Correlation between quality of life assessed by Thai-version of Skindex-29 scores and disease severity among patients with pemphigus

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ABSTRACT:

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Background: Pemphigus is an autoimmune blistering disease which has a great negative impact on the quality of life (QOL) in many aspects, such as physical, psychological and social status. Exploring associated factors might be beneficial and improve the QOL.

Objective: To examine the correlation between Pemphigus Disease Area Index (PDAI) score and Thai-version of Skindex-29 score in pemphigus and define other variable factors which affected the QOL.

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Materials and Methods: Fifty-nine pemphigus patients at the Institute of Dermatology in Thailand from July 2017 to September 2017 were enrolled. The QOL was evaluated by the Thai-version of Skindex-29. The clinical severity was assessed by PDAI scoring system. Correlation analysis was used to evaluate the correlation between Skindex-29 and PDAI scores.

Result: The disease severity scores assessed by PDAI had a low positive correlation with the Skindex-29 scores (r = 0.387, P = 0.002). Furthermore, the younger age group was significantly associated with lower QOL (odds ratio [OR], 0.923; 95% confidence interval [CI], 0.866 to 0.984; P = 0.013). Shorter duration of disease showed higher risk of poorer QOL in emotion (OR, 11.752; 95% CI, 1.093 to 126.285; P = 0.042) and functioning scores (OR, 22.131; 95% CI, 2.117 to 231.317; P = 0.010).

Conclusion: Our study demonstrated a significant negative effect on the QOL in pemphigus assessed by Thai-version of Skindex-29 score. The clinical severity, age and duration of the disease were important factors associated with poor QOL.

Key words: Quality of life, skindex-29, pemphigus

Introduction

Pemphigus is a rare, potentially life-threatening autoimmune blistering disease. The estimated incidence is about 1 to 16 new cases per 1 million people per year[1]. The pathogenesis is the autoantibodies against desmoglein 1 and desmoglein 3 which are specific epidermal antigens, resulting in loss of cell adhesion to epidermal keratinocytes. According to different target antigens, pemphigus can be separated into two major forms: pemphigus vulgaris (PV) and pemphigus foliaceus (PF). Autoantibodies against desmoglein 1 and 3 are associated with PV, which is characterized by mucosal erosions with/without cutaneous blisters. Autoantibody against only desmoglein 1 is associated with PF, which clinical manifestation is cutaneous blisters without mucosal involvement. Other subtypes include drug-induced pemphigus, IgA pemphigus and paraneoplastic pemphigus.

Due to the chronicity, pemphigus has a profound effect on both physical and psychological aspects of the patient's quality of life. The World Health Organization defined the term "quality of life" (QOL) as an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns[2]. The QOL is composed of many aspects, such as the physical, functional, social, and emotional well-being. To date, there were a few previous studies demonstrating the QOL assessment in pemphigus patients by using various tools. They had shown multiple variables

besides disease severity that might impact QOL, such as sex, age group, comorbidity, medication, etc[3,4]. Therefore, patients with mild disease severity may have poorer QOL compared with patients with severe disease due to different factors[5]. In this study we used the Thai version of the Skindex-29 questionnaire which was translated and tested by Tantanasrigul P, et al. at the Institute of Dermatology in Thailand in 2015[6]. The authors had formal permission from the original authors to use the validated Thai version of the Skindex-29[6,7]. The aim of this study is to examine the correlation between the disease severity and the QOL and define the variables that affect QOL in Thai pemphigus patients.

Methods

Study design and participants

A single-center cross-sectional study was performed from July 2017 to September 2017 at the immunology clinic and inpatient care unit of the Institute of Dermatology in Thailand. A total number of fifty-nine patients with pemphigus were enrolled in the study. The inclusion criteria were as follows: patients aged between 25 and 70 years who were diagnosed as PV or PF by clinical presentation, histopathology, direct immunofluorescence, indirect immunofluorescence and/or anti-desmoglein (ELISA); patients without other dermatoses for at least 6 months; patients with ability to read and write in Thai language. Information about the study was provided to all participants and their informed consent was obtained. Patients who had chronic and severe systemic diseases with major organ involvement, bed-ridden, cerebrovascular and coronary artery diseases were excluded from the study.

