

Clinical Characteristics and Treatment Outcomes of 53 Pustular Psoriasis Patients

Supanan Prasertyothin MD*, Leena Chularojanamontri MD*,
Chanisada Wongpraparut MD*, Narumol Silpa-archa MD*

* Department of Dermatology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand

Objective: To study clinical characteristics, treatment outcomes, and clinical course of pustular psoriasis in Thai patients.

Material and Method: A retrospective medical chart review of Thai pustular psoriasis patients aged older than 15 years attended the Dermatology Clinic at Siriraj Hospital during July 2002 to October 2014 was conducted. Clinical feature, treatment modality, treatment response, and clinical course were reviewed.

Results: Fifty-three cases of pustular psoriasis were identified and studied. Female to male ratio was 1.9:1. Mean age was 42.2 years (SD = 15.8) and mean age at onset was 36.6 years (SD = 17.2). Twenty-three patients had concomitant plaque-type psoriasis vulgaris preceded pustular psoriasis. A mean time from onset of plaque-type psoriasis to development of pustular psoriasis was 7.9 years. The most common manifestations of pustular psoriasis were exanthematic type (60.4%), followed by localized type (22.6%) which included acrodermatitis continua of Hallopeau (13.2%) and pustulosis palmaris et plantaris (9.4%). Three common precipitating factors were inadequate sleep (30.2%), infection (30.2%), and medication (28.3%). Systemic corticosteroids withdrawal, accounting for 60% of medication-induced pustular psoriasis, was found to induce pustular psoriasis with an average onset of 1.7 weeks after withdrawal. Topical corticosteroids were the most common prescribed treatment (98.1%) while acitretin was the most frequently used systemic treatment (75.5%), followed by methotrexate (43.4%). Generalized type (exanthematic, acute generalized pustular psoriasis of Von Zumbusch and annular types) had rapid clearance of pustules ranged from 2-4 weeks, except for localized type which extended until 18 weeks. At one month, generalized type showed significant clearance at 70% compared to 25% in localized type ($p < 0.05$). Recurrence was detected in 70% of all cases at a median time of 8 weeks. No mortality or severe complications from pustular psoriasis or treatment-related conditions was reported.

Conclusion: Pustular psoriasis is a rare condition in Thailand. Although some patients had severe forms of the disease, most patients had a benign clinical course that was controllable. However, relapse was common.

Keywords: pustular psoriasis, psoriasis, Thai, clinical characteristic

J Med Assoc Thai 2017; 100 (5): 565-72

Full text. e-Journal: <http://www.jmatonline.com>

Psoriasis is a chronic inflammatory skin disease with multifactorial etiologies, including genetic susceptibility, imbalance in T-cell-mediated immunity, and environmental factors⁽¹⁻⁵⁾. Pustular psoriasis is a rare variant of psoriasis characterized by sterile pustules on erythematous skin. Prevalence of pustular psoriasis among psoriasis types is low, as indicated by studies of Japan and Thailand of 2.1% and 1.2%, respectively^(6,7). Five clinical subtypes of pustular psoriasis have been classified; acute generalized pustular psoriasis (GPP) of Von Zumbusch, impetigo

herpetiformis, annular pustular psoriasis, exanthematic pustular psoriasis, and localized pustular psoriasis⁽¹⁾. To our knowledge and based on literature review, few studies have explored the epidemiology and clinical outcome of pustular psoriasis. This study aimed to investigate clinical characteristics, treatment modalities, and clinical course of pustular psoriasis in Thai patients.

Material and Method

This retrospective medical chart review included case record form of pustular psoriasis patients aged more than 15 years who attended the Dermatology clinic, Siriraj Hospital during July 2002 to October 2014. Diagnosis of pustular psoriasis was confirmed by dermatologist. In some cases, where definitive diagnosis could not be established from

Correspondence to:

Silpa-archa N, Department of Dermatology, Faculty of Medicine Siriraj Hospital, Mahidol University 2 Wanglang Road, Bangkoknoi, Bangkok 10700, Thailand.

