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Systemic Pemphigus Vulgaris: A Case Report

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Purpose: Pemphigus vulgaris, a rare autoimmune blistering disease of the skin and mucous membranes remains a challenging disease to treat. Management is focused on immunotherapy against autoimmune antibodies that target keratinocyte cell adhesion molecules, and antibiotics preventing secondary infections. There is no established dressing protocol and skin is usually manipulated the least amount possible in order to minimize irritation. The authors suggest that early initiation of aggressive bathing and debridement of skin lesions, with nutritional support, is essential in accelerating resolution.

Methods: A 40 year-old male previously diagnosed with pemphigus vulgaris was admitted due to exacerbation of mucocutaneous lesions involving the epidermis and mucosa of the whole body. Steroids, immunosuppressants, intravenous immunoglobulin and antibiotics were administrated, but infection and de-epithelialization progressed, while his general condition deteriorated with a weight loss of over 20 kilograms. The plastic surgery department intervened with daily bathing, debridement of unhealthy debris and non-traumatizing coverage of growing epithelium. Total parenteral nutrition and mobilization with rehabilitation therapy was initiated as early as possible.

Results: After bathing, healthy epithelium gradually covered the patient's entire body, while his general condition improved with a corresponding weight gain of 14 kgs.

Conclusion: Treatment of pemphigus vulgaris focuses on immunotherapy and infection control. However, an equal amount of attention should be laid on early intervention with daily dressings including bathing and irrigation, nutritional support, and exercise as this accelerates resolution of existing infections, promotes healthy epithelialization and leads to faster recovery.

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I. INTRODUCTION

Pemphigus vulgaris is a rare autoimmune disease characterized by the appearance of multiple bullae in the mucocutaneous surfaces of the body. Incidence is reported to be 0.5~3.2 in 100,000 of the general population, and mortality is cited to be 6 to 30%.[1,2] Pathophysiology is based on antibodies against desmoglein (Dsg) 1 and 3 in the desmosomes connecting keratinocytes in the epithelium, which disrupt adhesion between the cells (Fig. 1).[3] Consequentially, treatment focuses on high-dose corticosteroids, immunosuppressive agents or intravenous immunoglobulin targeted against these autoantibodies, although controversy remains on the treatment of choice.[2] Skin lesions are treated case by case, usually symptomatically due to the absence of a manage-

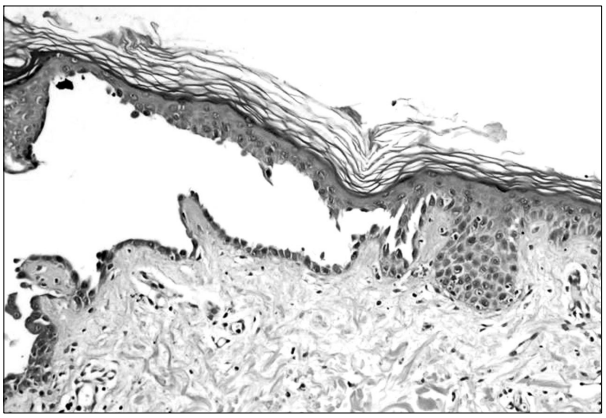

Fig. 1. Histological views of biopsy specimens (× 200, H & E stain) show detachment of the suprabasal layer of the epidermis and acantholysis.

ment protocol and various materials are utilized, minimizing trauma to the affected areas and encouraging epithelialization. Studies in the literature, most from the dermatology department, have reported use of different materials including Biobrane® or silver containing foam.[4-6] The authors have experienced successful treatment in this case of pemphigus vulgaris, using a systemic approach that included equal focus on early initiation of bathing and debridement of skin lesions, nutritional support, and conventional anti-inflammatory treatment.

II. CASE

A 40 year-old male who had a 2 year history of repetitive eruption and resolution of mucocutaneous lesions involving the epidermis and mucosa of the whole body presented with aggravation of the lesions. Exfoliation of epithelium and bullae began in the oral cavity and spread to the trunk and limbs. He had previously been admitted at four other institutions, receiving treatment with intravenous and oral steroids. Diagnosis of pemphigus vulgaris was confirmed through histopathological studies.