The demographic data including age, sex, education level, comorbidity, pemphigus subtype, duration of disease, type and dosage of medication were collected. A dermatologist assessed the disease severity of patients by using the Pemphigus Disease Area Index (PDAI) scoring system. The Thai version of Skindex-29 questionnaire was applied to all patients.

Disease severity assessment

For pemphigus severity assessment, many monitoring tools were developed, such as Pemphigus Disease Area Index (PDAI), Autoimmune Bullous Skin Disorder Intensity Score (ABSIS), Pemphigus Vulgaris Activity Score (PVAS), Pemphigus Area and Activity Score (PAAS) and Autoimmune Bullous Disease Quality of Life (ABQOL)[8]. Among these tools, the PDAI had higher reliability and validity compared with other tools[9,10]. Moreover, it was also more sensitive to low number of lesions and easier to use than body surface area (BSA) assessment[10,11].

PDAI score system was developed by the International Pemphigus Definition Committee in 2005[12]. Three main parts of the body, including skin, scalp and mucosa are examined. Twelve

anatomical areas of the skin, including ears, nose, face, neck, chest, abdomen, back, arms, hands, legs, feet and genitalia are assessed. Size and number of the lesions in each of the anatomical areas are recorded. For the skin, the score varies on disease extension; 0 (absent), 1 (1-3 lesions, all ≤ 6 cm; up to one lesion > 2 cm diameter), 2 (2-3 lesions, all ≤ 6 cm; at least two lesions > 2 cm diameter), 3 (> 3 lesions, all ≤ 6 cm diameter), 5 (> 3 lesions, and/or at least one lesion > 6 cm diameter) or 10 (> 3 lesions, and/or at least one lesion > 16 cm diameter or entire area). For the scalp assessment, the score depends on the presence of erosions, blisters or new erythema and affected area; 0 (absent), 1 (one quadrant), 2 (two quadrants), 3 (three quadrants), 4 (whole skull) or 10 (at least one lesion > 6 cm). For the mucous membrane, twelve areas are assessed, including eye, nose, buccal mucosa, hard palate, soft palate, upper gingiva, lower gingiva, tongue, floor of mouth, labial mucosa, posterior pharynx and anogenital area. The number of erosions and blisters are evaluated; 0 (absent), 1 (1 lesion), 2 (2-3 lesions), 5 (> 3 lesions or 2 lesions > 2 cm) or 10 (entire area). The PDAI total scores range from 0 to 263, which is composed of 250 points from the activity scores and 13 points from the damage scores. The activity scores reflect the number of erosions, blisters and new erythema while the damage scores reflect postinflammatory hyperpigmentation or erythema from resolving lesion. The higher PDAI scores represent more severe disease. The severity grading of PDAI scores are classified into three groups, mild, moderate and severe. Shimizu et al[13]. illustrated the cut-off severity scores of less than 8 as mild, 9 to 24 as moderate and more than 25 as severe.

Questionnaire

Skindex-29 is three dimensional, dermatology-specific QOL questionnaire which was developed by Chren et al. in 1996[7]. The Skindex was originally developed in 61 questions. Later, it was revised and simplified into Skindex-16 and Skindex-29. Skindex-29 had shorter length than the previous version and also improve the discriminative and evaluative capability of the instrument[14]. It was recommended as one of the instruments of choice in dermatology[15]. It had been translated and adapted into many languages such as German[16], Spanish[17], Polish[18], Turkish[19] and Thai[6], etc. There are three major domains, which are composed of seven items of symptoms, twelve items of functioning and ten items of emotion domains. Skindex-29 inquires about the frequency (never, rarely, sometimes, often, all the time) that the patient experienced. The responses are a 5-point scale and transformed to a linear scale of 100, varying from 0 (no effect) to 100 (always affected). The greater scores indicate lower QOL. The cutoff score of Skindex-29 was defined by Prinsen et al[20]. which ≥ 52 points of symptoms, ≥ 39 of emotions, ≥ 37 of functioning, and ≥ 44 of

the overall score represented severe QOL impairment.