Phone: +66-2-4194333, Fax: +66-2-4115031

E-mail: doctornarumol@gmail.com

clinical presentation, skin biopsies were performed. Collecting data in medical record form involved age, age onset of pustular psoriasis, gender, history of previous psoriasis vulgaris, family history of psoriasis, underlying diseases, precipitating factor of pustular psoriasis, type of pustular psoriasis, body surface area (BSA) involvement (1 palm included fingers = 1% BSA) and anatomical location (s) of pustules at first onset, treatment modalities, and treatment response. Type of pustular psoriasis was defined as [1] acute GPP of Von Zumbusch: characterized by sudden onset of fever and disseminated sterile pustules on erythematous skin all over the body, [2] impetigo herpetiformis: acute GPP occurs in pregnancy, [3] exanthematic pustular psoriasis: widespread pustules occur within psoriatic plaque lesions in plaque-type psoriasis patient, [4] annular pustular psoriasis: pustules presenting on annular erythematous skin, and [5] localized pustular psoriasis which divided to [5.1] acrodermatitis continua of Hallopeau: pustules presenting on fingertips and nails, and [5.2] pustulosis palmaris et plantaris: pustules presenting on palms and soles⁽¹⁾. In the present manuscript, the authors used the umbrella term “Generalized type” for acute GPP of von Zumbusch, impetigo herpetiformis, exanthematic and annular types. The term “Localized type” was used for acrodermatitis continua of hallopeau and pustulosis palmaris et plantaris. For treatment outcome, the median time to the first complete clearance of pustules was reported. Recurrence was defined as detection of new pustules after the first complete clearance.

The present study was approved by the Siriraj Institutional Review Board, Siriraj Hospital, Mahidol University, Bangkok, Thailand.

The PASW® Statistics version 18 (SPSS, Inc., Chicago, IL, USA) was used for statistical data analysis. Categorical data were described as frequencies and percentages. Continuous data were described as mean and standard deviation (SD). Kaplan-Meier analysis was applied for treatment response data.

Results

Patient characteristics

There were 53 pustular psoriasis patients, of which 35 (66%) were female and 18 (34%) were male. Table 1 demonstrated clinical characteristics of all 53 patients. Fourteen patients required skin biopsy for definitive diagnosis. Mean age onset of pustular psoriasis was 36.6 years (SD = 17.1). Dyslipidemia,

hypertension, and diabetes mellitus were the three most common comorbidities. Likewise, the three most common precipitating factors were inadequate sleep, infection, and medication. Systemic corticosteroids were the most common associated drug (9, 60%), which induced post-withdrawal pustular psoriasis with a mean duration of 1.7 weeks (1-4 weeks). Fourteen (26.4%) pustular psoriasis patients were admitted due to fever and large area of pustular lesions.

Of 53 patients, plaque-type psoriasis occurred before onset of pustular psoriasis was found in 23 (43.4%) patients with a mean duration to onset of pustular psoriasis at 7.9 years (SD = 5.6). Likewise, pustular psoriasis presented as the first manifestation in 30 (56.6%) patients, Of which 8 patients (36.4%) developed plaque-type psoriasis.

Clinical manifestation

Of the 5 classified types in 53 patients, the most prevalent type was exanthematic (32, 60.4%), followed by localized type (12, 22.6%), acute GPP of von Zumbusch (4, 7.5%) and annular type (4, 7.5%). One patient had impetigo herpetiformis, a 25-year-old primigravida female developed pustular lesions at 20th week of pregnancy. A mean % BSA involvement in all 53 patients was 26.7% (SD = 28.1).

Thirty-three patients (64.2%) had nail involvement. Nail bed lesions (onycholysis, subungual hyperkeratosis, oil spots) were more common than nail matrix (pitting, leukonychia). Twelve patients (36.4%) had both nail matrix and nail bed involvement. Seven patients (13.2%) had psoriatic arthritis, which was diagnosed by rheumatologists. Large joints (elbow, knee, wrist, ankle) and small joints (hand, foot, finger, toe) were affected in four and three patients, (7.5%, 5.7%), respectively. Leukocytosis was the most common abnormal laboratory investigation that can be detected in 18 patients (34%). Others included abnormal liver function test (transaminitis, bilirubinemia, hypoalbuminemia) at initial diagnosis in 13 patients (24.5%), hypercholesterolemia in 12 patients (22.6%) and high blood sugar in four patients (7.5%).