Physical examination revealed multiple bullae involving the trunk, all limbs, neck, face, oral cavity and conjunctivae (Fig. 2). Yellowish, puslike discharge emitting foul odor was found in skin folds of the popliteal fossa and axilla. Pinpoint bleeding was noted at the slightest stimulus to the oral mucosa, and oral feeding was impossible due to pain. The patient's body weight had dropped 20 kgs from 70 kgs to 50 kgs in 4 weeks. Pain from any type of movement stimulating the raw surfaces had the patient immobilized, and had led to contracture of the major joints, especially the knees. A grade II pressure sore had developed on his coccyx as a result of weight loss and persistent supine position.

Dexamethasone and prednisolone had been initiated on an outpatient basis, and solumedrol and deflazacort was administered upon admission, the dosage being increased when response was weak and decreased when signs of infection showed up. Azathioprine followed by cyclosporine was administered up to 100 mg. IV immunoglobulins were administered under stringent monitoring, and plasma exchange was contemplated. Approach to the cutaneous lesions was not as aggressive, and epithelial debris or discharge was wiped away with saline soaked gauzes in order to minimize pain or 'iatrogenic' exfoliation.

By the twelfth hospital day the mucocutaneous lesions had grown larger, involving almost 90% of the total body surface area. We immediately bathed him in warm water, and removed all necrotic epithelial debris. Raw surfaces were covered with foam dressing materials, and areas in the process of epithelialization were covered with petroleum gauze. Bathing was performed daily in warm, running water and dressings were changed once or twice daily depending on the amount of discharge. Healthy newly formed epithelium was left open (Fig. 3).

In order to prevent dehydration, a cradle covered with sterilized sheets was installed like a shallow tent sur

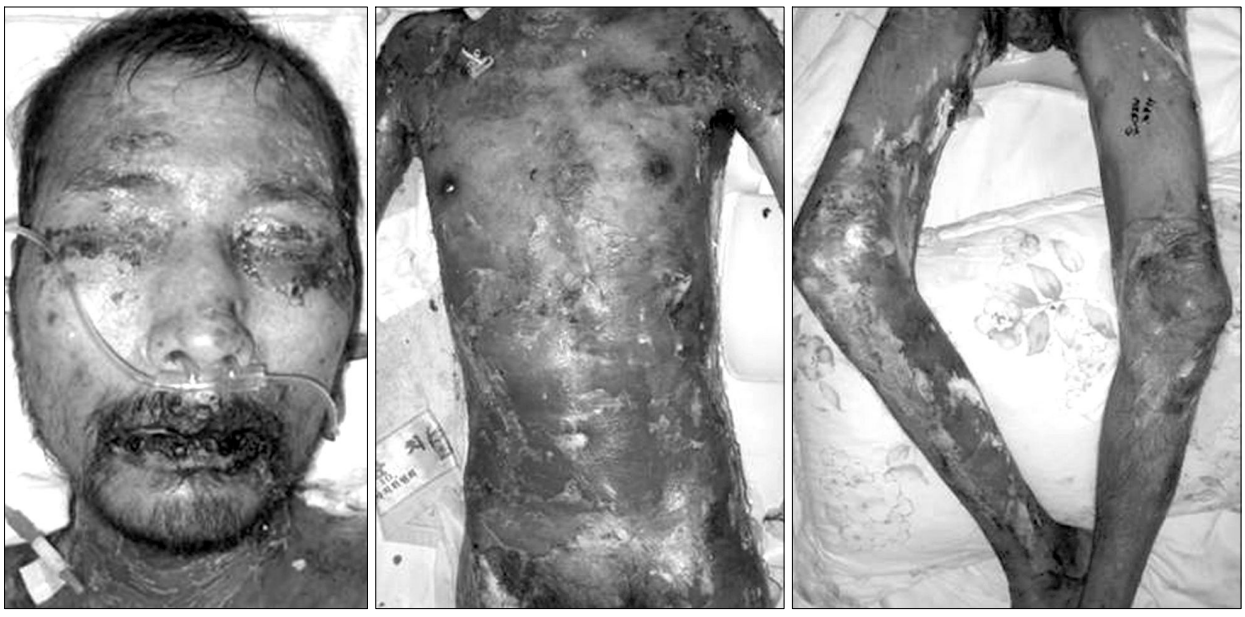

Fig. 2. The patient upon admission. Bullae formation with partial deepithelization was found on whole body skin, oral mucosa and conjunctiva. Nikolsky sign was positive, which indicates disruption of the epithelial layer.