Statistical analysis

Statistical analyses were performed by using the SPSS software (SPSS Inc., Chicago, IL, USA), version 17 for Windows. Descriptive statistics were used to analyze. Skindex-29 scores in each domain and the total which were the outcome variables, were calculated into a percentage. Demographic and clinical characteristics were used as independent variables and presented as numbers with percentages, means with standard deviations and median with interquartile range. One-way ANOVA was used to compare Skindex-29 scores in different PDAI severity, and a significant one-way ANOVA was assessed with multiple comparison tests by using Tukey's HSD test. Pearson's correlation test was used to measure the strength of a linear association between the Skindex-29 scores and PDAI scores. A value ranges from -1 to +1. A value of 0.00 to 0.30 (-0.00 to -0.30) indicates negligible correlation between the variables. A value of 0.30 to 0.50 (-0.30 to -0.50) implies low positive (negative) correlation. A value of 0.50 to 0.70 (-0.50 to -0.70) implies moderate positive (negative) correlation. A value of 0.70 to 0.90 (-0.70 to -0.90) implies high positive (negative) correlation. A value of 0.90 to 1.00 (-0.90 to -1.00) implies very high positive (negative) correlation[21]. Kruskal-Wallis and Mann-Whitney test was used to compare Skindex-29 scores in each of the variables. Univariate and multivariate logistic regression analyses were performed to investigate the impact of potential risk factors on the patient's QOL. A p-value less than 0.05 was used for all tests to indicate statistical significance.

Result

Demographic data and variables

The demographic data of all participants was shown in Table 1. A total number of 59 patients were enrolled in the study. There were 40 patients with PV and 19 patients with PF. There were 20 males and 39 females. The median age was 50. The median duration of the disease was 29 months. The disease duration in most patients was more than 24 months (57.6%). Thirty-one patients had comorbid conditions. The three most common comorbid diseases were hypertension (20.3%), dyslipidemia (13.6%) and diabetes mellitus (11.9%). For the treatment, a systemic steroid was used separately or concomitantly with other immunosuppressive drugs. The most commonly used drug was the combination of prednisolone and azathioprine (53.1%). The most common side effect was secondary bacterial infection (25.4%), followed by anemia, eczema herpeticum and leukopenia (data not shown).

Table 1 Demographic data and variables

Variables	Statistics data	
	N (%)	Median (interquartile range)
Pemphigus type		
Pemphigus vulgaris	40 (67.8%)	
Pemphigus foliaceus	19 (32.2%)	
Sex		
Male	20 (33.9%)	
Female	39 (66.1%)	
Age (year)		50 (41 - 57)
25-30	2 (3.4%)	
>30-40	11 (18.6%)	
>40-50	20 (33.9%)	
>50-60	16 (27.1%)	
>60-70	10 (16.9%)	
Education level		
Uneducated	2 (3.4%)	
Elementary school	28 (47.5%)	
Secondary school	18 (30.5%)	
Bachelor's degree	11 (18.6%)	
Duration of disease (month)		29 (12 - 72)
≤6 months	4 (6.8%)	
>6-12 months	11 (18.6%)	
>12-24 months	10 (16.9%)	
>24 months	34 (57.6%)	
Comorbidity	31 (52.5%)	
Hypertension	12 (20.3%)	
Diabetes mellitus	7 (11.9%)	
Dyslipidemia	8 (13.6%)	
Hyperthyroidism	2 (3.4%)	
Other	14 (23.7%)	
Systemic medication	49 (83.1%)	
Type of systemic medication		
Prednisolone	14 (28.6 %)	
Azathioprine	6 (12.2 %)	
Prednisolone + Azathioprine	26 (53.1%)	