Treatment and outcome

Treatment options based on pustular psoriasis type were shown in Table 2. Oral acitretin (0.25-1 mg/kg/day) was the most prescribed treatment in all types, followed by methotrexate (0.2-0.3 mg/kg weekly) and cyclosporine (2-4 mg/kg/day) in 75.5%,

Table 1. Demographic characteristics of pustular psoriasis patients (n = 53)

Characteristics	No. of patients (%)
Mean age 42.2±15.8 years	
Mean age at first onset 36.6±17.1 years	
Gender	
Female/male	35 (66) /18 (34)
Family history of psoriasis	4 (7.5)
Comorbid diseases	
Dyslipidemia	14 (26.4)
Hypertension	9 (17)
Diabetes mellitus	6 (11.3)
Coronary heart disease	3 (5.7)
Stroke	2 (3.8)
Obesity	2 (3.8)
HIV	2 (3.8)
Hepatitis	2 (3.8)
Cirrhosis	2 (3.8)
Precipitating factors	
Inadequate sleep	16 (30.2)
Infection	16 (30.2)
Drug (prednisolone ^[9] , herb ^[5] , over-the-counter analgesic ^[11])	15 (28.3)
Stress	12 (22.6)
Menstruation	7 (13.2)
Food	3 (5.7)
Smoking	2 (3.8)
Alcohol	1 (1.9)
Smoking history	
Current	5 (9.4)
Ever	2 (3.8)
Alcohol consumption history	
Current	2 (3.8)
Ever	4 (7.5)
Type of pustular psoriasis	
Generalized	41 (77)
Exanthematic	32 (60.4)
Acute GPP of Von Zumbusch	4 (7.5)
Annular	4 (7.5)
Impetigo herpetiformis	1 (1.9)
Localized	12 (22.6)
Acrodermatitis continua of Hallopeau	7 (13.2)
Pustulosis palmaris et plantaris	5 (9.4)
Symptoms	
Fever	18 (34)
Pain	16 (30.2)
Itching	7 (13.2)
Arthralgia	3 (5.7)
Myalgia	2 (3.8)
Anatomical locations at first presentation	
Trunk	34 (64.2)
Lower extremities	34 (64.2)
Upper extremities	31 (58.5)
Scalp	16 (30.2)
Hand	10 (18.9)
Fingers and toes	10 (18.9)
Face	8 (15.1)
Feet	7 (13.2)
Nail involvement	33 (64.2%)
Nail matrix (pitting, leukonychia)	8 (24.2%)
Nail bed (onycholysis, subungual hyperkeratosis, oil spots)	13 (39.4%)
Both nail matrix and nail bed	12 (36.4%)

Table 2. Treatment options based on pustular psoriasis type (n = 53)

	Total No.	Exanthematic n = 32	Localized n = 12	Annular n = 4	Von Zumbusch n = 4	Impetigo herpetiformis n = 1
Systemic treatment						
Acitretin	40	25	8	3	4	-
Methotrexate	23	17	3	1	2	-
Cyclosporine	4	3	1	-	-	-
Prednisolone	1	-	-	-	-	1
Topical treatment						
Corticosteroids	52	32	11	4	4	1
Tar	20	14	1	3	2	-
Vitamin D3 analogue	15	6	8	1	-	-
Phototherapy						
NB-UVB	2	2	-	-	-	-
PUVA	3	-	2	1	-	-
Biologics	2	-	1	-	1	-
Recurrence	37	22	9	2	3	1

NB-UVB, narrowband ultraviolet B; PUVA, psoralen plus ultraviolet A

Table 3. Median time to complete clearance of pustules based on type of pustular psoriasis

Types (n)	Weeks (range)
Generalized (41)	
Annular (4)	2 (1-4)
Acute GPP of Von Zumbusch (4)	2.5 (1-8)
Exanthematic (32)	4 (1-64)
Impetigo herpetiformis (1)	14
Localized (12)	18 (2-112)