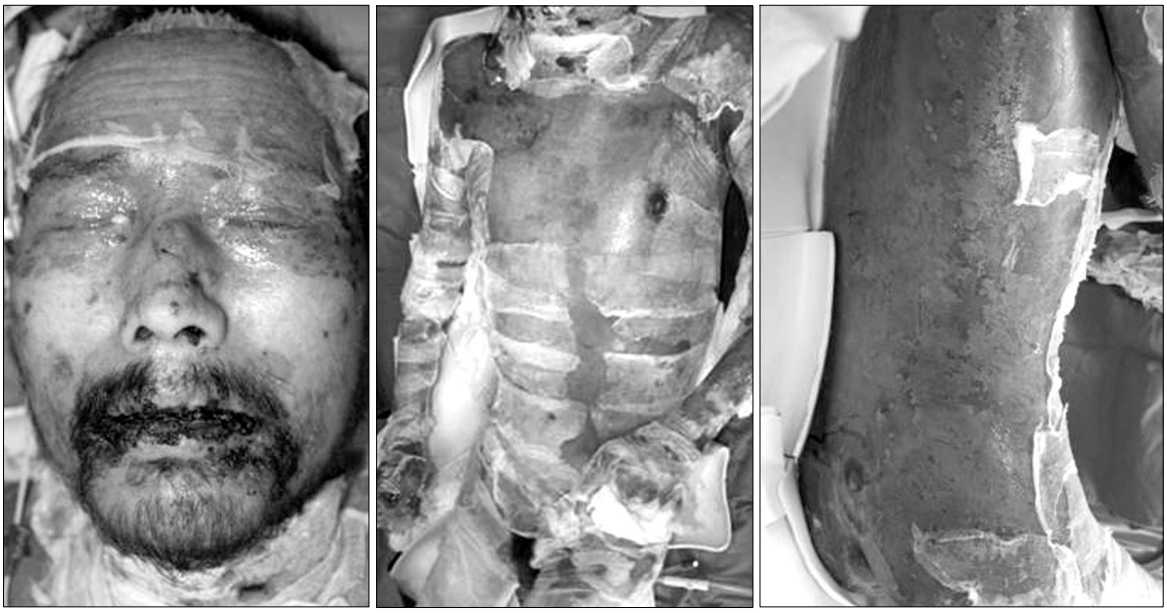

Fig. 3. The patient's 14th hospital day, 2 days after cleansing with bathing. Debridement of non-viable epithelium of trunk was perfomed and wound coverage was maintained with vaseline gauze and foam materials to prevent dehydration.

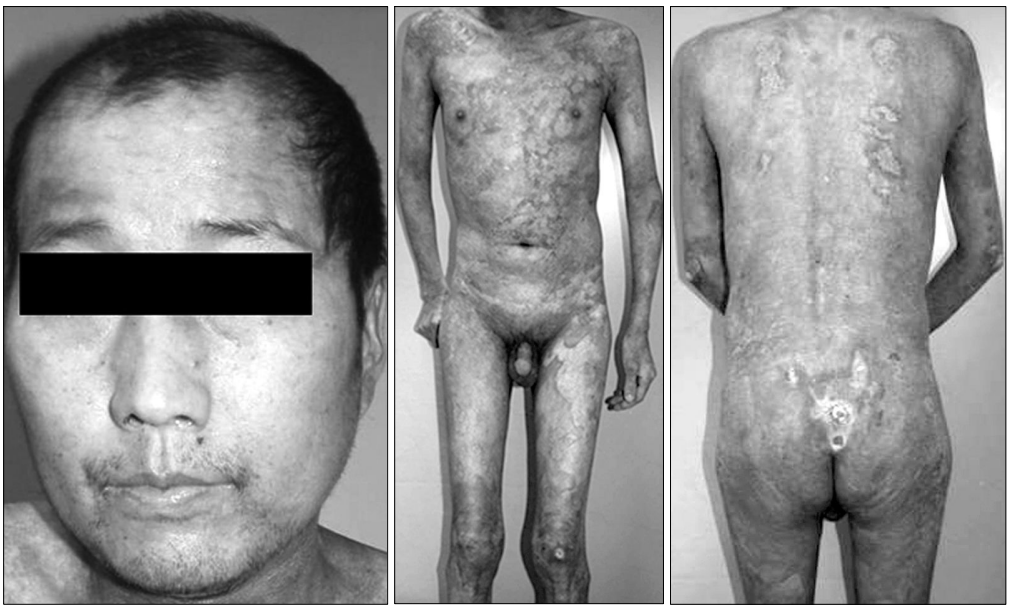

Fig. 4. 45 days of admission and 30 days after the first cleansing with bathing. Epithelization was completed on the whole body.

