Clinical features and course of pemphigus in Thai patients

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Summary

Background: **Pemphigus is a rare, organ-specific autoimmune disease. The epidemiology and clinical course vary between reports from different countries.**

Objective: **To evaluate clinical manifestations, investigation and clinical course of Thai patients with pemphigus.**

Methods: **Demographic data, clinical presentations, laboratory investigations and treatment outcomes in 124 pemphigus patients who had attended the specialized autoimmune skin clinic at Siriraj Hospital during the period from January 1991 to December 2009 were retrospectively studied.**

Results: **Of the 124 pemphigus patients, 79% were diagnosed with pemphigus vulgaris (PV) and 15.3% with pemphigus foliaceus (PF). The male to female ratio was approximately 1:2 in both groups. The mean age of onset was 45.4 years in PV patients and 57.4 years in PF patients. Oral mucosal involvement at the onset of disease was presented in 37.8% of PV patients. The sensitivity and specificity of DIF in the diagnosis of pemphigus was 97.8% and 98.3% while that of IIF was 94.7% and 98.4%. Disease control was achieved in 93.9% of PV patients and 94.7% of PF patients. Remission (off therapy) was achieved in 31.6% of patients in both groups.**

Conclusions: **PV is the most common subtype of pemphigus in Thailand and usually affects females more than males. The disease usually occurs in the fifth decade of life and mucosal involvement is common. Immunofluorescence studies yields very high sensitivity and specificity. Corticosteroids are the mainstay of treatment. The majority of patients attain disease control and one-third of them achieve remission (off therapy).** ***(Asian Pac J Allergy Immunol 2011;29:161-8)***

Key words: *clinical features course, pemphigus, Thai, treatment*

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Introduction

Pemphigus is an organ-specific autoimmune disease. It is caused by circulating autoantibodies directed against epidermal cell surfaces which result in disruption of epidermal cell adhesions and formation of intraepidermal blisters. Pemphigus is classified into 2 major forms depending on the level of blister located in epidermis; pemphigus vulgaris (PV), of which the blister is just above the basal layer, and pemphigus foliaceus (PF), the blister of which occurs more superficially. Other variants are pemphigus erythematosus (PE), pemphigus vegetans (PVeg), pemphigus herpetiformis (PH), paraneoplastic pemphigus (PNP) and IgA pemphigus.[1]

The incidences of pemphigus vary between 0.76 – 14 cases per million population per year depending on the location and tend to be higher in the countries located in lower latitudes.[2] Moreover, the proportion of each subtype of pemphigus, age of onset as well as sex distribution is also different from country to country. Reports from Asian countries such as Japan, Korea and Singapore show that PV is the most common subtype of pemphigus and male to female ratio ranges approximately from 1:1 to 1:2. The mean age of onset of PV patients is slightly higher than that of PF patients. The involvement of mucosa is about 75-84.4% of PV patients which is higher than the percentage in PF patients.[3-5] Corticosteroids are the mainstay of treatment in almost all studies but the adjuvant therapies used together with corticosteroids differ between studies. Azathioprine was the most common adjuvant drug used in a study from Korea whereas dapsone was widely used in a Singapore report.[3,4] In Japan, plasmapheresis is frequently used concurrently with systemic corticosteroids in

patients with severe symptoms and complete remission is achieved in 21% of cases.[5]

Piamphongsant et al.[6] studied 98 cases of pemphigus in Thai patients during 1978-1987. They were treated with corticosteroids alone or together with cyclophosphamide or dapsone and the reported remission rate was 32.8%. Moreover, another publication showed that prednisolone alone or the combination of prednisolone and cyclophosphamide have favorable outcomes in treatment of PV and PF in Thai patients.[7]

Although there are data concerning treatment aspect of pemphigus from Thailand, the clinical course and prognosis has not yet been documented. Thus, this study was designed awith the aim of evaluating clinical manifestations, investigations and the clinical courses of Thai patients with pemphigus.

