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Return to Work Program for a Chef with Pemphigus Vulgaris : a Case Report

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Abstract

Background: Pemphigus vulgaris is an autoimmune, intraepithelial, blistering disease, affecting the skin and mucosae, is defined by IgG antibody deposits in the keratinocyte membranes of the epidermis, leading to acantholysis with consequent generation of blisters. This disease has recurrence characteristics triggering by various risk factors such as medications, physical agents, infectious agents, hormones, and stress. The aim of this case report is to manage a return to work program for pemphigus vulgaris patient with numerous, widespread, blisters especially on both hands.
Case presentation: Fourty-three years old woman presents with a painful and itchy blisters appeared in the palms, feet and body surfaces which are widely spreading throughout this week. The patient reported that the lesions first appeared as an ulcer in the upper lip, followed by cutaneus involvement. On physical examination, vital signs are normal and nutritional status is overweight. Dermatology status found numerous, widespread, tense or flaccide blisters, erosions and pruritic erythema on chest, back, abdomen, both arms and both palms accompanied by black crust with a soliter erosions on the upper lip. She was diagnosed with Pemphigus Vulgaris since 10 December 2020 and was controlled very well since February 2021, when suddenly new blisters started to appear on May 2022.
She has got a medical and occupational management for her disease, which let her recovered and ready to go back to work again.
Conclusion: Pemphigus Vulgaris could affect patient's work life. Multiple hazard in the work place also can affect the nature of the disease. Occupational management is important to help patients return to their former work, and to prevent reccurence. As a return to work program, the author recommended several changes in the workplace to reduce the triggering factor, promote self care, did an education, and showed the way to work with a good ergonomic style. Work related stress which act as an exogenous factor triggering Pemphigus Vulgaris need to be controlled. With appropriate medications and interventions to control her occupational risk and exposure, patients can return to work with some modifications.
Keyword: pemphigus vulgaris, work stress, return to work, occupational management

Abstrak

***Latar belakang** : Pemphigus vulgaris adalah suatu penyakit autoimun, intraepitel, yang menyerang kulit dan mukosa, melepuh, dan ditandai dengan endapan antibodi IgG di membran keratinosit epidermis, menyebabkan akantolisis yang mengakibatkan timbulnya lepuh. Penyakit ini biasanya kambuhan dan dipicu oleh berbagai faktor risiko seperti obat-obatan, agen fisik, agen infeksi, hormon, dan stres. Tujuan dari laporan kasus ini adalah untuk mengatur program kembali bekerja bagi pasien pemphigus vulgaris dengan lepuh yang banyak dan meluas, terutama pada kedua tangan.*
***Presentasi kasus** : Seorang wanita berusia empat puluh tiga tahun datang dengan keluhan muncul lepuh yang nyeri dan gatal di telapak tangan, kaki, dan permukaan tubuh yang menyebar luas sepanjang minggu ini. Pasien melaporkan bahwa lesi pertama kali muncul sebagai ulkus di bibir atas, diikuti dengan keterlibatan kulit. Pada pemeriksaan fisik tanda vital normal dan status gizi overweight. Status dermatologis ditemukan lepuh yang banyak, luas, dapat tegang atau lembek, erosi dan eritema pruritus pada dada, punggung, perut, kedua lengan dan kedua telapak tangan disertai krusta hitam dengan erosi soliter pada bibir atas. Ia didiagnosis menderita Pemphigus Vulgaris sejak 10 Desember 2020 dan terkontrol dengan baik sejak Februari 2021, ketika tiba-tiba mulai muncul lepuh baru pada Mei 2022. Ia telah mendapatkan penanganan medis dan okupasi untuk penyakitnya, yang memungkinkan dia pulih dan siap untuk kembali bekerja.*
***Kesimpulan** : Pemphigus Vulgaris dapat mempengaruhi kehidupan kerja seseorang. Berbagai bahaya di tempat kerja juga dapat mempengaruhi perjalanan penyakit. Manajemen okupasi penting untuk membantu pasien kembali ke pekerjaan sebelumnya, dan untuk mencegah rekurensi. Sebagai program kembali bekerja, penulis merekomendasikan beberapa perubahan di tempat kerja untuk mengurangi faktor pemicu, mempromosikan perawatan diri, memberi penyuluhan, dan menunjukkan cara bekerja dengan ergonomi yang baik. Stres akibat kerja yang merupakan faktor eksogen pemicu Pemphigus Vulgaris perlu dikendalikan. Dengan pengobatan dan intervensi yang tepat untuk mengendalikan risiko dan paparan pekerjaan, pasien dapat kembali bekerja dengan beberapa modifikasi.*
***Kata kunci** : pemphigus vulgaris, stres kerja, kembali kerja, manajemen okupasi*