Variables	Statistics data	
	N (%)	Median (interquartile range)
Prednisolone + Cyclophosphamide	2 (4.1 %)	
Prednisolone + Azathioprine + IVIG	1 (2%)	
Prednisolone dosage		
≤ 10 mg/kg/d	25 (58.1%)	
> 10-20 mg/kg/d	10 (23.3%)	
> 20-30 mg/kg/d	6 (13.9%)	
> 30-45 mg/kg/d	2 (4.7%)	

Data presented as n (%) and median (interquartile range). IVIG: intravenous immunoglobulin.

The PDAI severity and Skindex-29 scores were shown in Table 2. The PDAI severity was classified into mild, moderate and severe groups. There were 22 patients (37.3%) with mild disease, 22 patients (37.3%) with moderate disease and 15 patients (25.4%) with severe disease. The mean PDAI total scores in mild, moderate and severe severity were 6.00 ± 1.85, 15.41 ± 4.14 and 33.80 ± 7.41, respectively. The mean Skindex-29 total score was 32.20 ± 21.15. The mean of emotions scores was 37.54 ± 26.24 which showed highest points compared with symptoms and functioning scores.

Table 2 Pemphigus Disease Area Index severity and Skindex-29 scores

	N (%)	Mean ± SD. or median (interquartile range)
PDAI severity		
Mild	22 (37.3%)	6.00 ± 1.85
Moderate	22 (37.3%)	15.41 ± 4.14
Severe	15 (25.4%)	33.80 ± 7.41
PDAI score		
Skin activity score		5 (0-12)
Skin damage score		7 (4-9)
Mucosa score		0 (0-0)
Scalp activity score		1 (0-2)
Scalp damage score		0 (0-1)
Total activity score		7 (1-14)
Total damage score		7 (4-9)

	N (%)	Mean ± SD. or median (interquartile range)
Total score		13 (8-25)
Skindex-29 scores		
Emotions		37.54 ± 26.24
Symptoms		34.86 ± 17.95
Functioning		29.02 ± 24.93
Total score		32.20 ± 21.15

Data presented as n (%), mean ± SD and median (interquartile range). SD: standard deviation

Table 3 showed Skindex-29 scores in different PDAI severities. There was a significant difference of all domains and Skindex-29 total scores between mild and severe groups ($p < 0.05$). There was also significantly different in symptoms, functioning and Skindex-29 total scores between moderate and severe groups ($p < 0.05$). On the contrary, any domains of Skindex-29 scores between mild and moderate PDAI severity was not significantly different. Figure 1 showed the minimum, maximum and mean of Skindex-29 total score in different disease severity.

Table 3 Skindex-29 scores in different Pemphigus Disease Area Index severity

	PDAI severity				
Skindex-29 scores	**Total (n=59)**	**Mild (n=22)**	**Moderate (n=22)**	**Severe (n=15)**	**P-value**
Emotions	37.54 ± 26.24	27.95 ± 25.39	38.18 ± 23.57	50.67 ± 26.9	0.032[b]
Symptoms	34.86 ± 17.95	29.54 ± 16.28	32.30 ± 13.74	46.42 ± 21.34	0.011[b,c]
Functioning	29.02 ± 24.93	18.50 ± 19.68	26.85 ± 22.71	47.86 ± 25.84	0.001[b,c]
Total score	32.20 ± 21.15	21.28 ± 13.57	32.05 ± 19.13	48.44 ± 23.61	<0.001[b,c]

Skindex-29 scores presented as mean ± SD. P-value corresponds to ANOVA with post-hoc Tukey Honestly Significant Difference. (a) mild vs. moderate, (b) mild vs. severe and (c) moderate vs. severe.

