

A Case Report on Pemphigus Vegetans: An Unusual Case of Pemphigus

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Abstract: *Pemphigus is a chronic epidermal immunobullous disease with potentially fatal outcomes. Among the many types of pemphigus, pemphigus vegetans (Pveg) is a rare (<5%) variant. We herein report a rare case of pemphigus vegetans presenting as multiple painful fluid filled lesions, pustules and erosions which needs documentation due to its extensive involvement. By ensuring accurate diagnosis and adherence to treatment regime a good prognosis can be achieved. This case report highlights the rarity of clinical presentation and need for early diagnosis to reduce morbidity associated with the disease.*

Keywords: Pemphigus, immunobullous disease, desmogleins, acantholysis.

1. Introduction

Pemphigus is a chronic epidermal immunobullous disease with potentially fatal outcomes, caused by autoantibodies directed against desmogleins, glycoproteins expressed on the epithelial cells of skin and mucosa, resulting in acantholysis.¹ Among the many types of pemphigus, pemphigus vegetans (Pveg), is characterized by the formation of vegetative plaques in intertriginous areas and oral mucosa. In the following case we present a case of pemphigus vegetans in a 23 year old female who presented with multiple painful erosions, eroded plaques with oozing and crusting. Clinical examination, histopathological and DIF findings were compatible with pemphigus vegetans and hence the diagnosis was confirmed. As corticosteroids are the mainstay of treatment, the side effects associated with long-term immunosuppressive therapy needs to be prevented and hence a multidisciplinary approach is necessary in following the treatment.

2. Case Report

A 23 year old healthy woman, presented to the dermatology department with multiple painful fluid filled lesions,

pustules and erosions over trunk, axilla, groin and genitalia with yellowish fluid discharge for a week. The skin lesions started with itching followed by eruption of clear fluid filled blisters which ruptured quickly and turned into crusted erosions. This later transformed into thick, elevated, moist vegetating masses.

The lesions developed initially 3 weeks ago in the oral cavity as painful oral ulcers for which she was treated at a local hospital with methylprednisolone 10mg once a day which healed the lesions but she had discontinued the medication. Thereupon the lesions reappeared and exhibited progressive enlargement, spreading across the trunk and axilla with genital and oral mucosal involvement.

Mucocutaneous examination revealed well - defined, multiple plaques of variable size with vegetating moist surface over both axillae, groin and perianal area. The lesions over the perianal and vaginal regions showed evidence of fissuring and oozing. Further on examination, multiple vesicles and excoriated erythematous plaques with pustules over both axilla, groin and genitalia were present, with foul smelling discharge.



Figure 1: a) Oral ulcers and b) multiple well - defined vegetative plaques of variable size with moist surface over axillae

Nikolsky's sign was negative. Tzanck smear revealed the presence of clusters of acantholytic cells, along with neutrophils and eosinophils. DIF revealed intercellular granular immunoglobulin G (IgG) deposits and C3 in the epidermis with a fishnet - like pattern. In addition to this her blood investigations were remarkable for peripheral eosinophilia

The histopathology sections displayed skin tissue with pseudoepitheliomatous hyperplasia in the epidermis. Additionally, an intraepidermal blister was observed, containing acantholytic cells, dyskeratotic cells, and a collection of eosinophils with a few neutrophils. Furthermore, 'tomb - stone appearance' of basal keratinocytes was observed in certain areas.

The diagnosis of pemphigus vegetans - Neumann type was confirmed through skin biopsy, direct immunofluorescence (DIF), and Tzanck smear. Initially she received intravenous dexamethasone 4 mg as part of the treatment. Additionally, topical emollients were prescribed for the skin lesions, saline gargle and candid mouth paint for the oral lesions. Condy's bath and compresses were also included in the management of the skin lesions. For mouthwash, Betamethasone 0.5mg tablets were crushed, mixed with water, and used.

The parenteral steroid was subsequently changed to a twice - daily dose of oral methylprednisolone, starting at 20 mg. After two weeks, the dosage was tapered to 20 mg in the

morning and 16 mg in the evening without the appearance of new lesions. The initial pus culture and sensitivity test revealed moderate growth of *S. aureus*, for which she was prescribed oral Amoxicillin - Clavulanate 625 mg. However, the antibiotic was later switched to Cefuroxime 500 mg upon a repeat pus culture and sensitivity test indicating superinfection with *E. coli*. Over time, the patient demonstrated significant improvement, with healing of the plaques, and no new blisters developing.

