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Multidisciplinary Approach in Treating Pemphigus Vulgaris by an Internist: A Case Report

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Abstract

Pemphigus vulgaris is an autoimmune disorder that affects the skin and mucosal membrane. Diagnosis of pemphigus vulgaris can take up to 10 months. Early treatment and management is vital to help decrease mortality. We describe a 69-year-old female with skin lesions who presented to a hospital with no inpatient dermatology specialty. The patient was found to have pemphigus vulgaris. The shortage of dermatologists in the United States and the inability to transfer the patient to a higher level of care, lead to innovative and collaborative approaches between specialties. This case study highlights the importance of a multidisciplinary and collaborative approach in treating pemphigus vulgaris in an institution without a dermatology department. Hence, collaboration between internal medicine hospitalists, burn care specialists, and infectious disease helped successfully treat the patient with pemphigus vulgaris.

Keywords: Pemphigus Vulgaris; Internist; Burn Care; Infectious Disease; Collaborative Approach.

Introduction

Pemphigus vulgaris (PV) is a rare acantholytic autoimmune disorder that affects the skin and mucosal membrane. In the United States, the incidence is 16.1 per million per year [1]. The age of onset is between 45 and 65 with no sex predominance. The cause of PV is thought to be an interaction between the host's genetics and environmental factors, such as diet, drugs, viruses. [2]. Manifestation of PV differs greatly from person to person [3]. Some have mucosal involvement, while others have only oral involvement. Face, eyelids, and scalp can also be involved. Usually, mucosal involvement occurs first followed by a delay in weeks to months before skin involvement occurs. These blisters and erosions are painful and can cause septicemia, which is responsible for the high morbidity and mortality associated with PV [2-3]. On average, it can take 10 months to make the diagnosis of PV [4]. Therefore, early intervention is vital to decrease mortality. We present a 69-year-old female with no past medical history who presented with blistering lesions throughout her body. We detail her medical management in a hospital that does not have a dermatology subspecialty, highlighting the multidisciplinary approach to managing PV.

Case Synopsis

A 69-year-old female with no medical problems presents to the emergency department with skin lesions for the past three weeks. The patient describes the skin lesions as painful blisters that are intermittently itchy and painful. The patient had an episode of confusion, where she was not oriented to place. She was previously seen at a smaller community hospital, where she was advised to come to a larger hospital for higher level of care. When the skin lesions were first present, she saw her primary care physician and was treated with acyclovir and Keflex. The patient had also seen a dermatologist, who suspected bullous pemphigoid and started her on doxycycline and clobetasol cream. However, despite these

medications, her rash had not improved.

Upon admission, temperature was 37.4°C (99.3°F), blood pressure was 110/54, heart rate was 90 beats per minute, respiratory rate was 17, and oxygen saturation was 100% on room air. She had multiple scattered, crusted lesions on her face, ear, right knee, and bilateral hands without evidence of bleeding. Skin biopsy was performed. Initial laboratory values were significant for leukocytosis (11,800), thrombocytosis ($479x10^3$µL), hyponatremia (132 mmol/L), and hypocalcemia (7.6 mg/dL).

The patient was attempted to be transferred to a hospital that has inpatient dermatology for two days. During this time, the patient became febrile with a temperature of 38.4°C (101.1°F) and tachycardic at a heart rate of 109 beats per minute. Repeat labs demonstrated an increase in white blood count (WBC) to 12,500. The patient became altered and only oriented to self. A chest x-ray was performed to rule out any pulmonary cause of increase in WBC and computed tomography (CT) scan of head was performed due to altered mental status, which were unremarkable. The patient was admitted to internal medicine for sepsis and management of the blistering and crusted lesions.

The patient was empirically started on vancomycin and ceftriaxone for broad antibiotic coverage. Burn unit was consulted for the blistering skin, who started the patient on xeroform and mupirocin. As the patient did not improve properly and due to high concern for autoimmune etiology of skin lesion, the patient was started on IV methylprednisolone. Blood cultures demonstrated MSSA, sensitive to oxacillin. Therefore, vancomycin and ceftriaxone were deescalated to oxacillin.

On day 10 of hospitalization, final skin biopsy resulted in pemphigus vulgaris (Figure 1). Despite having a final diagnosis, transfer to higher level of care with both burn unit and dermatologist was declined by 6 hospitals. Case management broadened the search to include other states, applying to 15 more hospitals for possible transfer.