43.4%, and 7.5% of cases, respectively. In most patients (98.1%), topical corticosteroids were combined with systemic treatment. The single impetigo herpetiformis case was commenced on systemic corticosteroids for 6 months, with an initial dose of 1 mg/kg/day that was gradually reduced over the course of treatment. She carried to full term and delivered a normal birth weight child without any abnormality. In the postpartum period, the disease relapsed and treatment was switched to acitretin with partial response. There were two cases that received biologic treatments. One refractory case of acrodermatitis continua of Hallopeau who failed to topical corticosteroids, local psoralen plus UVA (PUVA) therapy, acitretin, and methotrexate had initially excellent responded to etanercept 25-50 mg/week with acitretin 25-50 mg/day. In a subsequent relapse of the disease, this combination was unable to control her condition. The patient was later given two separate doses of ustekinumab 90 mg with no

significant improvement. This case was previously reported⁽⁸⁾. The other patient had plaque-type psoriasis with psoriatic pan-uveitis that developed into Von Zumbusch type when oral prednisolone dose was tapered off due to development of ocular lesions. She received topical corticosteroids and acitretin 10-25 mg/day with poor response. Patient was given infliximab 200 mg once a month for 6 months and her ocular and skin lesions improved. When her clinical condition converted to erythrodermic psoriasis, she was given methotrexate 7.5 mg/week with partial response.

Clinical course of pustular psoriasis was demonstrated in Table 3. The median time to complete clearance of pustules in all types was one month. Exanthematic, acute GPP of Von Zumbusch and annular types had rapid clearance ranged from 2-4 weeks, while localized type prolonged until 18 weeks. From statistical analysis, one and two months following treatment, 60% and 90% of all patients had complete clearance of pustules, respectively. At one month, exanthematic, annular, acute GPP of Von Zumbusch types and impetigo herpetiformis showed significant complete clearance of pustules at 70% compared to 25% in localized type ($p < 0.05$). Recurrence was detected in 70% of all pustular psoriasis cases at a median time of 8 weeks after complete clearance. However, none of the patients developed mortality, severe complications from pustular psoriasis or treatment-related conditions.

Table 4. Comparing clinical characteristics of pustular psoriasis with other published studies

	Baker and Ryan ⁽¹²⁾ n (%)	Zelickson and Muller ⁽¹⁴⁾ n (%)	Tay YK, et al. ⁽⁹⁾ n (%)	Borges-Costa, et al. ⁽¹⁵⁾ n (%)	Choon SE, et al. ⁽²²⁾ n (%)	Jin H, et al. ⁽¹¹⁾ n (%)	This study n (%)
No. of patients	104	63	28	34	102	33	53
Gender							
Female	61(59)	32(50)	16 (57)	13 (38)	68 (67)	18 (55)	35 (66)
Male	43(41)	31(50)	12 (43)	21 (62)	34 (33)	15 (45)	18 (34)
Mean age, years (range)	³ / ₄ of patients aged >40	50 (N/A)	37.5 (4-77)	58 (N/A)	48 (22-89)	45 (N/A)	42 (15-78)
Precipitating factors							
Upper respiratory tract infection	N/A	11(17)	9 (32)	N/A	16 (16)	6 (18)	16 (30)
Systemic corticosteroids	37(36)	N/A	N/A	N/A	45 (44)	2 (6)	9 (17)
Generalized type	104 [#]	56	19	34	102	33	41
Von Zumbusch	92 (88)	35 (56)	7 (25)	34 (100)	78 (76)	21 (64)	4 (8)
Annular	10 (10)	15 (24)	2 (7)	-	3 (3)	7 (21)	4 (8)
Juvenile	-	-	6 (21)	-	-	3 (9)	-
Impetigo herpetiformis	-	-	1 (4)	-	17 (17)	-	1 (2)
Exanthematic/ Localized of GPP	-	-	3 (11)	-	4 (4)	2(6)	32 (60)
Mixed type*	-	6 (9)	-	-	-	-	-
Localized type	N/A	7	9	N/A	N/A	N/A	12
Acrodermatitis continua of Hallopeau		N/A	4 (14)				7 (13)
Pustulosis palmaris et plantaris		N/A	5 (18)				5 (9)
Recurrence	N/A	N/A	N/A	N/A	102 (100)	19 (76)	37 (70)

N/A: not available, GPP: generalized pustular psoriasis

Two patients were defined as unclassified type

*Patients had some localized area of pustular psoriasis that spread to become a generalized flare

Discussion

To our knowledge, very few studies have been undertaken to investigate the prevalence and clinical aspects of pustular psoriasis in Asians⁽⁹⁻¹¹⁾. Based on our data, pustular psoriasis was commonly found in the fifth-decade women, which is consistent with Asian studies^(9,10).