Methods

Ethical approval was granted by Siriraj Institute Review Board, Siriraj Hospital, Mahidol University, Bangkok, Thailand. Data from case record forms of patients over the age of 18 with a diagnosis of pemphigus who had attended the specialized autoimmune skin clinic at Siriraj Hospital during the period from January 1991 to December 2009 were retrospectively studied. The diagnosis of pemphigus was based on clinical manifestations, histopathology and immunological findings as previous reported.[1] Patients who were diagnosed with other autoimmune vesiculobullous diseases or had never been followed up at the clinic were excluded from the study.

Data were recorded in a case record form that was particularly designed for this study. Data concerning age of onset, sex, presenting symptoms, characteristic of skin and mucosal lesions, laboratory investigations, treatment outcomes and clinical course were obtained.

Disease severity was classified according to the severity scale created by Herbst and Bystryn.[8] The scale is based on the compilation of the extent of disease and the intensity of therapy. The extent of disease was classified by the number of body areas involved, including scalp, face/neck, upper trunk, lower trunk, arms, legs, oral mucosa and genitals. A score 0 was given for no lesions, ½ for lesions healed within 48 hours, 1 for 1 site involved, 2 for 2 to 3 sites involved, 3 for 4 to 5 sites involved and 4 for ≥ 6 sites involved. The score for the intensity of therapy was given as follows: 0 for no treatment required, ½ for only topical treatment needed, and 1, 2, 3 and 4 for ≤ 15, 16 to 49, 50 to 89 and ≥ 90 mg of prednisolone (or equivalent) per day, respectively. If ≤ 100 mg/day of azathioprine/cyclopho-phamide, or gold, dapsone, or cyclosporine was used, an additional score of 1 was added. An extra score of 2 was added if > 100 mg/day of azathioprine/cyclophosphamide or plasmapheresis were used. Then the sums of these scores of ≤ 2+, 3 to 6+ and ≥ 7+ were used to classify the disease as mild, moderate and severe disease respectively.[8]

The definitions and observation end points for assessment of disease activity and treatment response, including control of disease activity, remission (on therapy), remission (off therapy) and relapse were defined by using criteria from International Pemphigus Committee.[9] To illustrate, the control of disease activity is the time at which existing lesions begin to heal and new lesions stop forming. Remission (on therapy) is defined as no lesions having developed for at least two months while the patients are receiving prednisolone ≤ 10 mg/day or adjuvant drugs at half of the maximum ineffective dosage. Remission (off therapy) occurs while the patients are off all systemic therapy and free of lesions for at least 2 months. Relapse takes place when patients develops ≥ 3 new lesions per month that do not heal without treatment.[9]

Statistical analysis was performed using the SPSS software version 17.0. Descriptive statistics and contingency tables were used to describe demographic data, clinical manifestations, laboratory investigation results and treatment outcome. Duration from treatment to each observation end point was analyzed by using Kaplan-Meier method of survival analysis. Furthermore, comparison of survival pattern in PV patients and PF patients was done by using log-rank test.

Results

A total number of 124 pemphigus patients diagnosed from 1991 to 2009 were studied. PV was the most common subtype followed by PF, PE and PH. We had no patients with PVeg, PNP and IgA pemphigus. Due to the small number of PE and PH patients, data from PV and PF patients was the primary focus. The PV to PF ratio was 5.3:1. The percentage of females was approximately double that of the male patients in the PV, PF and PH group. The mean age of onset was 45.4 years in PV and 57.4 years in PF patients, respectively (Table 1). The mean duration from the beginning of symptoms

Table 1. Demographic data of pemphigus patients

Disease	Gender			Total; n=124 Number (%)	Mean age of onset; years (range)
	Male Cases	Female Cases	M : F ratio		
PV	29	69	1 : 2.4	98 (79)	45.4 (18-104)
PF	6	13	1 : 2.2	19 (15.3)	57.4 (31-87)
PE	2	2	1 : 1	4 (3.2)	61.8 (53-65)
PH	1	2	1 : 2	3 (2.4)	39 (31-87)

PV, pemphigus vulgaris; PF, pemphigus foliaceus; PE, pemphigus erythematosus; PH, pemphigus herpetiformis

to time of diagnosis was 5.7 months (range 0-37 months) in PV patients and 6.6 months (range 1-24 months) in PF patients.