Upon discharge, she received the following recommendations: a tapering dose of Methylprednisolone (20 mg in the morning, 16 mg in the evening, followed by further tapering), Triamcinolone mouth paste, Vitamin C and multivitamin tablets, crushed betamethasone 0.5mg for use as a mouthwash, saline gargle, and Clotrimazole mouth paint. Furthermore, the patient was instructed to steer clear of foods containing thiols, phenols, thiocyanates, and tannins, as these substances have the potential to induce symptom exacerbation.

During a subsequent follow - up, she exhibited Cushingoid features (moon face), a potential adverse effect of prolonged corticosteroid use. However, the lesions were healing and did not reoccur. Later on, she was started on steroid sparing agent, oral Azathioprine 25 mg once daily and was increased to 50 mg on later followup and stopped in view of leukopenia and thrombocytopenia.



Figure 2: a) Resolving oral ulcers and b) Healing vegetative plaques

3. Discussion

Pemphigus represents a group of chronic inflammatory disorders characterized by autoantibodies that target components of desmosomes, leading to the loss of intercellular adhesion between keratinocytes and causing intraepithelial blistering. Autoantibodies are produced against structural desmosomal proteins in the skin and mucous membranes, mainly desmogleins, desmocollins and plakins.²

There are two clinically recognized forms of pemphigus vegetans, the Hallopeau type, and Neumann type. The Hallopeau type has an indolent course characterized by pustules that heal as vegetative plaques, also the oral mucosa is frequently uninvolved. The Neumann type is more severe and refractory to treatment, with vegetations that develop during an eruption of vesiculobullous lesions and the oral mucosa is usually involved.³ The patient in discussion had an extensive oral mucosal involvement and vegetations

present over the body which were congruent with the Neumann type of pemphigus vegetans.

The treatment options classically include immunosuppressive agents, such as corticosteroids and corticosteroid-sparing agents such as azathioprine, mycophenolate mofetil, cyclophosphamide, methotrexate or dapsone. Newer therapies focus on blocking cell signaling events induced by pathogenic autoantibodies and/or targeting specific autoantibodies.²

Here the patient was initially treated with an immunosuppressive agent like dexamethasone and further with methylprednisolone, which resulted in resolvment of the lesions and vegetations.

The goal of treatment is to obtain a complete remission off therapy, although many patients may only achieve a partial remission off therapy, or a complete remission on minimal therapy. The natural history of untreated pemphigus vulgaris

is to progress to life - threatening disease; therefore the risk - benefit ratio favors treatment. Patients should ideally be maintained in complete remission for at least 1 year before all immunosuppressive therapy is discontinued.

Typically, corticosteroids are tapered off first, then adjunctive immunosuppressants are slowly tapered over the course of one year. However, the tapering regimen should be tailored for each patient depending on side effects and response to therapy. It is common for patients to have a small flare with each dose taper; as long as lesions heal within one week and no further lesions form, the taper can be continued.

4. Conclusion

An interprofessional approach is essential for the accurate diagnosis and effective management of all autoimmune diseases. Pemphigus vegetans, in particular, is often overlooked and frequently misdiagnosed. Therefore healthcare professionals must collaborate to identify and treat individuals with pemphigus. Given that these patients require a range of immunosuppressive medications, it is crucial to educate them about potential adverse effects and monitor them for signs of infection. Despite being the least common type of pemphigus, untreated pemphigus vegetans can be potentially life - threatening. By fostering open communication among team members, the morbidity and mortality associated with this disorder can be minimized. In a nutshell, pemphigus vulgaris requires a multidisciplinary team approach to achieve optimal patient care and outcomes, reduce medical errors and healthcare costs.

Patient Consent for Publication

Consent was provided by the patient to use any clinical image that did not reveal personal identity.

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Conflict of Interest

There are no conflicts of interest.

Abbreviations

DIF - Direct Immunofluorescence

IgG - Immunoglobulin G

S. aureus - Staphylococcus aureus

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