A study from England, however, reported an age range of 40-60 years in generalized pustular psoriasis⁽¹²⁾. Table 4 demonstrated clinical characteristics of published studies.

Obesity, hypertension, diabetes mellitus, and dyslipidemia are known co-morbidities in patients with plaque-type psoriasis⁽¹³⁾. However, only limited associated morbidity information has been reported in pustular psoriasis. The co-morbidities found in the present study (dyslipidemia, hypertension, diabetes mellitus) were similar to plaque-type psoriasis. These metabolic conditions may affect selection of treatment, especially in localized pustular psoriasis which required longer course of treatment. Among commonly

reported factors that aggravated pustular psoriasis (infection or withdrawal of systemic corticosteroids^(2,3,9,14), the present study found that upper respiratory tract infection is the most common precipitating factor. Pustular psoriasis can occur as a first onset (de novo) manifestation⁽¹⁰⁾ or be preceded by plaque type psoriasis^(14,15). The authors found 56.6% patients had de novo pustular psoriasis while 43% patients had previous psoriasis vulgaris prior to onset of pustular psoriasis. In the latter group, mean duration to onset of pustular psoriasis was 7.9 years, which is longer than the Ohkawara et al. study, which reported a mean duration of 6 years⁽¹⁰⁾. Interestingly, studies from Japan proposed that de novo generalized pustular psoriasis is a distinct subtype caused by deficiency of interleukin (IL)-36 receptor antagonist^(16,17). Further investigations should be done to discover new treatment that target to this defect.

Acute GPP of Von Zumbusch is more common than localized pustular psoriasis in adult^(9,12,18), while annular pustular psoriasis is the most common form of

pustular psoriasis in children. Our study found exanthematic pustular psoriasis to be the most common type. This condition was commonly observed in lesions of plaque-type psoriasis with disease exacerbation.

A rare condition of impetigo herpetiformis was observed in one patient, and this low prevalence was consistent with Singapore study⁽⁹⁾. Systemic corticosteroids, cyclosporine, and topical corticosteroids are the first-line treatment⁽¹⁹⁾. Resolution of symptom occurs after delivery and recurs in subsequent pregnancy. Maternal-fetal complications include stillbirth or neonatal death caused by placental insufficiency⁽²⁰⁾.

Liver function test abnormality was found in 24.5% in all types of pustular psoriasis, which is lower than a Portuguese study which reported 47% in only acute GPP type⁽¹⁵⁾. These abnormalities were an associated findings which returned to normal value after disease resolution⁽¹⁵⁾. Therefore, screening for liver function test should be done at baseline before commencing treatment.

Treatment of pustular psoriasis is determined according to disease type, severity, underlying comorbidities, and laboratory abnormalities⁽¹⁹⁾. Oral acitretin is the first-line therapy, with methotrexate and cyclosporine also delivering effective results⁽¹⁹⁾. In our patients, oral acitretin was the most frequent treatment, followed by oral methotrexate in all types of pustular psoriasis. Cyclosporine was prescribed in only 3 patients, due to high drug cost compared to methotrexate. Moreover, some evidence indicates that the efficacy of methotrexate in pustular psoriasis is slightly better than cyclosporine⁽²¹⁾. Overall, the present study found satisfactory response from systemic treatment with 60% and 90% of patients showed complete clearance of pustules at one and two months, following treatment with significant rapid clearance in generalized type (exanthematic, acute GPP of Von Zumbusch and annular types). Similarly, Korean study showed lesion clearance of 68% in two months after treatment initiation⁽¹¹⁾. However, the authors found common recurrence of disease (70%) which was similar to Korean and Malaysian studies that reported recurrence of 76% and 100%, respectively^(11,22). No mortality or severe complications (e.g., sepsis) was observed in this study. The clinical course of pustular psoriasis was relatively benign with report of mortality in few cases^(9,14,15).

