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(CASE REPORT)

A case report on pemphigus vulgaris with secondary infection

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Abstract

Pemphigus vulgaris is an autoimmune disorder characterized by ulceration of the skin or mucous membrane, most commonly affecting the oral mucosa which is derived from Greek word pemphix which means blisters or bubbles. We present a case of a 59-year-old male with a known history of pemphigus vulgaris, was managed with Oral steroids and Azathioprine. He experienced relapses in April 2023 and April 2024 after discontinuing allopathic treatment. Patient presented with widespread erosions and lesions on the face, neck, trunk, upper arm, axilla and oral cavity. He showed a positive Nikolsky's sign and multiple oral erosions. The case highlights the importance of early diagnosis, adherence to immunosuppressive therapy and regular monitoring to prevent relapses.

Keywords: Pemphigus Vulgaris; Autoimmune disorder; Nikolsky's sign; Immunosuppressive therapy; Oral steroids

1. Introduction

Pemphigus Vulgaris is the commonest type of pemphigus, is an autoimmune disease marked by erosion and ulceration of mucous membrane. Almost a majority 80-90 % patients with pemphigus vulgaris develop oral lesions occur in response to the autoantibodies which react with the desmoglein component of keratinocytes. These most commonly seen in the age group of 50-60 years old.[1, 2]

Etiological factors like diet, stress, viral infection, medications, UV radiation, ionising radiation, pesticides and allergens these factors can induce immune dysregulation can hence cause flare in the symptoms. Certain medications that can trigger development of pemphigus vulgaris include penicillin, cephalosporins, captopril, NSAIDs (Diclofenac).[3, 4]

The pathogenesis of Pemphigus vulgaris (PV) is IgG autoantibodies—primarily of the IgG4 subclass during active disease, target desmosomal cadherins, particularly desmoglein 3 (Dsg3) and, in later stages, desmoglein 1 (Dsg1). This leads to acantholysis, or the loss of keratinocyte adhesion, resulting in blistering of the mucocutaneous layer. Oral lesions typically appear first due to predominant Dsg3 expression in mucosa, while cutaneous involvement arises with the appearance of Dsg1 antibodies. Pathogenic mechanisms include steric hindrance, antibody-induced signalling (involving calcium influx, PKC activation, and phospholipase C), endocytosis of desmogleins, and possibly apoptosis and protease activation. Recent hypotheses, reveal "super-compensation" and desmoglein non assembly depletion theories, suggest that both antibody crosslinking and interference with desmosomal assembly contribute to disease.[3, 4]

The clinical picture of PV shows both mucosal and mucocutaneous involvement. About 50-60% of cases have oral lesions manifesting as either erosions or blisters that can break easily. Buccal and palate mucosa, lips and gingiva is the most common parts that are affected. The mucous membrane that can get affected include nasal mucosa, pharynx, larynx, oesophagus, vagina, penis [5]. These lesions can interrupt feeding and cause poor nutritional status.

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The diagnosis of PV includes clinical, laboratory and histopathological findings. The clinical findings is based on physical examination of blisters, Nikolsky sign, and proper medical history. The laboratory findings include Direct immunofluorescence (suggest Ig3 and C3 deposits, shows apple green colour), Indirect immunofluorescence, ELISA (detects IgG and anti Dsg1), complete blood count, a metabolic panel, ANA profile. The histological examination includes biopsy - a recent blister or perilesional area is selected and the findings indicate epidermal cleavage.[6, 5]

Treatment goal include decreasing blister formation, promote healing by making use of minimum dose of medication to control disease progress. The steroids are the mainstay in the treatment of PV because of its anti- inflammatory and immunosuppressive action. The most commonly used oral steroids include prednisolone followed by deflazacort. Prednisolone 40-60 mg /day for mild cases and 60-100/day for severe cases. Pulse therapy is used where control with prednisolone above 1 mg/kg/day cannot be achieved. Methylpred (1 g/day), dexamethasone (300 mg /day) IV are used both for 3 consecutive days. The pulse therapy minimises the side effects and also provides faster prednisolone dose.[5, 7]