Table 2 shows clinical manifestations and disease severity of patients with PV and PF. The majority of PV and PF patients had lesions starting on skin. However, 37 of 98 patients (37.8%) in PV group had lesions beginning on the oral mucosa at the onset of disease and seven of them still had only oral mucosal involvement during the follow-up period (pure oral pemphigus). In contrast, none of PF patients had lesions confined only to the oral mucosa from the beginning of disease through to the last visit. Focusing on the disease severity, none of the patients had mild severity, but two-thirds of the patients in both groups had moderate severity of disease. Itching symptoms were presented in more than half of both diseases (50.8% and 60% respectively).

The most common abnormal full blood count finding in the PV group was leukocytosis (28.1%) while that in PF group was eosinophilia (22.2%). Tzanck's smear test demonstrated acantholytic cells in nearly two-thirds of both group (65.1% and 60% respectively). However, histopathologic findings revealed acantholytic cells in 89.3% of PV patients and 58.3% of PF patients. Regarding the type of cell infiltration, mixed cells were most frequently found in Tzanck's smear, which corresponded to the histological findings. The percentages of positivity for direct immunofluorescence (DIF) and indirect immunofluorescence (IIF) in both patient groups were over 90 as shown in table 3.

Table 4 demonstrates that the sensitivity of DIF for diagnosis of pemphigus in this study was slightly higher than that of IIF (97.8% and 94.7%), while the specificity of both tests was the same (98%). Furthermore, DIF and IIF yield high positive predictive values (PPV) and high negative predictive values (NPV) in patients with pemphigus. Nevertheless, among oral pemphigus patients, the specificity and PPV of both tests were much lower. The IIF titer in PV patients ranged from 1:10 to 1:5,120 (mean 1:202) whereas it ranged from 1:40 to 1:2,560 (mean 1:349) in PF patients. Serum antinuclear antibody was detected in two PV patients at the titer of 1:40 and 1:160 and both had a speckle pattern.

Table 2. Clinical manifestations and disease severity of patients with PV and PF

Clinical findings	Number of patients (%)	
	PV n=98	PF n=19
Morphology of skin lesions		
Vesicle/bullae	98 (100)	19 (100)
Base : Erythematous base	85/98 (86.7)	19/19 (100)
Normal skin base	66/98 (67.3)	14/19 (73.7)
Patch/plaque	43 (43.9)	13 (68.4)
Erosion/ulcer	61 (62.2)	9 (47.4)
Pustule	5 (5.1)	0 (0)
Scar	4 (4.1)	0 (0)
Site of involvement at onset		
Oral mucosa alone	37 (37.8)	0 (0)
Skin involvement alone	40 (40.8)	17 (89.5)
Both oral mucosa and skin	21 (21.4)	2 (10.5)
Site of involvement during clinical course		
Oral mucosa alone	7 (7.1)	0 (0)
Skin involvement alone	27 (27.6)	16 (84.2)
Both oral mucosa and skin	64 (65.3)	3 (15.8)
Disease severity[8]		
Mild	0 (0)	0 (0)
Moderate	71 (72.4)	13 (68.4)
Severe	27 (27.6)	6 (31.6)

Prednisolone was the only drug prescribed in 40 of 98 PV patients (40.8%) and 10 of 19 PF patients (52.6%), while adjuvant drugs in addition to prednisolone were used in 57 of 98 PV patients (58.2%) and 9 of 19 PF patients (47.4%). The most frequently prescribed adjunctive drug was cyclophosphamide followed by dapsone in both groups. In contrast, one PV patient received only dapsone during the follow up periods but also achieved remission. Mean initial doses of prednisolone were equal in both groups (0.9 mg/kg/day). The mean accumulative dose of prednisolone (or equivalent) required to achieve remission was 0.13 g/kg in patients with PV and 0.11 g/kg in patients with PF.