Conclusion

Pustular psoriasis is a rare condition. Clinical

subtype influences different clinical outcome. Generalized type (exanthematic, acute GPP of Von Zumbusch, and annular types) had rapid response to treatment compared to localized type. Recurrence was common in all types, however, most of the cases had benign clinical course and the disease was controllable. Overall, the authors highlighted the clinical characteristics, treatment outcome, and clinical course of pustular psoriasis in Thailand.

What is already known on this topic?

- Pustular psoriasis is a rare form of psoriasis which required systemic treatment.

What this study adds?

- Oral acitretin is the most commonly prescribed treatment.
- Type of pustular psoriasis influenced the clinical outcome.
- Generalized type of pustular psoriasis (exanthematic, acute generalized pustular psoriasis of Von Zumbusch, and annular types) had rapid response than localized type.
- Disease recurrence was common in both generalized and localized forms.

Acknowledgment

The authors gratefully acknowledge Mr. Suthipol Udompunthurak for his assistance in statistical analysis.

Potential conflict of interest

The authors hereby declare no personal or professional conflicts of interest regarding any aspect of this study.

References

1. Gudjonsson JE, Elder JT. Psoriasis. In: Goldsmith LA, Katz SI, Gilchrist BA, Paller AS, Leffell DJ, Wolff K, editors. Fitzpatrick's dermatology in general medicine. 8th ed. New York: McGraw-Hill; 2012: 197-231.
2. Naldi L, Chatenoud L, Linder D, Belloni FA, Peserico A, Virgili AR, et al. Cigarette smoking, body mass index, and stressful life events as risk factors for psoriasis: results from an Italian case-control study. *J Invest Dermatol* 2005; 125: 61-7.
3. Elston GE, Charles-Holmes R, Carr RA. Precipitation of generalized pustular psoriasis by