Infectious disease was consulted for the open skin lesions and concerns that the antibiotics used might have exacerbated the pemphigus vulgaris. Infectious disease recommended to increase the dosage for oxacillin and continue antibiotics for 14 days. Per infectious disease, antibiotics used are not known to be triggers of pemphigus vulgaris.

On day 15, the patient failed to improve with methylprednisolone for 11 days. Based on research, rituximab would be the next line of treatment. Before starting rituximab, hepatitis panel and quantiferon tests were performed. At the hospital, the machines used to run the hepatitis panel were broken, delaying treatment. Also, quantiferon results were indeterminate. Chest x-ray was performed to rule out any active tuberculosis, which was negative.

On day 16, a small amount of neon green drainage and foul smell from the skin lesions were witnessed by the burn unit. This continued on day 17 and 18. Due to the increasing amount of neon green drainage, wound culture was obtained, demonstrating pseudomonas. Cefepime was added to oxacillin.

On day 18, hepatitis results were negative and quantiferon results were indeterminate. Telemedicine infectious disease was consulted, recommending rituximab and tuberculosis treatment. The patient was at first hesitant and wanted to hold treatment until in house infectious disease was consulted. Repeat quantiferon was performed.

On day 21, the patient agreed to start rituximab although repeat quantiferon was still pending. The patient was made aware of risks and benefits of starting rituximab and possibility of activation of latent tuberculosis while on rituximab. The patient agreed to continue treatment with rituximab and was started on rituximab 1000 mg infusion and was closely monitored for any adverse reactions.

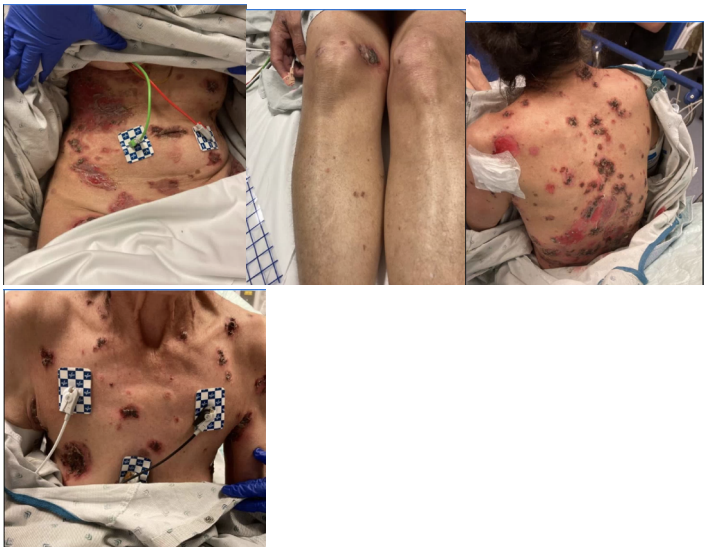
Tapering dose of methylprednisolone was initiated. Repeat quantiferon resulted in indeterminate again. Repeat chest x-ray was not performed at this time. Infectious disease was consulted and recommended purified protein derivative test (PPD test), which showed no induration. After the patient's first dose of rituximab, her skin was healing slowly. Also, the patient was able to participate in physical therapy, was able to move without difficulty, and have more of an appetite. Furthermore, patient mentation improved.

On day 33, the patient started to become delirious with confusion about place and time. Urinary tract infection (UTI) was suspected as the patient has recurrent UTI and patient endorsed suprapubic pain and dysuria. The patient was empirically treated with ceftriaxone. Urine cultures were sent, which later was positive for candida glabrata. Ceftriaxone was switched to fluconazole. However, on day 37, the patient started having red-tinged blood in urine. Infectious disease was consulted and recommended to increase the dose of fluconazole. Infectious disease also recommended to start PJP prophylaxis with Bactrim due to extensive steroid use.

On day 35, the patient received another treatment with rituximab 1000 mg. The patient tolerated the infusion well without any complications. The patient continued to report noticeable differences in her skin with improvements in overall pain.

On day 37, the patient developed hematuria. Infectious disease recommended an increase in the dose of fluconazole and to start pneumocystis jirovercii pneumonia prophylaxis with sulfamethoxazole and trimethoprim due to extensive steroid use. The patient thereafter received her second dose of rituximab 1000 mg with further improvement in her lesions.

Table 1 demonstrates the treatments and pictures of the patient's lesions. After the second dose of rituximab, the patient had considerable improvements of her lesions and pain.

