin) 250 mg per oral (PO)/BD; T. Hicope (hydroxyzine) 10 mg PO/H/S; Cap. Zevit (zinc, vitamin B complex, and vitamin C) 150 mg PO/OD; Gentian violet, topical OD; T. Cliaheart (cilnidipine) 5 mg PO/OD; Inj. Decadron (dexamethasone) 1 cc intramuscular/BD; and T. Cifran (ciprofloxacin) 500 mg PO/OD. These medications were prescribed for 10 days. On use of these medications, the severity of the condition had not decreased. So, the physicians opted for pulse therapy.

Prognosis

After following-up the patient for 3 months, the patient had shown good prognosis with the pulse therapy, but the

patient had an irregular heart rate during the time of the therapy, which was monitored and regulated.

Discussion

In PV, the lesions at first comprise small asymptomatic blisters, although they are very thin walled and can easily rupture, giving rise to painful and hemorrhagic erosions. In 70%-90% of the cases, the first signs of the disease appear on the oral mucosa.

PV typically begins on mucosal surfaces and often progresses to involve the skin. The blister cavities contain acantholytic epidermal cells, which appear as round homogenous cells containing hyperchromatic nuclei. Basal keratinocytes remain attached to the epidermal basement membrane. Direct immunofluorescence microscopy of lessional or intact patient skin shows deposits of IgG on the surface of keratinocytes.

In PV, auto-antibodies are produced against desmosomes (adhesion proteins), especially desmoglein 3 (Dsg 3). Another important component of desmosomes is desmoglein 1. The first target affects the subcutaneous site only. Dsg 3 is expressed in the oral mucosa and Dsg 1 is expressed in the skin [9]. The loss of adhesive function among the spinous cells due to anti-Dsg 3 antibodies results in a bullae formation immediately in the suprabasal region in PV [10].

The etiology of this case is still unknown. These groups of diseases are characterized by the production of antibodies against intercellular substances; therefore, they are classified as autoimmune diseases [11]. Other initiating factors reported included certain foods (garlic), infections, neoplasms, and some drugs, like captopril, penicillamine, and rifampicin [12].

In this case, the patient was prescribed antibiotics like azithromycin and ciprofloxacin, and anti-inflammatory agents, like dexamethasone for 10 days. Despite these medications, the patient did not show any improvement in his condition. So, the physicians suggested pulse therapy for the patient, which included a combination of an immunosuppressant (cyclophosphamide) and a corticosteroid (dexamethasone). However, this combination therapy has many side effects, like bradyarrhythmias, electrolyte imbalance, and seizures. This case informs that PV can also be treated well with pulse therapy for recovery of the patient's condition.

Conclusion

PV is a rare autoimmune cause of chronic ulceration of the mucosa. The severity and the natural history of PV are variable, but before the advent of steroids, most patients with PV die. PV can be treated with systemic steroids which have shown a reduced mortality rate. If PV is untreated, it may be fatal because of the susceptibility to infection and fluid and electrolyte disturbances. Most deaths occurred during the first few years of the disease. If the patient survives for 5 years, then the prognosis is good. Early disease is probably easier to control than widespread disease, and mortality rates may be higher if therapy is delayed.

What is new?

PV is a rare autoimmune disease, in which the cause is unknown. In this case report, the patient undergoes pulse therapy and recovers from the condition within 3 months.

List of Abbreviations

BP	Blood pressure
IgG	Immunoglobulin G
PO	Per oral
PR	Pulse rate

Consent for publication

Written informed consent was taken from the patient.

Ethical approval

Ethical approval is not required at our institution for publishing an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Male, 42 year old
2	**Final diagnosis**	Pemphigus vulgaris
3	**Symptoms**	Crusted plaques all over face, chest, and on scalp associated with itching
4	**Medications**	Glucocorticoids with immunosuppressant
5	**Clinical procedure**	N/A
6	**Specialty**	Dermatology