- prednisolone. *Clin Exp Dermatol* 2006; 31: 133-4.
4. Thurber M, Feasel A, Stroehlein J, Hymes SR. Pustular psoriasis induced by infliximab. *J Drugs Dermatol* 2004; 3: 439-40.
 5. Gregoriou S, Kazakos C, Christofidou E, Kontochristopoulos G, Vakis G, Rigopoulos D. Pustular psoriasis development after initial ustekinumab administration in chronic plaque psoriasis. *Eur J Dermatol* 2011; 21: 104-5.
 6. Takahashi H, Nakamura K, Kaneko F, Nakagawa H, Iizuka H. Analysis of psoriasis patients registered with the Japanese Society for Psoriasis Research from 2002-2008. *J Dermatol* 2011; 38: 1125-9.
 7. Jiamton S, Suthipinittharm P, Kulthanan K, Chularojanamontri L, Wongpraparut C, Silpa-archa N, et al. Clinical characteristics of Thai patients with psoriasis. *J Med Assoc Thai* 2012; 95: 795-801.
 8. Silpa-archa N, Wongpraparut C. A recalcitrant acrodermatitis continua of Hallopeau successfully treated with etanercept. *J Med Assoc Thai* 2011; 94: 1154-7.
 9. Tay YK, Tham SN. The profile and outcome of pustular psoriasis in Singapore: a report of 28 cases. *Int J Dermatol* 1997; 36: 266-71.
 10. Ohkawara A, Yasuda H, Kobayashi H, Inaba Y, Ogawa H, Hashimoto I, et al. Generalized pustular psoriasis in Japan: two distinct groups formed by differences in symptoms and genetic background. *Acta Derm Venereol* 1996; 76: 68-71.
 11. Jin H, Cho HH, Kim WJ, Mun JH, Song M, Kim HS, et al. Clinical features and course of generalized pustular psoriasis in Korea. *J Dermatol* 2015; 42: 674-8.
 12. Baker H, Ryan TJ. Generalized pustular psoriasis. A clinical and epidemiological study of 104 cases. *Br J Dermatol* 1968; 80: 771-93.
 13. Horreau C, Pouplard C, Brenaut E, Barnette T, Misery L, Cribier B, et al. Cardiovascular morbidity and mortality in psoriasis and psoriatic arthritis: a systematic literature review. *J Eur Acad Dermatol Venereol* 2013; 27 (Suppl 3): 12-29.
 14. Zelickson BD, Muller SA. Generalized pustular psoriasis. A review of 63 cases. *Arch Dermatol* 1991; 127: 1339-45.
 15. Borges-Costa J, Silva R, Goncalves L, Filipe P, Soares dA, Marques GM. Clinical and laboratory features in acute generalized pustular psoriasis: a retrospective study of 34 patients. *Am J Clin Dermatol* 2011; 12: 271-6.
 16. Sugiura K, Takemoto A, Yamaguchi M, Takahashi H, Shoda Y, Mitsuma T, et al. The majority of generalized pustular psoriasis without psoriasis vulgaris is caused by deficiency of interleukin-36 receptor antagonist. *J Invest Dermatol* 2013; 133: 2514-21.
 17. Wolf J, Ferris LK. Anti-IL-36R antibodies, potentially useful for the treatment of psoriasis: a patent evaluation of WO2013074569. *Expert Opin Ther Pat* 2014; 24: 477-9.
 18. Liao PB, Rubinson R, Howard R, Sanchez G, Frieden IJ. Annular pustular psoriasis--most common form of pustular psoriasis in children: report of three cases and review of the literature. *Pediatr Dermatol* 2002; 19: 19-25.
 19. Robinson A, Van Voorhees AS, Hsu S, Korman NJ, Lebwohl MG, Bebo BF, Jr., et al. Treatment of pustular psoriasis: from the Medical Board of the National Psoriasis Foundation. *J Am Acad Dermatol* 2012; 67: 279-88.
 20. Karen JK, Pomeranz MK. Skin change and diseases in pregnancy. In: Goldsmith LA, Katz SI, Gilchrist BA, Paller AS, Leffell DJ, Wolff K, editors. *Fitzpatrick's dermatology in general medicine*. 8th ed. New York: McGraw-Hill; 2012: 1204-12.
 21. Ozawa A, Ohkido M, Haruki Y, Kobayashi H, Ohkawara A, Ohno Y, et al. Treatments of generalized pustular psoriasis: a multicenter study in Japan. *J Dermatol* 1999; 26: 141-9.
 22. Choon SE, Lai NM, Mohammad NA, Nanu NM, Tey KE, Chew SF. Clinical profile, morbidity, and outcome of adult-onset generalized pustular psoriasis: analysis of 102 cases seen in a tertiary hospital in Johor, Malaysia. *Int J Dermatol* 2014; 53: 676-84.

ลักษณะทางคลินิกและผลการรักษาของผู้ป่วยโรคสะเก็ดเงินชนิดคุ่มหนอง 53 ราย ในประเทศไทย

ศุภานัน ประเสริฐโยธิน, สีนภา จุฬาโรจน์มนตรี, ชนิษฐา วงษ์ประภารัตน์, นฤมล ศิลปอาชา

วัตถุประสงค์: เพื่อศึกษาลักษณะทางคลินิก ผลการรักษาและการดำเนินโรคของผู้ป่วยโรคสะเก็ดเงินชนิดคุ่มหนองในประเทศไทย

วัตถุประสงค์และวิธีการ: สํารวจข้อมูลย้อนหลังเกี่ยวกับ ลักษณะทางคลินิก การรักษาที่ได้รับ การตอบสนองต่อการรักษา การดำเนินโรคของผู้ป่วยโรคสะเก็ดเงินชนิดคุ่มหนอง ที่อายุมากกว่าหรือเท่ากับ 15 ปี ซึ่งเข้ารับการรักษาที่หน่วยตรวจโรคผิวหนัง โรงพยาบาลศิริราช ตั้งแต่เดือนกรกฎาคม พ.ศ. 2545 ถึงเดือนตุลาคม พ.ศ. 2557

