

**Table SI. Demographic data of patients with adult-onset immunodeficiency due to anti- interferon- $\gamma$  autoantibodies**

Characteristics	<i>n</i> = 41
Age, years, mean $\pm$ SD, (range)	54.24 $\pm$ 12.68 (25–78)
Total follow-up time, months, mean $\pm$ SD, (range)	42.66 $\pm$ 41.51 (1–180)
Sex, <i>n</i> (%)	
Male	25 (61)
Female	16 (39)
Marital status, <i>n</i> (%)	
Married	32 (89)
Single	4 (11)
Residential area in Thailand, <i>n</i> (%)	
North	5 (12)
Northeast	9 (22)
Central	21 (51)
East	3 (7)
South	3 (7)
Occupation, <i>n</i> (%)	
Government officer	5 (19%)
Business owner	1 (4%)
Employee	6 (23%)
Merchant	1 (4%)
Farmer	6 (23%)
Others	7 (27%)
Underlying diseases, <i>n</i> (%)	
Hypertension	4 (10)
Dyslipidemia	2 (5)
Diabetes mellitus	1 (2)
Gouty arthritis	1 (2)
Stroke	1 (2)
Chronic hepatitis B virus infection	1 (2)
Hemoglobin E trait	1 (2)
Hyperthyroid	1 (2)
Poliomyelitis	1 (2)
Overlapping SLE and RA	1 (2)
Rheumatoid arthritis	1 (2)
No	28 (68)
Outcome, <i>n</i> (%)	
Alive	38 (93)
Dead	3 (7)

Some patients have more than one organ involvement. Patients were suspected of having autoantibodies based on the presence of at least two episodes of infections caused by uncommon intracellular organisms including NTM, disseminated fungal infections (e.g., infection due to *Talaromyces marneffe*i, cryptococcosis, histoplasmosis), non-typhoidal Salmonella bacteremia, disseminated herpes zoster infection with a negative test for anti-HIV antibody.

**Table SII. Comparison of laboratory data between patients with and without reactive skin lesions in patients with adult-onset immunodeficiency due to anti-interferon- $\gamma$  autoantibodies**

		<i>n</i>	Mean $\pm$ SD	<i>p</i>
Hemoglobin (g/dl)	Without RS	141	11.29 $\pm$ 1.94	0.751
	With RS	23	10.04 $\pm$ 2.07	
White blood cell count (*10 <sup>3</sup> /ul)	Without RS	140	12.47 $\pm$ 7.67	<0.001
	With RS	23	25.09 $\pm$ 13.67	
Absolute neutrophil count (*10 <sup>3</sup> /ul)	Without RS	141	8.65 $\pm$ 7.06	<0.001
	With RS	23	20.03 $\pm$ 12.55	
Absolute lymphocyte count (*10 <sup>3</sup> /ul)	Without RS	141	2.62 $\pm$ 1.25	0.730
	With RS	23	2.97 $\pm$ 1.23	
Absolute eosinophil count (*10 <sup>3</sup> /ul)	Without RS	141	0.64 $\pm$ 0.69	0.004
	With RS	23	1.01 $\pm$ 1.04	
Platelet count (*10 <sup>3</sup> /ul)	Without RS	186	349.04 $\pm$ 150.07	0.989
	With RS	34	431.94 $\pm$ 134.20	

Statistical differences between the two groups were analyzed by paired *t*-test and was completed by GraphPad Prism 6 software (GraphPad Software Inc., La Jolla, CA).  
RS: reactive skin diseases.

**Table SIII. Clinical features, histopathology and laboratory data of non-neutrophilic dermatoses in patients with adult-onset immunodeficiency due to anti-IFN  $\gamma$  autoantibodies**