Introduction

Pemphigus vulgaris (PV) is an autoimmune, intraepithelial, blistering disease, affecting the skin and mucosae, is defined by IgG antibody deposits in the keratinocyte membranes of the epidermis, attacking desmoglein 1 (Dsg1) and 3 (Dsg3), which are responsible for cell-cell adhesion.1 Binding of autoantigens to the intercellular connections leads to disruption of cell-cell adhesion, called acantholysis with consequent generation of blisters on cutaneous and mucosal surfaces. The mean age of onset is 50 to 60 years of age, and it affects both sexes equally. It is a life-threatening autoimmune disease for which treatment is indicated with the goal of treatment is to induce complete remission with minimal treatment-related adverse effects. Cutaneous and mucosal involvement in Pemphigus Vulgaris can cause significant pain and functional impairment.2

PV's incidence varies from 0,5-3,2 cases per 100,000 populations and it is increasing by certain race.[2] This disease has recurrence characteristics, according to Shimizu et al, 61,9% of cases experienced relapse, ussually when corticosteroid was tapered off.[1] Interestingly, some evidence found that PV's relapse could be triggered and aggravated by various risk factors other than medications, those are physical agents, infectious agents, hormones, and stress.[3] The aim of this case report is to manage a return to work program for pemphigus vulgaris patient with numerous, widespread, blisters especially on both hands.

Case Presentation

A 43-year-old woman presented to the Department of Dermatology and Venereology, Cipto Mangunkusumo Hospital, for evaluation of one-month history of painful and itchy new-blisters appeared in the palms, feet and body surfaces which are widely spreading within a week. The disease was controlled very well since February 2021, when suddenly new blisters started to appear on May 2022. The patient reported that the lesions first appeared as an ulcer in the upper lip, followed by cutaneus involvement. Physical examination found numerous, widespread, tense or flaccid blisters, erosions and pruritic erythema on chest, back, abdomen, both arms and both palms accompanied by black crust (Figure 1) with a solitary erosions on the upper lip. No other mucosal involvement was found in the oral, pharyngeal, laryngeal or oesophageal membranes. She was diagnosed with Pemphigus Vulgaris since 10 December 2020 and was treated with mycophenolate 360mg twice daily, systemic methyl prednisolone 16 mg/day morning and 4 mg/day in the afternoon, combined with topical corticosteroid (bethamethasone) twice daily on the lesions. She has a family history of autoimune, and she has never got a Covid-19 infection before. Now she got a series of medications and has filled out an informed consent statement that her case will be presented.

Other medications were given as a supportive treatment, such as Calsium carbonat three times daily, omeprazole once daily before eat, cetirizine as an anti allergy, and vaseline album twice daily on the lesions. Then she made an occupational medicine consultation in order to find and control hazard in her workplace and finally to make a recommendation regarding her ability to back to work. A visitation and some interviews has been done. She is working as a chef who makes local street snacks such as, risoles, lupis cakes and rice cakes. She works in a kitchen of 9 meters square, no air conditioner, with two windows and some ventilations on the wall, the air temperature is warm especially when 2 big stove is on. She usually makes 200-400 pc of cakes every day, and could be doubled on high season. She usually starts making cakes at around 3 am until 6 am in the morning and continues it at 3pm until 6pm in the afternoon. She usually wakes up at 2 am and preprares the ingredients.

During April – May 2022, there were overload orders because of Idul Fitri celebrations therefore she got an increasing workload and works overtime. We used Stress Diagnostic Survey to measure work stress and the result showed a medium stress on qualitative workload subscale. Occupational exposure for this patient as a chef includes high air room temperature (with the variation around 25-36^{0}C), chemicals used for washing and cleaning (natrium lauryl ether sulphate and natrium alkyl benzene sulfonate), biological agent (bacteria and fungus), ergonomic exposure (prolonged sitting in a non-ergonomic chair, repetitive movement on both hands, grasping motion, bending over), long working hour and work stress.