ผลการศึกษา: ผู้ป่วยโรคสะเก็ดเงินชนิดคุ่มหนอง 53 ราย มีอัตราส่วนผู้หญิงต่อผู้ชาย 1.9:1 มีอายุเฉลี่ย 42.2 ปี (SD = 15.8) และอายุเฉลี่ยขณะเกิดโรคครั้งแรก 36.6 ปี (SD = 17.2) ผู้ป่วย 23 รายมีสะเก็ดเงินชนิดปื้นหนามาก่อนสะเก็ดเงินชนิดคุ่มหนองโดยมีเวลาเฉลี่ยในการเกิดสะเก็ดเงินชนิดคุ่มหนองภายหลังที่ 7.9 ปี ชนิดย่อยของสะเก็ดเงินชนิดคุ่มหนองที่พบมากที่สุดคือ ชนิดเอกแซนทีมาติก (exanthematic, 60.4%) รองลงมาคือชนิดเฉพาะที่ (localized, 22.6%) โดยสะเก็ดเงินคุ่มหนองชนิดเฉพาะที่ประกอบด้วยชนิดย่อย 2 ชนิด คือ อะโครเดอร์มาไคติส คอนทีนัว ออฟ ฮาโลโพร (acrodermatitis continua of Hallopeau, 13%) และสะเก็ดเงินคุ่มหนองที่ฝ่ามือและเท้า (pustulosis palmaris et plantaris, 9%) การพักผ่อนไม่เพียงพอ (30.2%) การติดเชื้อ (30.2%) และยา (28.3%) เป็นปัจจัยกระตุ้นการเกิดโรคที่สำคัญ 3 ลำดับแรกโดยการหยุดยาสเตียรอยด์ชนิดรับประทานเป็นสาเหตุที่สำคัญถึง 60% ในกลุ่มของยาที่กระตุ้นการเกิดโรคสะเก็ดเงินชนิดคุ่มหนองซึ่งเวลาเฉลี่ยในการเกิดคุ่มหนองหลังหยุดยาสเตียรอยด์คือ 1.7 สัปดาห์ ยาสเตียรอยด์ชนิดทายังเป็นยาที่ใช้มากที่สุดในผู้ป่วยโรคสะเก็ดเงินชนิดคุ่มหนอง (98.1%) ส่วนอะซิเตรติน (acitretin) เป็นยารับประทานที่ใช้มากที่สุด (75.5%) รองลงมาคือเมทโทเทรกเซต (methotrexate, 43.4%) ผู้ป่วยสะเก็ดเงินชนิดคุ่มหนองชนิดกระจายทั่วตัว (generalized type) ส่วนใหญ่ตอบสนองอย่างรวดเร็วต่อการรักษาภายในระยะเวลา 2-4 สัปดาห์ ยกเว้นสะเก็ดเงินชนิดคุ่มหนองชนิดเฉพาะที่ (localized type) ซึ่งตอบสนองช้าที่เวลาเฉลี่ย 18 สัปดาห์ภายหลังจากการรักษา 1 เดือน ผู้ป่วยสะเก็ดเงินคุ่มหนองชนิดกระจายหายจากคุ่มหนอง 70% ส่วนสะเก็ดเงินคุ่มหนองชนิดเฉพาะที่หายจากคุ่มหนอง 25% ซึ่งมีความแตกต่างกันอย่างมีนัยสำคัญทางสถิติ อัตราการเกิดโรคซ้ำของทั้ง 2 ชนิดพบได้ 70% ของผู้ป่วยที่ระยะเวลาเฉลี่ย 8 สัปดาห์ภายหลังตอบสนองต่อการรักษา ไม่พบมีรายงานผู้ป่วยเสียชีวิตหรือได้รับผลข้างเคียงที่รุนแรงจากตัวโรคหรือจากการรักษา

สรุป: ผู้ป่วยโรคสะเก็ดเงินชนิดคุ่มหนองพบได้น้อยในประเทศไทย บางรายมีอาการรุนแรง แต่ผู้ป่วยส่วนใหญ่มีการดำเนินโรคที่ไม่รุนแรงและสามารถควบคุมโรคได้หลังได้รับการรักษา อย่างไรก็ตามอัตราการกลับเป็นซ้ำพบได้บ่อย