Almost all PV and PF patients achieved disease control (93.9% and 94.7%, respectively) as shown in

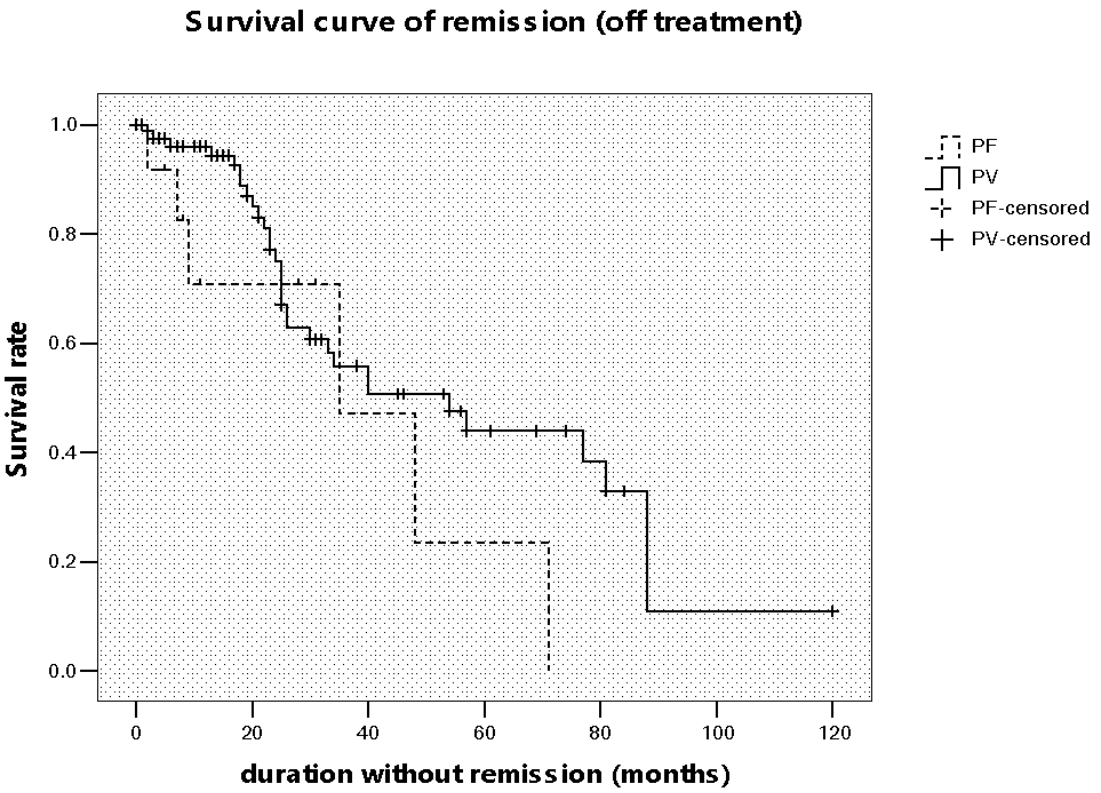

Figure 1. Kaplan Meier survival curve of remission (off treatment) in patients with PV and PF. One-year remission rate is 4% in patients with PV and 29.3% in patients with PF, whereas five-year remission rate is 56.1% and 76.4% in patients with PV and PF, respectively.

Table 5; however, only one-third of them (31.6%) attained remission (off therapy). The median duration to disease control, remission on therapy and off therapy in patients with PV was longer than that in patients with PF; nevertheless there was no statistically significant difference. Among the patients who achieved remission (off therapy), relapse occurred in 10 of 31 PV patients (32.3%) and 3 of 6 PF patients (50%). Mean follow-up period was 33.3 months in PV patients and 20.8 months in PF patients.

Among patients with PV, corticosteroid was used as a single agent in patients with moderate disease severity more often than patients with severe disease. In contrast, both corticosteroid and adjuvant drugs were used in patients with severe disease more than in patients with moderate disease severity (p = 0.001). The therapeutic outcome was not different between patients treated with corticosteroid alone and patients treated with corticosteroid together with adjuvant drugs when analyzed by disease severity ($p > 0.05$). However, the median duration to achieve disease control, remission (on therapy) and remission (off therapy) in patients treated with corticosteroid alone tended to be shorter than that of patients treated with both corticosteroid and adjuvant drugs.

Since there is no definitive consensus on the definition of dose of corticosteroid, this study defined the use of prednisolone or equivalent $\geq$ 1 mg/kg/day as a high dose, and < 1 mg/kg/day as a low dose. Focusing on the initial dose of corticosteroid prescribed to PV patients, there was no discrepancy in the treatment outcome of patients treated with low dose as compared to a high dose of corticosteroid.