This pandemic season has made a lot of changes that could be a stresfull situation for some people. At first she opened her own cake shop and it was quite busy,

Figure 1. Numerous blisters, erosions and pruritic erythema on the body and mucous on the first visit

but sales decreased drastically since the pandemic so her employees have been laid off and she struggled to switch to online selling with a pre-order system. She usually opens pre-orders cakes three times a week. however, the scarcity and increasing prices of raw materials, the sudden increase in orders and the sudden cancellation of orders have become a challenge for her, because she do it all alone with only a little help from her husband. Besides, she got pressure from annoying customers and some of her neighbours who said that she had a contagious skin disease.

From the dermatologist, she got a series of treatment and education how to maintain the moist of the skin. From occupational medicine, as a result of the return to work analysis, she has got a temporary unfit status and recommendation to stop her work at least 2 weeks until the pemphigus flare up reduce, her work stress and psychological stress released. It was due to her lession on the arms and palms which made her couldn't hold the kitchen utensils very well, while the demand from her job is to hold it tight when fried the cakes, cutting the vegetables, and making dough. And the risk for widen the lession because of warm temperature kitchen and work stress.

During the break time, she got an occupational medicine management. To overcome her stressor, we arrange a discussion with her husband and son, made details of existing problems and looked for solutions; For annoying customers, sudden increase in orders and sudden cancellations from customers, clear pre-ordering rules are made, with down payment, with cancellation fees, and we asked her son to help manage these things. For the scarcity and increase in the price of raw materials, we advise her to find a distributor, so she can get a lower price, lower transportation fees with guaranteed quality to avoid sudden shortages of goods that result in sudden order cancellations.

To overcome the risk factor on her job proccess, we do a Hierarchy of Control, which are elimination, engineering controls, and administrative controls. For ergonomic exposures, we have done educations for a good posture at work, and provided some easy stretching exercise to do before and during her work. For the high air room temperature, we have done an environmental measurement and it has never exceed the temperature limit which is at risk of increasing Pemphigus recurrence, but since she is working in the kitchen long enough, we suggest to always open the window, install an mosquito nets upon it and it would be better to put a fan near the stoves to speed up the hot air flow to wide open windows.

For this patient, self care behaviors which are recommended to manage her condition in correlation with her job, include the following; Avoiding activities that could cause skin to become damaged, such as too many usage of chemical for washing dishes; Keep any sore patches clean to reduce the risk of infection, this can be achieved by wearing plastic gloves when working and if needed use a cotton glove beneath (to reduce the risk of sweating and hot inside the plastic gloves), use mask

and proper cotton clothing and apron while cooking; Using a soft toothbrush and mint-free toothpaste and avoid crisp, hard, hot or spicy foods to reduce the risk of oral ulcer; Gels containing local anesthetics for application at the oral ulcer, particularly prior to eat or brush the teeth, if needed; Maintaining good oral and skin hygiene with brush teeth, taking bath and wash hand regularly; Use talcum powder to prevent the skin from sticking to the bedsheets while she has to use vaseline first to cover the lession (powder could induce dry skin and more prone to infections); Finally, frequently changing and laundering towels, sheets and body linen in hot water to prevent secondary infection.

Fourteen days after treatment, she came back to the hospital with a better condition. No new blisters and no itching complaints. All lession has became macula hyperpigmentation, no pus, and no wound. Four weeks later, we have done a re-assessment of return to work, and found that she was fit to work with some notes. These notes are due to control the risk in the workplace. The author found that after coping her stress, she got a new bright perspective of her self, more confident and has done some modifications to avoid the reccurence of PV. It is now mandatory for her to maintain the cleanliness of the work place and tools, avoiding direct contact with chemicals by using rubber glove while washing and cleaning the kitchen utensils and plastic gloves while cooking, avoiding stress and still taking medications from Dermatovenereology department.