Hospital day	Treatment	Clinical Presentation
Day 1	No treatment initiated	
Day 13	Patient received the following medications: ● Methylprednisolone 125 mg ● Oxacillin 1g ● Xeroform and Bactroban dressing changes	
Day 18	● Methylprednisolone 80 mg ● Oxacillin 2g ● Cefepime	

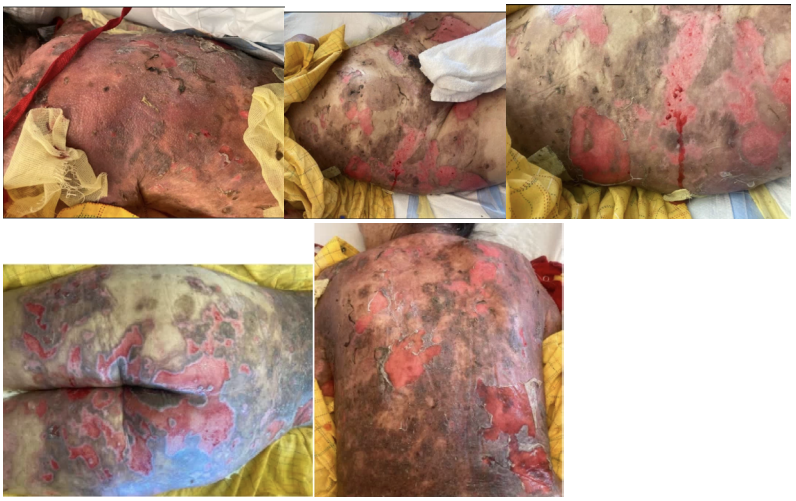
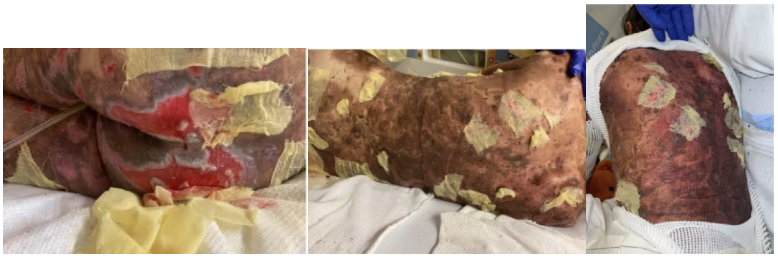
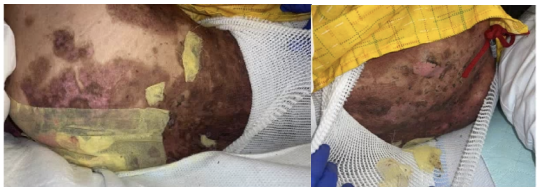
Day 29	• Rituximab 1 g (1st dose) • Oxacillin 2g • Cefepime	
36	Rituximab 1 g (2nd dose)	
38		

Table 1: Patient's clinical course throughout hospital stay from days 1-38, highlighting treatment provided and overall clinical improvement in her condition.

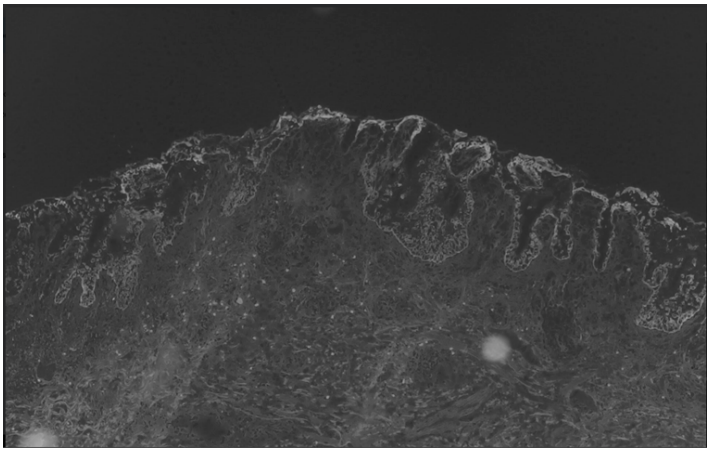


Figure 1: Direct immunofluorescence of pemphigus vulgaris. Image obtained from University of California Irvine Health, Department of Dermatology.