The correlation between PDAI and Skindex-29 scores was presented in Table 4. There was a low positive correlation between the PDAI total scores and Skindex-29 total scores (r = 0.387, $p = 0.002$) as shown in Figure 2. In addition, the PDAI total score was also low positively correlated functioning scores (r = 0.353, $p < 0.05$). In contrast, the PDAI total score was negligible correlated with the symptoms scores (r = 0.294, $p < 0.05$). Moreover, the indirect immunofluorescence titer showed a low positive correlation with Skindex-29 scores. (r = 0.485, $p < 0.05$, data not shown)

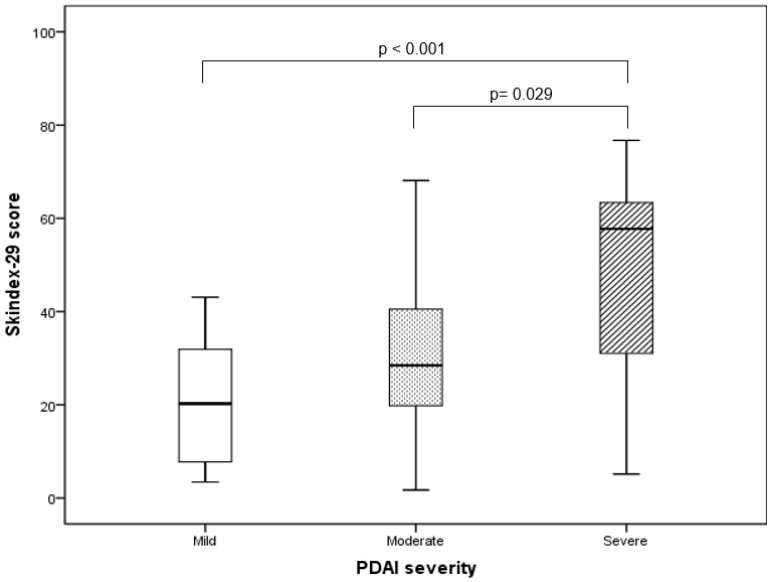

Figure 1 Skindex-29 total score in different disease severity

Table 4 The correlation between Pemphigus Disease Area Index severity and Skindex-29 scores

	PDAI severity			
Skindex-29 scores	Mild (n=22)	Moderate (n=22)	Severe (n=15)	Total score
Emotions	-0.094	-0.181	-0.405	0.222
Symptoms	-0.028	-0.004	-0.289	0.294
Functioning	0.008	-0.295	-0.278	0.353*
Total score	0.198	-0.222	-0.348	0.387*

Data presented as the correlation coefficient (r). * P-value corresponded to Pearson correlation test.

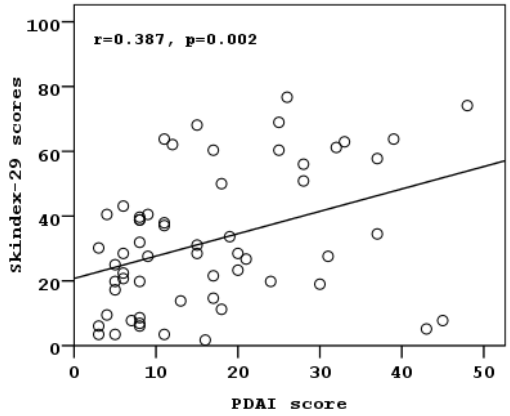

Figure 2 The correlation between PDAI scores and Skindex-29 scores

Figure 3 showed Skindex-29 score in pemphigus patients with different ages. The different age groups were significantly different in emotions, symptoms, functioning and Skindex-29 total scores ($p < 0.05$). The age group of 30-40 years had the highest Skindex-29 total scores while the age group of 60-70 years showed the lowest scores. In contrast, the other variables showed no statistically significant difference in Skindex-29 scores. Patients with PV had no statistically significant difference of Skindex-29 total score compared with PF ($p = 0.390$, data not shown). The Skindex-29 score of patients with comorbidity was not statistically significantly different compared with those without comorbidity ($p = 0.832$, data not shown). Moreover, the different types of comorbidity, sex, education level, duration of disease, type and dosage of medication did not affect the Skindex-29 scores.

