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CASE REPORT
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CASE OF PEMPHIGUS VULGARIS: MULTIDISCIPLINARY APPROACH

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Abstract
Pemphigus is a life-threatening mucocutaneous immunobullous disorder affecting the skin and mucous membrane which is clinically presented by vesicle formation and histologically by intraepithelial cleft and acantholysis. Pemphigus vulgaris is a common autoimmune blistering disease always affecting the oral mucosa and it can be the initial site of presentation before involving the skin or other mucous membranes. The oral manifestations of pemphigus vulgaris typically run a chronic course causing blisters, erosions and ulcers of the oral mucosa. The dermal manifestation is not a common occurrence and many times the oral lesions are the primary clinical presentation of pemphigus vulgaris.
In this case report, we present the case of a 76-year-old woman who was admitted to our hospital with extensive pemphigus vulgaris. The patient presented with a complex and severe form of the disease, characterised by widespread blisters and lesions on her back, chest, pelvis, left eye, and abdomen, leading to septic shock. The case highlights the challenges faced by patients with this condition in rural areas of India, where access to specialised healthcare services may be limited.
The patient's treatment plan was managed by a team of healthcare professionals from various disciplines, who worked together to provide comprehensive care. This case report aims to highlight the importance of aggressive treatment and multidisciplinary care in managing patients with pemphigus vulgaris in rural areas of India.

Keywords: Pemhigus, Desmoglein 3, Mycophenolate mofetil , Occular involvement, Sepsis

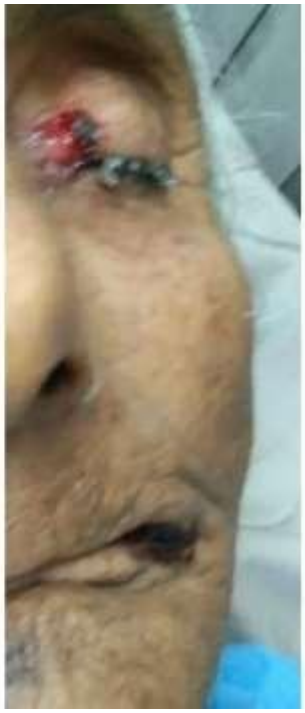
Introduction

Pemphigus vulgaris is a rare and debilitating autoimmune disorder characterised by the formation of painful blisters and lesions on the skin, mucous membranes, and other parts of the body. In India, pemphigus vulgaris is a significant public health concern however there is lack of population based studies and most studies are hospital based. The incidence of pemphigus among the dermatology outpatient attendees has varied widely, 0.09-1.8% [1].
The prevalence and incidence of pemphigus vulgaris vary significantly across different age groups and genders in India.Unfortunately, in India pemphigus occurs at a younger age as compared to western countries and tends to be more severe. A significant proportion of pemphigus patients have been less than 40 years of age[1]. Mortality due to pemphigus which was as high as 90% decreased remarkably, with aggressive and widespread use of corticosteroids but such high doses of

corticosteroids are often associated with severe side effects, and were responsible for the death of nearly 10% of patients [2]. The incidence of pemphigus, however, varies as per the geographic area and ethnic population. Literature data suggest that the incidence of pemphigus vulgaris ranges from 0.76 to 16 per million population per year in Europe. In India, the prevalence of pemphigus vulgaris is lesser than the rest of the world and is in the range of 0.09% to 1.8% [1]. Pemphigus vulgaris has a prolonged clinical course with significant morbidity and mortality. If pemphigus is left untreated, for two years the mortality is 50% and at the end of 5 years, it is nearly 100%.The causes of mortality includes extensive skin involvement, sepsis, bronchopneumonia, electrolyte imbalance and secondary systemic infections.

Patient details and clinical findings

A previously healthy 76 year old female presented to the Department of Dermatology with a chief complaint of multiple large painful skin lesions with crusting over chest, back, underarms and groin with oral involvement (buccal mucosa) for past 3 months. The patient was in her usual state of health 3 months back when she started with ulcerative skin lesions in form of vesicles and bullae on the chest followed by widespread mucocutaneous blisters affecting torso ,axilla ,pelvis and oral mucosa with lesions on the left eyelid for which she tried home remedies. These lesions broke down following minimal contact leaving behind painful raw eroded areas on back along with thick crusting over the lesions on the chest. There was history of fever, reduced appetite, malaise and foul smell from the lesions. There was no history of allergy to drugs, chemicals and food substances. There was no history of similar complaints in the past or in the family. On General examination patient was conscious but not oriented to time place and person with multiple erosions over back, mammary area, inframammary folds including upper limb, oral cavity and scalp. On oral examination erosions were seen on hard palate and buccal mucosa. Nikolsky's sign was positive in perilesional areas and negative at distant sites.Considering the oral and dermal manifestations the condition was provisionally diagnosed as pemphigus. The initial assessment of the severity of the disease required the following immediate measures: o Intravenous fluid administration to correct the circulatory and electrolyte imbalance due to excess fluid loss from the skin wounds. o Intravenous broad spectrum antibiotics to correct sepsis secondary to infection of the exposed wounds. o Cleaning of the wounds with normal saline followed by Antiseptic Bactigrass Dressing . o Half hourly monitoring of vitals and urine output.