The second line agents include steroid sparing immunosuppressants which include Azathioprine, Mycophenolate Mofetil, Cyclophosphamide, Methotrexate, Dapsone, Cyclosporin, and rituximab. Azathioprine (AZA) and mycophenolate mofetil (MMF) both act by inhibiting the purine synthesis and proliferation of cells. The recommended dose of AZA is 1-3 mg/kg/day.The recommended dose of MMF is 2-3 mg/kg/day. The main side effects include leukopenia, thrombocytopenia, pancytopenia. Cyclophosphamide affects B lymphocytes and antibody production, administered as pulse therapy with IV dexamethasone, orally or intravenously at a dose of 1-3 mg/kg/day. Methotrexate has both anti-inflammatory action and also inhibits proliferation of cells, given in PV at a dose of 10-20 mg/week. Rituximab [5, 3, 7]

Intravenous human immunoglobulin is used when other therapies are failed. It has higher safety profile with fewer side effects. It is always used in combination with a corticosteroid at a dose of 0.4 g/kg/day for 5 days.[7]

2. Case report

A 59-year-old male with a known history of pemphigus vulgaris, initially diagnosed in April 2020. He is also a k/c/o hypertension and type II diabetes mellitus for the past 15 years and was managed on T.Voglibose 0.3 mg 1-0-1, Dapagliflozin and Metformin 500 mg 1-0-0, Inj.H Actrapid 10-0-10, Inj.NPH 10-0-10, T.Cilacar 10 mg 1-01. He was managed with steroid pulse therapy, 1 g methylprednisolone in the first admission then tapering doses of oral steroids (methyl pred 24 mg), and azathioprine 50 mg OD until April 2022. The patient experienced relapse in April 2023, requiring replacement of steroids (T.Methyl prednisolone 8 mg BD, then 8 mg OD), which were tapered and continued for a year. In April 2024, the second relapse occurred. However, the patient discontinued all allopathic medications 2 weeks ago and opted for homeopathic treatment, following which his skin lesions significantly worsened following admission on 30/9/24. On admission he had extensive erosive skin lesions with pus discharge and crusting. On local examination

- Oral cavity showed-erosions over the buccal mucosa, tongue and erythema over the hard palate.
- Multiple erosions with crusting were seen in the face, neck, arms, axillae, chest, back and thigh.
- Genitalia -erosions over prepuce, maceration over groin flexures.

Nikolsky sign was positive

Table 1 laboratory findings

Laboratory test	**Impression**
Nikolsky sign	Positive
Biopsy	Suprabasal acantholysis with formation of an intraepidermal blister;
Complete blood count (TOTAL COUNT)	12, 300
T.Protein	5.12

Pus culture and 2 site blood cultures were sent that showed no growth further the patient was started Antibiotics Inj. Piptaz 4.5 g Q6h for 12 days, IV Inj. dexamethasone 4 mg twice daily with other supportive measures. The patient was managed with Condy's compresses and bath, Fucidin cream for lesions on scalp, trunk and limbs. Ketoconazole cream for lesions over the prepuce and groin. Saline nasal sprays were given for the lesions in the nasal cavity, for lesions on the upper back and steroids 5 mg OD was given. The patient was started T. Levocet 5mg OD for controlling allergy symptoms. He was also managed with candid mouth paint and chlorhexidine mouthwash for the erosion in the oral cavity. A pus swab was sent from the erosion over the gluteal region which showed MSSA cones and antibiotics were escalated to T. Linezolid 600 mg BD for7 days. IV dexamethasone was changed to oral Prednisolone 50 mg OD and hence lesions showed improvement. As second line agent T. Cyclophosphamide 50 mg 1-0-0 was started which was later stopped due to altered RFT.

At the time of admission the patient also had hyponatremia (Na-128), and hypoalbuminemia (2.6), and low protein 5.12 hence this was managed with salt and high protein diet. The patient was also started on IV albumin infusion for 4 hours for 3 days. The patient was also advised to continue T. Cilacar 5 mg 1-0-1 for hypertension. The patient's Renal function worsen with Creatinine 1.4mg/dl and hence cyclophosphamide was stopped. The patient also had HbA1c of 9.8 and was managed with Inj. H Actrapid 26-20-24 and Inj. Basalog 0-0-20 along with T. Metformin 500 mg 1-0-1 and T. Voglibose 0.3 mg 1-0-1.

At discharge his vitals were stable, his lesions were improving T. Prednisolone was tapered to 10 mg 3-2-0, all the previous medications and creams were asked to be continued until the next review.