Since some patients were lost to follow up, the Kaplan Meier survival curve was used to study the time to remission (off treatment) in patients with PV and PF as shown in figure 1. One-year and five-year

Table 3. Laboratory investigations in patients with PV (n=98) and PF (n=19)

Investigations	Number of patients (%)	
	PV	PF
Complete blood count		
Total studied cases	89	18
Anemia (hematocrit ≤ 35%)	11 (12.4)	3 (16.7)
Leukocytosis (WBC ≥ 12,000 cell/µL)	25 (28.1)	3 (16.7)
Neutrophilia (neutrophil > 80%)	15 (16.9)	2 (11.1)
Eosinophilia (eosinophil > 600 cell/µL)	10 (11.2)	4 (22.2)
Tzanck's smear		
Total studied cases	43	5
Positive acantholytic cells	28 (65.1)	3 (60)
Neutrophil predomination	11/28 (39.3)	0 (0)
Eosinophil predomination	2/28 (7.1)	1/3 (33.3)
No predominant cell infiltration	15/28 (53.6)	2/3 (66.6)
Direct immunofluorescence		
Total studied cases	91	17
Positive	86 (94.5)	16 (94.1)

DIF, direct immunofluorescence; IIF, indirect immune-fluorescence; WBC, white blood cell

Table 4. Sensitivity, specificity, PPV and NPV of DIF and IIF in diagnosis of pemphigus in this study

Test	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)
DIF				
Pemphigus group	97.8	98.3	98.9	96.6
Oral pemphigus	100	40.4	2.3	100
IIF				
Pemphigus group	94.7	98.4	99.1	90.9
Oral pemphigus	71.4	38.1	4.6	97

PPV, positive predictive value; NPV, negative predictive value

remission rate in patients with PV was 4% and 56.1% respectively, whereas they were higher in patients with PF (29.3% at one-year and 76.4% at five-year). However, there was no statistical significant difference between the two groups of patients.

Of 124 pemphigus patients, 60 (48.4%) experienced side effects from drugs. A Cushinoid appearance was the most common adverse drug reaction found (55%), followed by steroid-induced acne (20%), hyperglycemia (18.3%), infection (15%), weight gain (11.7%), myopathy (6.7%), amenorrhea (3.3%) and anemia (1.7%). Serious infection associated with mortality occured in 3 patients (2.4%). All of them were PV patients treated with high dose corticosteroid and had been admitted to the hospital. During the hospital stay, they later developed septic shock which resulted in death.

Regarding the 4 patients with PE, the number of males and females was equal. The mean age of onset was 61.8 years and mean duration from the beginning of symptoms to time of diagnosis was 4.2 months. All of them had itching symptoms and the skin was affected at the first presentation. Moreover, IIF was positive in 3 of 4 patients (75%) while DIF was positive in all of them. One patient treated with prednisolone and cyclophophamide achieved remission (off therapy) whereas the others only achieved control of disease activity.

Among the patients with PH, the mean age of onset was 39 years and the mean duration from onset to diagnosis was 4.5 months. All three of them had itching and skin lesions at the first visit. DIF and IIF tests were positive in all. Furthermore, prednisolone together with dapsone was prescribed and they all reached disease control. However, only one of them achieved remission (off therapy) in 4 years.

Discussion

Pemphigus has a variety of epidemiological profiles in different regions of the world. Our study reveals that PV is the most common subtype of pemphigus diagnosed in Thailand, which is similar to reports from most countries.[3,4,10-16] The PV to PF ratio was 5.3:1 which was comparable with reports from India and Croatia.[15,16] Our data show that pemphigus affected females more than twice as often as males; however, a male preponderance is found in Bangladesh and Switzerland (male to female ratios 1.3:1 and 2.5:1, respectively).[12,13] In our study, the mean age of onset of PV was in the fifth decade of life which is similar to the previous studies of Piamphongsant et al.[6] and those from other Asian countries, except for the report from Japan in which the mean age was in the sixth decade (52.8 years).[3-5,15] On the other hand, the reported mean age of onset was higher in European countries and was between the sixth and seventh decade of life.[11,12,16,17]