Discussion

This study emphasizes the value of treating pemphigus vulgaris (PV) in a multidisciplinary and cooperative manner, particularly in hospital settings where specialized dermatological services are absent. Pemphigus vulgaris was treated in collaboration by the departments of internal medicine, burn care, and infectious disease. The case also emphasizes the critical importance of cutting-edge therapies, including Rituximab, in successfully controlling pemphigus vulgaris.

In handling PV, the case study underscores the crucial role of a multidisciplinary approach, particularly in hospital settings where specialized dermatological services are absent. Given the current dermatological scarcity in the United States, this topic is very important. Only 500 residency seats are available per year for aspiring dermatologists [5], which is insufficient to fulfil the rising need for skin care specialists. There are about 11,000 dermatologists in the United States, with about 420 of them located in California [6]. A fraction of the dermatologist practice in Southern California.

Moreover, studies have shown that inpatient dermatologist consults have changed diagnosis, decreased hospital stay, and reduced readmission rate, improving the best care possible to patients [7]. The amount of inpatient dermatology physicians has decreased throughout since the 1990s and is expected to continue to decrease [8]. Currently it is unknown how many inpatient dermatologists practice in the United States. In 2024, a journal paper was published to help establish the inpatient dermatologist workforce between 2013-2019. This study looked at Medicare and dermatologist who billed 11 or more services in a year. There were 782 inpatient dermatologists between 2013-2019. The lack of inpatient dermatologists will hinder patient care.

This deficit is more than just a logistical issue; it has important effects on how patients are treated. The 69-year-old patient's story highlights the risks of improper treatment and delayed diagnosis, which resulted in sepsis and growth of pseudomonas bacteria. During the days she waited to be transferred to a hospital with both inpatient dermatological and burn services, her health deteriorated, highlighting the urgent need for quick medical attention.

Our medical team treated the patient with Rituximab at a dosage of 1g, for 2 doses that were given two weeks apart in the absence of timely dermatological expertise. Clinical trials showing the efficacy of this dose schedule in bringing PV patients into remission provide support for it. Rituximab has been discovered to be an effective and safe treatment for PV, leading to considerable clinical improvement and remission in many cases, according to a study published in the Journal of the American Academy of Dermatology [9]. Rituximab has been shown to significantly improve or completely cure the majority of patients with refractory PV, according to research published in the British Journal of Dermatology [10].

Following the start of Rituximab, the patient in our case study showed appreciable clinical improvement, as seen by the photos capturing the healing process above. Her body was initially covered in gruesome erosions and blisters that worsened over time without adequate treatment, but they were starting to heal with treatment, and the chance of problems like secondary infections was greatly reduced.

Rituximab was used, along with the combined efforts of several medical teams, to help this patient's PV be successfully managed. This instance provides a testament to the effectiveness of Rituximab and the significance of a multidisciplinary approach in addressing difficult disorders like PV, particularly in situations where professional dermatological treatment is not easily accessible. Additionally, it draws attention to the pressing need to solve the dermatology deficit, particularly in areas where their knowledge is scarce but essential for the best possible patient outcomes.

Lastly, the bacterial organisms frequently linked to these illnesses are essential for successful management of skin infections, a common consequence of PV. Staphylococcus aureus is the main pathogen in charge of skin infections in PV patients [11]. This bacterium is infamous for its antibiotic resistance and can cause anything from serious illnesses like sepsis to skin infections. Pseudomonas, the second most frequent bacteria, is another opportunistic pathogen that can cause serious infections, especially in those with impaired immune systems [12]. The clinical course of PV can be severely complicated by these bacterial infections, which can contribute to the high rates of morbidity and mortality linked to the condition. Hence, it is necessary to be aware of these organisms for effective treatment of the patient.

Conclusion

This paper highlights the multidisciplinary and collaborative approach in treating pemphigus vulgaris, especially when an inpatient dermatology specialist is unavailable. Internal medicine, burn care team, and infectious disease worked collaboratively to treat pemphigus vulgaris.

Declarations

Contribution: All authors contributed to the manuscript write-up, literature review, and editing of the manuscripts. All authors have reviewed and agreed on the final manuscript.

Competing interests: None

Patient consent for publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The patient was also appropriately de-identified for this manuscript. Informed consent form available upon request.

Ethics approval and consent to participate: Ethical approval for this publication by IRB at Arrowhead Regional Medical Center.

Availability of date and materials: All data in our report was obtained from patient's hospitalization. Any inquiries regarding supporting data availability of this study should be directed to corresponding author.

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Potential conflicts of interest: The authors declare no conflict of interest.

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