Case 2

A 15-year-old Thai boy

Chief Complaint: Generalized erythematous rash for 3 months.

Present Illness: The patient developed dry erythematous rash since he was 2 years old and was diagnosed as Atopic dermatitis. His skin condition was well controlled with topical and oral medication. In the past 3 month, he developed multiple scaling erythematous itchy plaques at trunk and extremities which was not improved by the previous modalities. He came to Ramathibodi hospital on 9th OCT 2009. The skin biopsy was done on 15th OCT 2009. The pathological diagnosis was compatible with Psoriasis. During follow up we noticed that he always has persistent upper respiratory tract infections. Thus,we consulted Pediatric allergy clinic to work up for this condition.

Past History: He is the first child, term born with cesarean section. Birth weight 3,000 gm. He had history of recurrence otitis media since he was 2 year-old.

Family History: All family members are healthy.

Physical examination

BW 43 kg, Height 153 cm.

HEENT: slightly pale nasal mucosa, no lymphadenopathy

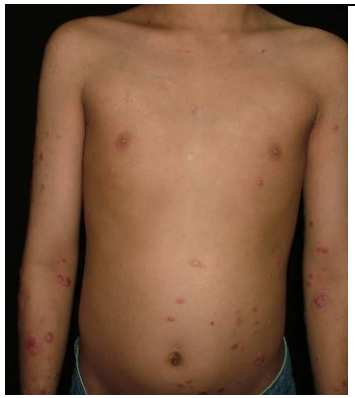
Heart: normal s1 s2, no murmur

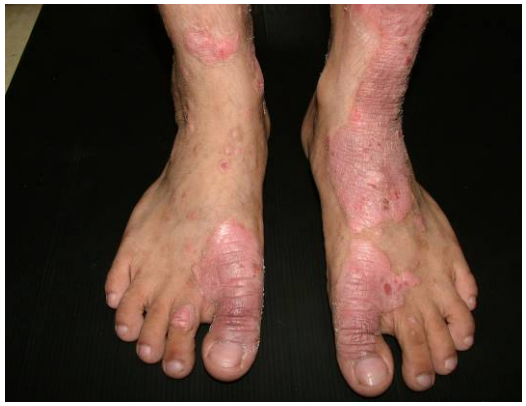
Lung: course crepitation at right lower lung

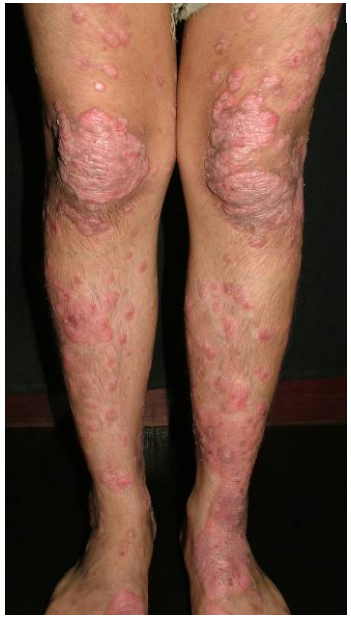
Abdomen : Liver and spleen not palpable

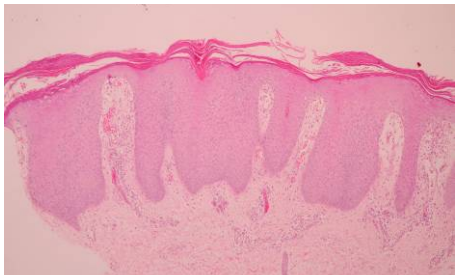
Dermatological Examination:

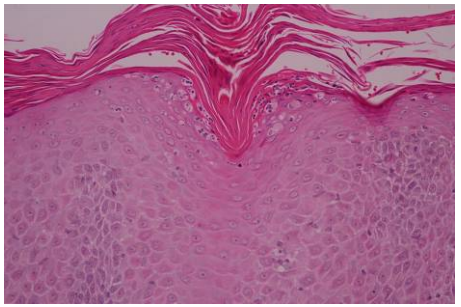
Generalized discrete well- defined erythematous plaques with silvery scale at face, trunk, genitalia and extremities.











Laboratory Investigation

IgG 571 mg/ml (822-1070) ↓

IgM 6.94 mg/ml (39-79) ↓

IgA 131 mg/ml (85-211) ↔

CD19 - 0 ↓

Chest X-ray: RML bronchiectasis

Histopathology (S09-16200)

Mounds of parakeratosis with neutrophils, spongioform pustules, hypogranulosis and psoriasiform hyperplasia in the epidermis. superficial perivascular inflammatory-cell infiltrate of lymphocytes with dilated, tortous blood vessels in the papillary dermis.

Diagnosis: Psoriasis with Common Variable Immunodeficiency (CVID)

Treatment: Psoriasis: NBUVB then MTX(2.5) 2 tab/wk (Dec 2009 – Feb 2011) with topical treatment and Acitretin(10) 1 tab (Mar 2010 – Sep 2010)

CVID: IVIG 300 – 400 mg/kg/dose every 4 weeks. (Dec 2010 until present)

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Discussion:

Common variable immunodeficiency (CVID) is a primary immunodeficiency which is defected in quantitative immunoglobulin levels. The prevalence of this disease is estimated at 1:25,000 with male and female affected equally.[1] Most cases are sporadic. CVID mostly present in young adult characterized by chronic recurrence of sinopulmonary tract infections, malignancies and granulomatous inflammation.[1-3] Laboratory investigations will show low level of IgG, IgA and/or IgM.[4-6] Periodicaly Intravenous immunoglobulin administration is a standard treatment. Dermatologic finding which were reported associated with this condition were alopecia areata, vitiligo, cutaneous granuloma and atopic dermatitis.[7-9] There was no reported case of psoriasis with CVID.

In this case, clinical and laboratory finding are compatible with Common variable immunodeficiency and X-linked agammaglobulinemia (XLA). To get the definite diagnosis, genetics work up should be done. (Table 1)

Table 1 shows clinical and laboratory features of XLA, CVID, and our patient.[10]

	XLA	CVID	Patient
Immunoglobulin level	all subtype markly decrease	IgG +/- IgA, IgM, just below normal	IgG and IgM
CD19 (B cell)	Absent	low	absent
Family history	+	- or +	-
Gene defect	BTK gene	ICOS, BAFFR, CD-19: AR <1% TACI-AR,AD:(10-15%) Unknown:75-80%	pending
Skin manifestation	Unusual	Granulomatous, AA, vitiligo, AD	AD, Psoriasis

Gurmin et al. reported three patients with psoriasis and psoriatic arthritis who improved after IVIG therapy for other underlying diseases.[11] The same as Yoshinaru et al reported that psoriasis skin lesions of their chronic inflammatory demyelinating polyneuropathy (CIDP) patient also improved dramatically with IVIG and disease free for 6 months.[12] Since Psoriasis was diagnosed, he was treated with topical medications, phototherapy and also systemic drug. These modalities improved psoriasis lesions gradually but not clear. Two months after first injection of IVIG, all the skin lesions subsided and gradually clear. Several mechanisms of action have been proposed to explained immunomodulatory effects of IVIG in psoriasis. [6, 11-13]

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