Risk factors for severe QOL impairment from Skindex-29 scores, which was identified by multivariate logistic regression analysis, was shown in Table 5. The age was the significant factors associated with severe QOL impairment (odds ratio, 0.923; 95% confidence interval [CI], 0.866 to 0.984; $p = 0.013$). There was 7.7% reduction in severe QOL impairment per year of the increased age. Duration of disease was another associated factor for poor QOL. Shorter disease duration, especially less than 6 months showed higher risk of poorer QOL as shown in emotion (odds ratio, 11.752; 95% confidence interval [CI], 1.093 to 126.285; $p = 0.042$) and functioning scores (odds ratio, 22.131; 95% confidence interval [CI], 2.117 to 231.317; $p = 0.010$). Other variables did not show a risk for severe QOL impairment.

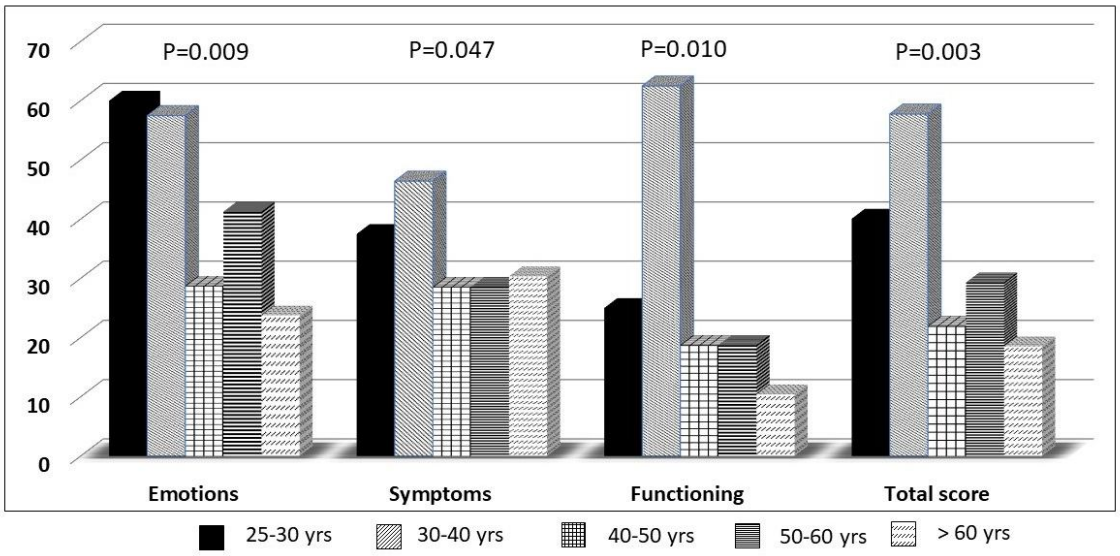

Figure 3 Skindex-29 score of different age groups in pemphigus patients

Table 5 Risk factors for quality of life impairment by multivariate logistic regression analysis

Variables	Emotions		Symptoms		Functioning		Total score	
	OR (95% CI)	p-value	OR (95% CI)	p-value	OR (95% CI)	p-value	OR (95% CI)	p-value
Constant	14.144	0.132	3.415	0.515	7.672	0.268	14.825	0.072
Age[a]	0.929 (0.868-0.993)	0.031	0.901 (0.822-0.987)	0.025	0.932 (0.868-1.001)	0.054	0.923 (0.866-0.984)	0.013
Duration of disease[b]								
≤ 6 months	11.752 (1.093-126.285)	0.042			22.131 (2.117-231.317)	0.010		
>6-12 months	0.864 (0.113-5.612)	0.878			1.185 (0.168-8.357)	0.865		
>12-24 months	1.651 (0.327-8.349)	0.544			1.994 (0.395-10.076)	0.404		
>24 months (Ref.)	1				1			

[a] Entered as a continuous variable by year. [b] Entered as a continuous variable by month.