Patients with PV often have mucous membrane involvement but the initial sites of lesions vary depending on the geographic area. Oral lesions were reported to be present at the onset of PV in approximately two thirds of patients (59.3–82.1%); however, a study from Bangladesh revealed only 26% of PV patients had lesions which started on oral mucosa.[13,18-21] In our study, 72.4% of patients with PV had oral involvement during the clinical course while 59.2% had oral lesions present at the

Table 5. Clinical course of PV and PF patients

Clinical course	PV	PF	p value*
Disease control†			
Total studied cases	98	19	
Number of patients (%)	92 (93.9)	18 (94.7)	
Duration between treatment and disease control; median (days)	43	28	0.35
Remission (on therapy)‡			
Total studied cases	98	19	
Number of patients (%)	64 (65.3)	10 (52.6)	
Duration between treatment and remission on therapy; median (months)	10	9	0.80
Remission (off therapy)§			
Total studied cases	98	19	
Number of patients (%)	31 (31.6)	6 (31.6)	
Duration between treatment and remission off therapy; median (months)	54	35	0.16
1-year remission rate (%)	4	29.3	
5-year remission rate (%)	56.1	76.4	
Relapse‖			
Total studied cases	31	6	
Number of patients (%)	10 (32.3)	3 (50)	
Duration between off treatment and relapse; median (months)	41	32	0.38

*Log-rank test

†Disease control: the time at which existing lesions begin to heal and no new lesions occur

‡Remission on therapy: no lesion having developed for at least two months while the patients have received prednisolone ≤ 10 mg/day or adjuvant drugs at half of the maximum ineffective dosage.

§Remission off therapy: lesion free while the patient has been off all systemic therapy for at least 2 months

‖Relapse: development of ≥ 3 new lesions/month that do not heal without treatment in a patient who has achieved remission (off therapy)

onset of the disease. Even though mucosal involvement is uncommon in patients with PF, our study demonstrates that three PF patients (15.8%) had oral lesions during the course of the disease. Additionally, reports from Tunisia and Kuwait show mucosal involvement in 6.7% and 13% of patients with PF, respectively.[14,20] Approximately half of patients with PV and PF in our study had itching symptoms. Perhaps the warm weather and high humidity in Thailand may account for this.

Tzanck's smear is a useful tool in making the diagnosis of pemphigus. Durdu et al.[22] report 100% sensitivity and 43.4% specificity of Tzanck's smear in diagnosis of pemphigus. On the contrary, our data show the presence of acantholytic cells in only 65.1% of PV patients and 60% of PF patients. The lower sensitivity of Tzanck's smear in our patients may be due to a variety of stages of lesions and techniques of taking Tzanck's smear. Concerning the immunofluorescence study, DIF results from our study yielded a high sensitivity and a high PPV in the diagnosis of pemphigus compared to previous studies.[23] The sensitivity of IIF in pemphigus patients varies according to disease activity and the substrate used.[24,25] Our result showed relatively high sensitivity of IIF in pemphigus patients (94.7%) owing to the fact that serum was obtained while the disease was active in the patients. Focusing on our data from patients with pure oral pemphigus, DIF sensitivity was as high as 100%, which is similar to preceding reports.[26,27] However, the sensitivity of IIF was much lower and related data are limited.

Corticosteroids are the major drugs used in pemphigus patients because they are able to reduce autoantibody levels and also dramatically decrease the mortality rate.[1,28] Nevertheless, high dose administration and prolonged usage of corticosteroids may bring about some serious complications. All patients in our study, except one patient with PV, received prednisolone at a mean initial dose of 0.9 mg/kg/day. Adjuvant drugs were additionally prescribed to 58.2% of PV patients and 47.4% of PF patients to achieve disease control. The advantages of adjuvant drugs are the steroid-sparing effect and the augmentation of steroid efficacy.[1,6,29] The most widely used adjuvant drugs are azathioprine and cyclophosphamide. Other reported adjunctive drugs included cyclosporine, gold, dapsone, nicotinamide and tetracycline.[1,29] Mycophenolate mofetil, a relatively new agent, has been reported by Beissert et al.[30] to have a similar efficacy as azathioprine. Other treatment modalities used are plasmapheresis, extracorporeal photopheresis and intravenous immunoglobulin.[1] Cummins et al.[31] used oral cyclophosphamide in combination with prednisolone in 23 refractory cases of patients with PV and PF. As a result, nineteen of them (83%) achieved complete remission, defined as no lesions for at least 4 weeks while a low dosage of prednisolone was used (up to 0.15 mg/kg/day). In our study, cyclophosphamide was the most frequently prescribed adjuvant drug.