She has managed her work stress by making rules for pre-ordering cakes, fee for sudden cancellation, and help by her son managing the pre-order system to finish the raw material regularly. Besides she has done some exercise to recognize and manage her emotions by being kind to others, be open and accept her conditions, talk to friends and get some supports, tell the village elder that this disease is not contagious so he can convince the residents not to afraid ordering cakes from her.

Discussion

Pemphigus vulgaris is an autoimmune, intraepithelial, blistering disease, affecting the skin and mucosae, is defined by IgG antibody deposits in the keratinocyte membranes of the epidermis, leading to acantholysis with consequent generation of blisters.[3] Pemphigus is derived from pemphix, the Greek word for blister, it was first described in 1788 by Stephen Dickson, who observed a patient with a blister on her tongue.[2] Its affects the skin and mucosae and the autoantibodies linked to both desmoglein 1 and 3, respectively. Desmoglein-1 is found in desmosomes in the keratinocytes near the top of the epidermis, leading to the loss of intercellular adhesion between keratinocytes and are replaced by fluid, causing intraepithelial blistering, while the desmoglein-3 prevails in the lower layers of epidermis and in the mucosae.[3]

Genetic factors are involved in the pathogenesis, with HLADR4 (DRB1*0402) and HLADRw6

Figure 2. Her palms after treatment and she can back to work with some modifications.

(DQB1*0503) allele more common in patients with pemphigus vulgaris.[4] A female predominance is reported in most epidemiological studies, most of the cases occurred in the second decade of life with a peak age between 5060 years, and very rare in juvenile patients.[4] Family history in this patient showed a family eczema-history on her father which has been going on for a long time with a high relapse.

The common implicated triggers for Pemphigus Vulgaris include medications, physical agents, infectious agents, and pesticides. Physical agents which tend to induce PV include UV radiation, burn injury, high air temperature and ionizing radiation. Numerous other exogenous factors have been reported to affect the onset and aggravated the course, including hormones, stress, some kind of vaccination, topical medications, thiurams, and the allium group of plants, including onion and garlic.[5] The odd ratio for high air temperature exposure as a risk factor for pemphigus patients is 2.05 ($p < 0.001$). It has previously been shown, that an increase of temperature from 37°C to 42°C stimulates the production of lL-1. This represents an important interaction between hyperthermia and immunoregulation, because small temperature elevations may provoke T cell proliferation in response to IL-I.[6]

This patient has undergone a work related stress because of high demand and limitation in her ability to fulfill it, many cancelled order and pressure from neighbors. Based on Perry and Brunsting research, symptoms of PV were aggravated due to stress and emotional issues of the patient.[3] Furthermore, Brenner and Bar-Nathan described two patients who developed PV under great emotional stress and four years later, Cremniter et al. pointed that, despite the genetic predisposition to pemphigus, other aspects may be important in the disease's pathogenesis.[1] Work-related stress can induce a break in homeostasis by placing demands on the body that are met by the activation of two different systems, the hypothalamic–pituitary–adrenal axis and the sympathetic adrenomedullary system.[7]

It is important to recognize the influence of psychological stress on the development and evolution of pemphigus, since the health condition of the patients can be improved through the recognition, validation and treatment of their psychological issues, associating psychological assistance to the immunosuppressive treatment.[8] A study researched the impact of stressful life events which took place up to one year before the symptoms of pemphigus first appeared, the presence of personality disorders and symptoms of anxiety or depression, it is concluded that emotional stress is a factor which affects the prognostic of the disease.[3]

Night works has several disadvantages for health and autoimmune. This patient has to start her work very early in the middle of the night at 2am. According to ILO definition, night work means all work which is performed during a period of not less than seven consecutive hours, including the interval from midnight to 5 a.m. and this night shift work will alters the body's exposure to the natural light–dark schedule and disrupts circadian rhythms. These disruptions may have an impact on the pathogenesis of many diseases, including autoimmune.[9]

Based on occupational medicine management, that focus on controlling hazard in the workplace, several manipulation including her working hour, need to be changed. Without appropriate manipulation, her recurrence rate will be increased. These must be accompanied with appropriate medical treatment, without treatment, PV has a mortality rate ranging from 60 to 90%. Moreover, multiple life-threatening complications may occur, such as sepsis, fluid and electrolyte imbalances, impaired thermoregulation, as well as cardiac and renal failure. Systemic corticosteroids and adjuvant therapies have reduced the mortality rate of PV patients to approximately 10%.[4]