Discussion

Pemphigus is a rare life-threatening autoimmune blistering disease characterized by vesicles, bullae and erosions on skin and mucous membrane. Various methods of disease severity assessment were widely used. We used PDAI to evaluate disease severity due to higher reliability and validity comparing to other tools in this study[9,10]. From a previous study, we found that pemphigus had a significantly negative impact on the patient's QOL not only about general health status, but also psychological and social aspects. Body image embarrassment and the problem of socialization resulted in more psychological problems, such as anxiety and depression[3]. To date, there were a few previous studies of the QOL in pemphigus patients, but there had not been a study in Thailand[4,5,22]. This study measured the QOL in Thai pemphigus patients by using the Thai-version of Skindex-29 questionaires, explored the correlation with the disease severity and also determined the variables that probably affected the QOL.

Our result showed the disease severity scores by PDAI had a positive correlation with Skindex-29 scores as same as the previous study which used Physician Global Assessment (PGA) and Ikeda index[3]. The mental status was the most affected aspect in the QOL due to the highest score in the emotion domain. Therefore, psychological support has an essential role in patient care. The significant difference of the QOL was shown between two groups: mild and severe groups and moderate and severe groups. While the mild and moderate severities were not statistically significantly different in the QOL. This indicates that the more severe disease will cause a great impact and result in poorer QOL.

In addition to the disease severity, our study showed other two variables that related to severe QOL impairment. First, the younger age was a significant factor associated with severe QOL impairment. On the contrary, older patients were associated with better QOL. This result was similar to the previous report on pemphigus and other skin diseases such as port-wine stain[3,23]. The age group of 30 to 40 years, who were the population of working age, had the most QOL impairment. They socialized and often stayed in the public and workplace. This might be the reason why the functioning score in this group was strikingly high compared with others. The disease might disturb the function and make them feel embarrassed or depressed with their appearance. While the older patients had lower social interaction and better coping mechanism. Therefore, the function and mental status might be less affected compared with the younger ones. Secondly, shorter duration of disease, especially less than 6 months was another associated factor for poor QOL while patients with disease duration longer than two years showed better QOL. This might be

explained by active disease in the initial phase which leaded to QOL impairment. Other variables, such as sexes, education level, subtypes of pemphigus, duration of disease, comorbidity, type and dosage of medication, were not significantly associated with severe QOL impairment. In contrast, Paradisi et al[3] showed that pemphigus foliaceus subtype, female sex and multiple comorbid diseases had a great negative impact on QOL. Sung et al[5]. reported a negative correlation between the dose of corticosteroid and the QOL, which might be from more adverse effects in the higher corticosteroid dosage group. In contrast to our study, the age and disease duration had no effect in both studies.

There were a few limitations in this study. First, the participants might not represent the pemphigus population due to data collecting from a single center and small sample size. Secondly, there might be a selection bias because the selected patients in the tertiary care center tended to have more severe disease. Finally, there are small numbers of patients in some treatment subgroups, such as cyclophosphamide and IVIG. Therefore, the association between medication type and QOL impairment was not able to clearly illustrate.

In conclusion, pemphigus is a chronic dermatologic disease that causes a great burden in physical, psychological and social aspects. Clinical severity, age and duration of disease were important factors related to QOL impairment. Therefore, the physician should evaluate clinical severity, concomitantly provide psychological and social support to achieve a holistic care approach and lead to a better patient's QOL.

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