3. Discussion

Pemphigus vulgaris is an autoimmune disease characterised by the formation of blisters and erosions over the oral mucosa and mucous membrane. This mainly affects the elderly from age of 40-60 years. The pathophysiology of the disease is involves the circulation of antibodies against keratinocytes cell surface. The binding of autoantibodies to keratinocytes desmosomes were Dsg3 and Dsg1 results in formation of blisters via loss of cell to cell adhesion (acantholysis). Patient with Pemphigus vulgaris having mucocutaneous disease have both anti Dsg1 and anti Dsg3 antibodies. In oral PV, Dsg3 are expressed predominantly whereas Dsg1is expressed more in mucocutaneous involvement.[6, 1]

The clinical picture of PV often begins in the oral mucosa with superficial, flaccid blisters that rapidly rupture, leaving slow-healing, painful, moist erosions making oral intake difficult and causing pain.[10] Sometimes sialorrhea and bloody saliva can be seen. Involvement of other skin areas may occur from weeks to months later. Furthermore other parts, mucocutaneous area of the mouth, the pharynx, vocal folds, and anogenital mucosa may also be affected. Scalp involvement is observed in up to 60% of patients.[8] In this case report he had lesions over Oral cavity showing -erosions over the buccal mucosa, tongue and erythema over the hard palate on local examination. Multiple erosions with crusting were seen in the face, neck, arms, axillae, chest, back and thigh, Over the Genitalia -erosions over prepuce, maceration over groin flexures.

The diagnosis of PV is based on clinical picture, medical history, laboratory investigation and histopathological studies. The clinical picture include Nikolsky sign as positive.The laboratory test include a Direct immunofluorescence (suggest Ig3 and C3 deposits, shows apple green colour), Indirect immunofluorescence, ELISA(detects IgG and anti Dsg1), complete blood count, a metabolic panel, ANA profile. The histological examination includes biopsy - a recent blister or perilesional area is selected and the findings indicate epidermal cleavage.[8, 9] Here the patient had Nikolsky sign as positive. The patient underwent a biopsy test in 2020 which showed Suprabasal acantholysis with formation of an intraepidermal blister, it confirmed the diagnosis of pemphigus vulgaris. The patient underwent Complete blood count were the total count was 12300 cells/mm^3, metabolic panel showed Na-126mmol/l T.Protein was 5.12g/dl, Hypoalbuminemia -2.6g/dl were seen.

The management of PV was done with systemic corticosteroids, like Prednisolone 40-60 mg/day and deflazacort.Pulse therapy was initiated when control with prednisolone above 1 mg/kg/day cannot be achieved, reducing side effects and providing faster prednisolone dose.Methylpred (1 g/day), dexamethasone (300 mg /day) IV are used both for 3 consecutive days. The second line agents include immunosuppressants that Azathioprine, MMF, Cyclophosphamide, Methotrexate, Dapsone, Rituximab, Intravenous human immunoglobulins.[5, 7]. Here in this case report the patient was managed with IV Inj.dexamethasone 4 mg twice daily with other supportive measures. As second line agent T. Cyclophosphamide 50 mg 1-0-0 was started which was later stopped due to altered RFT. The patient was managed with Condy's compresses and bath Fucidin cream for lesions on scalp, trunk and limbs. Ketoconazole cream for lesions over the prepuce and groin. Saline nasal sprays were given for the lesions in the nasal cavity, for lesions on the upper back

steroids 5 mg once daily was given. The patient was given Levocet 5mg OD for controlling allergy symptoms. He was also managed with candid mouth paint and chlorhexidine mouthwash for the erosion in the oral cavity.A pus swab was sent from the erosion over the gluteal region which showed MSSA cones at first he was on Inj.Piptaz 4.5 g Q6h for 12 days, which were later escalated to T.Linezolid 600 mg BD for7 days. IV dexamethasone was changed to oral Prednisolone 50 mg OD, hence lesions improved and was tapered on discharge.

Abbreviations

- PV-Pemphigus vulgaris
- MSSA- Methicillin-Susceptible Staphylococcus aureus
- Dsg- desmoglein
- OD-once daily
- BD-twice daily

4. Conclusion

This case highlights the importance of early diagnosis and prompt beginning of immunosuppressive therapy in pemphigus vulgaris management. In optimizing medication therapy, minimizing side effects, and ensuring patient compliance, our clinical pharmacists' intervention is pivotal. A corticosteroid steroid-sparing regimen with drugs like azathioprine or rituximab can induce and maintain remission. Pharmacovigilance, patient counselling, and constant monitoring are essential to improve outcomes and prevent recurrence.

Compliance with ethical standards

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Disclosure of conflict of interest

No conflict of interest to be disclosed

Statement of informed consent

Informed consent was obtained from all the individual participants included in the study.

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