Occupational analysis are important to assess working patients, especially those with chronic illness and autoimune disease which are relapsing time by time. We need to control triggering and aggravating factors which can take place either in the work place or at home. At this patient we found several triggering factors in the workplace that can be at risk to induce PV relapse. High air room temperature, onion, garlic, chemicals, and stress could have an impact. The way she uses kitchen utensils, could be risky too. In order to avoid direct contact with chemicals she has to use rubber glove to wash the kitchen utensils and plastic gloves while cooking, while in the other hand using that kind of gloves will increase the temperature of the palms, and palms will be sweating so easily. So it is also advisable to use cotton gloves first to absorb sweat on the hands, beneath the plastic or rubber gloves on the outside.

Since PV is a chronic disease and regarding its autoimmune nature, patients need to adopt self-care behaviors. Many studies have been conducted to access

the effects of educational interventions on adopting self-care behaviors.[10] A study in Razi Hospital, a dermatology referral center, assessed the impacts of an educational program based on health belief model (HBM) on adopting self-care behaviors among patients with PV. The educational program was not based just on lecturing techniques but also applied group discussion and problem solving methods in the program that could have been more effective in educating the patients.[10] After the intervention, the scores of perceived susceptibility, severity, and perceived benefits increased and the rising was greater in the intervention group compare to the control group ($p<0.001$). Perceived barrier scored decrease significantly in intervention group compare to the control group ($p<0.001$; ANCOVA).[10] This shows the positive impact of HBM-based educational program as a tertiary preventive measure on adopting self-care behaviors in patients that can help them achieve self-efficacy in controlling their disease and enhancing their treatment process.[10]

This educational program has been done to this patient, as a part of occupational medicine management. We assess the patients' motivation to adopt selfcare behavior and build new concepts that affecting health behaviors, including perceived susceptibility, perceived severity, perceived benefits, perceived barriers, cues to action, and selfefficacy. Moreover, stress management has been taken into account regarding chronic and daily sources of psychological stress caused by this disease. Concerning the recurrence of this disease, the impact of selfcare behavior and stress management have beneficial effects.

Self care behavior is one of the key concept in health promotion and prevention, refers to decisions and actions to cope with a health problem, it includes seeking information, exercising, and lifestyle changes. Self care is generally viewed as a complement to professional health care for person with chronic health conditions.[11] For this patient, it is very distressing for having a rare and serious disease, plus bad opinion from her neighborhood, it can trigger feelings of distress, bewilderment, loneliness and fear, therefore we propose a stress management program to help her coping with this. A good way to start coping is building self awareness by giving her as much information as possible about her condition, support her so she can receive the condition, build positivity and tell her how to do exercise and relaxation techniques, suggest her to join a support groups which provide informations and offer support to help them manage the stress and the unpredictable remissions and exacerbations.[11]

Supportive treatment also recommended in various cases included lessions in many places throughout the body. These treatments can be soothing and reducing the symptoms,[11] for this patient we recommend to do proper dental care due to risk of oral lession, topical treatment with potent corticosteroids or calcineurin inhibitors applied directly to the lesions, antiseptic baths, covering erosive lesions using low-adhesive wound dressings whenever she is working in the kitchen and local emollients.

Conclusion

Our patient has a flare up Pemphigus Vulgaris since May 2022, she is a chef and her lession on her arms and palms, made difficulties for her job. In the other hand, multiple hazard in the work place may affect the nature of the disease. Occupational management is important to help patients return to their former work, and to prevent reccurence. As a return to work program, the author recommended several changes in the workplace to reduce the triggering factor, did an education, promote self care and showed the way to work with a good ergonomic style. Work related stress which act as an exogenous factor triggering Pemphigus Vulgaris need to be controlled. With appropriate medications and interventions to control her occupational risk and exposure, and taking into account her job description, job demand and risk of relapse, we assess that she can return to work with some modifications. Various considerations of risk factors for recurrence, including occupational exposure, work stress, psychological stress, and other personal factors, need to be controlled when making return to work decision in pemphigus vulgaris patients.

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