Investigations:

Routine blood investigations were carried out which came out to be normal. Perilesional incision biopsies were taken from oral mucosa and culture sample was taken from ocular lesion which were sent for histopathology and culture sensitivity respectively. Histopathology confirmed acantholysis in the epidermis with intra- epidermal blistering. Culture sensitivity came out positive for Proteus mirabiilis. Anti desmoglin 3 antibody tittre was >200 RU/ml.Both these findings confirmed a diagnostic Pemphigus vulgaris.

Following stabilisation , the patient was shifted to dermatology ward and the required consultations were made.

- Physician consult for unstable vitals and antibiotic review, following medications were started:

Inj. Linezolid 600mg I/v BD

Inj.MVI in 100ml NS I/v slowly

Inj. Piperacillin + Tazobactam 4.5gm I/v TDS

- Ophthalmologist consultation for ulcer on left eyelid ointment Polymixin and chloramphenicol l/a , moxifloxicillin eye drops TDS l/a

- Physician Consult for CECT done for mild pleural effusion seen on chest X-ray. Mild B/L pleural effusion with adjacent atelectasis seen. Bulky and heterogenous B/L lobes of thyroid with multiple small non enhancing cystic areas seen.

- ENT consult for the above mentioned CT findings Colloidal thyroiditis. No active intervention required from their side.

Treatment received in hospital:

1. Injection Metronidazole 500mg I/V BD for 7 days
2. Injection Piperacillin+ Tazobactam 4.5 gm I/V BD for 15 days
3. Injection dexamethasone 1/2cc I/M for 12 days
4. Injection Linezolid 600mg I/V BD for 7 days F/B
5. Tablet linezolid 600 mg BD for 7 days
6. Tablet Acyclovir 800mg TDS for 7 days
7. Tablet Mycophenolate Mofetil 500 mg OD for 5 days
8. Bactigrass dressing done on alternate days

Discussion:

This case report presents a 76-year-old woman who was admitted to the hospital with extensive pemphigus vulgaris, a severe and debilitating autoimmune skin disorder. The patient's condition was characterised by extensive blisters and lesions over her back, chest, pelvis, left eye, and abdomen, which led to septic shock, severe pain.There was loss of hope for recovery or survival. The patient and her family were struggling to cope with the devastating impact of the disease on her quality of life.

The case report highlighted the importance of a multi-disciplinary approach in managing patients with complex and chronic conditions like pemphigus vulgaris. The patient's treatment plan involved a team of dermatologists, medicine specialists, ENT specialists, eye specialists, and radiologists working together to provide comprehensive care. Regular monitoring and adjustments to the treatment plan were crucial in achieving the desired outcome.

The use of immunosuppressive medications was critical in reducing the severity of the disease and preventing further complications. The patient's lesions gradually improved, reducing in size and number, and she was relieved of severe pain. She was able to gradually return to her daily activities, regaining her independence and quality of life.

Conclusion:

This case report demonstrates the importance of a multi-disciplinary approach in managing patients with complex and chronic conditions like pemphigus vulgaris. The patient's remarkable recovery from a life-threatening condition is a testament to the effectiveness of collaborative care and the importance of regular monitoring and adjustments to the treatment plan.

This report highlights the devastating impact of pemphigus vulgaris on patients' quality of life and emphasises the need for early intervention and aggressive treatment. Pemphigus has a considerable effect on quality of life of patients as well as their family members. Patients experience relapses, remissions, and a mortality rate between 5-10%, posing a challenge for treatment [3].The use of immunosuppressive medications was critical in controlling the disease and preventing further complications.

The case report also underscores the importance of patient education and empowerment.Effective therapy for pemphigus lies in combining various agents to maximize efficacy and minimize side effects. Corticosteroids continue to play a central role in pemphigus therapeutics while we continue our search for safe and effective adjuvants[3]. In conclusion, this case report demonstrates the importance of a multi-disciplinary approach in managing patients with complex and chronic conditions like pemphigus vulgaris. Early intervention, aggressive treatment, patient education, and ongoing support can deliver optimal outcomes for patients

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