rounding the patient's body, the room was kept warm and a sterilized humidifier let vapor inside the tent. Fluid replacement and calorie input via total parenteral nutrition was increased, while steroids and immunosuppressants were continued with dose modification. As soon as the oral mucosa was strong enough, oral nutrition was initiated with fluid, increased step by step to soft solids. Bedside physical therapy was started under pain control and the patient began practicing light movements such as rolling side to side.

Pus discharge decreased within the first week of bathing, and serous discharge decreased slowly while epithelialization progressed. Skin on the hands and feet recovered first, then the trunk and limbs. Pain decreased as raw surface area decreased, and combined with the increase in muscle power and hyperalimentation, range of motion in all joints increased.

By the sixth week of bathing, healthy epithelium

covered the patient's entire body (Fig. 4) except for the 3 × 3 cm sized grade II coccygeal sore. Body weight was recovered at 64 kgs, and normal daily activities could be performed. The patient was discharged from the hospital with a maintenance dosage of prednisolone tablets.

III. DISCUSSION

Pemphigus vulgaris is a rare, blistering, autoimmune disease caused by antibodies against desmoglein, components of the desmosomes between keratinocytes of the epithelium. Incidence is reported to be 0.5~3.2 in 100,000, and morbidity rates range from 6~30%.[1,2]

IgG autoantibodies bind to desmoglein 1 and 3 and induce intercellular disintegration directly, or indirectly through a cascade of molecular events. The concentration of immunoglobulin is usually in proportion to disease activity. In almost all cases mucosal lesions exist, and 50~70% of those cases involve oral mucosa. Skin lesions appear afterwards covered with flaccid bullae or with a positive Nikolsky's sign or Asboe-Hansen sign. Loss of epithelium results in increased risk of infection, and may lead to sepsis.

As serious as this disease is, there is no solid treatment protocol for pemphigus vulgaris though high-dose steroids, immunosuppressants, and immunoglobulins have succeeded in lowering the mortality and morbidity rates. However, whereas our understanding of the pathogenic mechanisms and corresponding treatments of this disease has increased tremendously, there is much to be learned about the various other factors that affect the prognosis and course of therapy. There is a paucity in the literature on studies discussing management of the cutaneous lesions. This is partially due to the fact that pemphigus patients are initially managed by the dermatology or infection departments that focus on medical treatment. Stimulation of the skin is minimized to prevent further exfoliation, and consequentially prevent dehydration. There have been case reports using hydrocolloids or silver containing products, but there are no controlled studies on the effects of these materials.[4-6]

The dilemma in such patients is based on this question. How do we remove exfoliated debris without carving away at healthy but friable newly formed epithelium? Occlusive dressings may entrap necrotic debris or discharge, causing infection. Even dressing materials that absorb discharge have a limit in extent of fluids they can hold. Frequent changes may be an answer, but may financially burden the patient. Open dressing may dehydrate the patient. Wiping with gauze materials, can simultaneously remove healthy new skin.

Bathing, the process of washing the surface of the body using flowing fluids is a clear answer to this question. Flow pressure allows necrotic debris to be washed away while discharge is irrigated from the skin surfaces. As no direct friction is applied, healthy tissue is protected from being rubbed off, and pain from such stimulus is minimized. Fluid reaches surfaces and creases of the body that may be missed during individual wipings. Bathing fluid can be modified, normal saline or antibiotic mixed fluids, lotion baths have been reported.[6]

Pemphigus vulgaris is a systemic disease whose course is altered by multiple factors like nutrition, fluid therapy, physical therapy and dressing methods. The authors believe that early initiation of a wholistic, systemic approach that focuses equally on immunotherapy, infection management, dressings that include daily bathing, and nutritional aspects is essential for accelerated recovery.

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