Skin conditions	No. of episodes	Clinical features	Histopathology
Leukocytoclastic vasculitis	1	Multiple hemorrhagic tense bullae on erythematous base with pustules at periphery on both hands	Fibrinoid necrosis of blood vessel walls and extravasated red blood cells with neutrophils and nuclear dusts
Erythema nodosum	6	Subcutaneous nodules on the shins and posterior legs	Septal panniculitis with inflammatory cells infiltration predominantly composed of neutrophils
Diffuse plane xanthoma	1	Multiple, soft, well-defined yellow-orange plaques on medial upper and lower eyelids, anterior neck and upper chest with telangiectasia	Dense aggregates of foamy histiocytes in upper dermis. AFB and mAFB: negative
		Note: Serum protein electrophoresis: Albumin 2.10 g/dl (3.5–5.0), Alpha-1 0.34 g/dl (0.1–0.3), Alpha-2 1.00 g/dl (0.6–1.0), Beta-1 0.58 g/dl (0.7–1.0), Beta-2 0.74 g/dl (0.3–0.6), Gamma 4.03 g/dl (0.7–1.6), Total protein 8.80 g/dl (6.4–8.3) Immunofixation: Polyclonal gammopathy Free Kappa light chain 256 mg/l (3.30–19.4), Free Lambda light chain 64.60 mg/l (5.71–26.30) K/L ratio 3.963 (0.26–1.65)	
Papulovesicular eruption	1	Generalized papulovesicles on trunk and extremities	Superficial perivascular infiltration with eosinophils
Pancreatic panniculitis	1	Multiple erythematous subcutaneous nodules on abdomen, back and all extremities	Necrotic fat cells (ghost adipocytes) with neutrophilic infiltrate and calcification in subcutaneous fat tissue AFB, mAFB, GMS, PAS: negative
Suppurative granulomatous dermatitis	3	Ulcerated nodules on the buttocks	Epidermal hyperplasia with collection of neutrophils, lymphocytes, histiocytes and multinucleated giant cells AFB, mAFB, GMS, PAS: negative cultures: neg
		Hyperpigmented fluctuated mass on left arm	Large aggregation of neutrophils and lymphocytes, histiocytes and multinucleated giant cells AFB, mAFB, GMS PAS: negative
		Multiple infiltrative erythematous papules and nodules on the face and trunk	Diffuse suppurative granulomatous infiltration composed of plasma cells, histiocytes, eosinophils neutrophils lymphocytes and giant cell AFB and PAS: negative
Lichen amyloidosis	1	Hyperpigmented keratotic papules in ripple pattern on arms, legs and back	Not done
Seborrheic dermatitis	1	Hypopigmented patches with scales on scalp and nasolabial folds	Not done
Other: non-specific dermatosis, clinical suspicious of having cutaneous vasculitis	1	Erythematous papules on the face and trunk	Non-specific chronic inflammation, no evidence of vasculitis

AFB: Acid-Fast Bacillus staining; mAFB: Modified-acid fast staining; GMS: Gomori Methenamine-Silver Nitrate Stain; PAS: Periodic acid-Schiff stain.

**Table SIV. Episodes of neutrophilic dermatoses (ND) that coincide with the infections. Types of ND, sites and species of the infections are shown**

Neutrophilic dermatoses	Sites and species of the concurrent infections			
	Blood stream	Lymph nodes	Bone	Lung
1 Sweet syndrome	<i>Mycobacterium abscessus</i>			
2 Sweet syndrome		<i>Mycobacterium chelonae</i>		
3 Sweet syndrome		<i>Mycobacterium abscessus</i>		
4 Sweet syndrome	<i>Mycobacterium non-photochromogen</i>			
5 Sweet syndrome	<i>Salmonella</i> group B	<i>Mycobacterium kansasii</i>	<i>Mycobacterium kansasii</i>	
6 Sweet syndrome		<i>Mycobacterium abscessus</i>		
7 Sweet syndrome	<i>Salmonella choleraesuis</i>			
8 Generalized pustular eruption	<i>Salmonella</i> group D, <i>Mycobacterium abscessus</i>			
9 Generalized pustular eruption	<i>Mycobacterium kansasii</i>			
10 Generalized pustular eruption		<i>Mycobacterium abscessus</i>		
11 Generalized pustular eruption		<i>Mycobacterium fortuitum</i> , <i>Mycobacterium chelonae</i> complex		
12 Generalized pustular eruption				<i>Mycobacterium tuberculosis</i>
13 Generalized pustular eruption	<i>Mycobacterium abscessus</i>			
14 Neutrophilic reactions NOS*		<i>Mycobacterium abscessus</i>		
15 Neutrophilic reactions NOS*		<i>Mycobacterium abscessus</i>		
16 Lobular panniculitis		<i>Mycobacterium abscessus</i>		

Neutrophilic reactions NOS\*, the condition was diagnosed when erythematous papules, nodules or pustules were detected, and histopathology revealed neutrophilic infiltrations in whom the criteria for diagnosis of Sweet syndrome were not met.