The second most common prescribed adjunctive drug in our center was dapsone, especially for patients who have contraindications to the use of immunosuppressive drugs. One patient with PV was treated with dapsone monotherapy and achieved remission (off therapy). Gurcan et al.[32] analyzed 55 pemphigus patients treated with dapsone from the literature published between 1969 and 2008. They state that 86% of PV patients and 78% of PF patients responded to treatment either with dapsone alone or dapsone concurrently with prednisolone.

Since corticosteroids and adjuvant drugs have been introduced as a treatment, more pemphigus patients achieve remission.[8,28,29,33] There are many studies concerning the results of pemphigus treatment; nevertheless, interpretation and comparison of outcomes is difficult because the definitions of remission are inconsistent when defined by different authors. Using the definitions of the disease, end points and therapeutic response defined by International Pemphigus Committee[9], a study of 155 pemphigus patients in Israel revealed that all pemphigus patients achieved disease control at an average time of 3 months and one-third of them reached complete remission (off treatment) at the end of follow-up.[34] However, our data shows that disease control was achieved in close to 95% of patients while the mean time to control was shorter. Besides, one-third of our patients were in remission (off treatment). We looked at the patients who did not achieve disease control and found that two had died due to sepsis, four were referred to other hospitals closer to their residence and the other one was lost to follow up. Thus, the actual control rate may be higher than stated. The different ethnicity, younger age of onset and dissimilar treatment regimen may be resposible for the shorter time to control.

Zaraa et al.[20] studied the clinical course of PV and PF and concluded that both diseases share the same prognosis. Relapse occurred in 32% of PV patients and 53% of PF patients. Even though our patients with PV required a higher dose of prednisolone to achieve remission than patients with PF, we found no difference in time to disease control, remission and relapse. The percentage of patients with relapse was about the same as in previous reports.

Our results reveal that the additional use of adjuvant drugs to corticosteroid did not significantly alter the therapeutic outcome in PV patients. Surprisingly, patients who received both corticosteroid and adjuvant drugs had longer durations to disease control, remission (on therapy) and remission (off therapy) than patients treated with only corticosteroid. This may be owing to the fact that adjuvant drugs were added to corticosteroid when there were complications from corticosteroid use, or in patients with severe disease were partially responsive to corticosteroid, or when the disease flared up while lowering corticosteroid dose.

The mortality rate of pemphigus patients has decreased to less than 10% since adjuvant drugs have been used. Unfortunately, complications from such treatment are the most common causes of death.[3,29,35] Our data also supports these reports. We found 2.4% mortality rate in our patients and all of them had been treated between 1993 and 1999. They all had PV, had been treated with high dose corticosteroids and died of sepsis which resulted from the complications of corticosteroids. The low to moderate dose of corticosteroids that were prescribed in our practice may account for the lower mortality rate. On the other hand, side effects were encountered by half of our patients and most were the consequence of corticosteroids. A Cushinoid appearance was the most common adverse reaction, followed by steroid-induced acne and hyperglycemia. Although infection was seen in only 15% of our patients, prompt treatment of infection is necessary because serious infection can lead to death. A limitation of our study was that slit lamp examination and bone scans were not performed in all cases so that cataracts and osteoporosis could have been missed; nonetheless, a calcium supplement was prescribed to all patients receiving prednisolone.

In conclusion, the present study is the first to reveal the clinical manifestations, laboratory investigations, management and clinical course of pemphigus in Thai patients. PV is the most common form and affects females more often than males. The disease usually occurs in the fifth decade of life and mucosal involvement is common in PV patients. Immunofluorescence studies yield a very high sensitivity and specificity. Corticosteroids are the mainstay of treatment and cyclophosphamide is the most common adjuvant drug prescribed. The majority of patients attain disease control and one-third of them achieve remission (off therapy).

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Conflicts of interest

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