Case 3

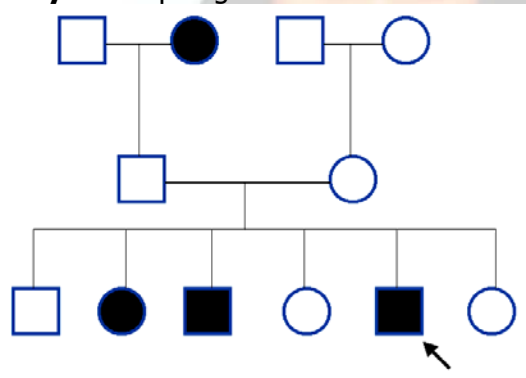
A 36 year-old man from chachoengsao

Chief complaint:

Present illness 4 months PTA, he worked in a prawn farm, had chronic contact with chemicals (formaline, chrlorine and water) Since then, he became itchy and developed rash on trunk and extremities, gradually progressed to generalized scaly, peeling, erythematous rash all over the body

Past history He had icthyotic skin and red skin since birth

Family history The pedigree was shown as below



Physical examination Afebrile man

HEENT Extropion of both eyes. Swelling of the lips.

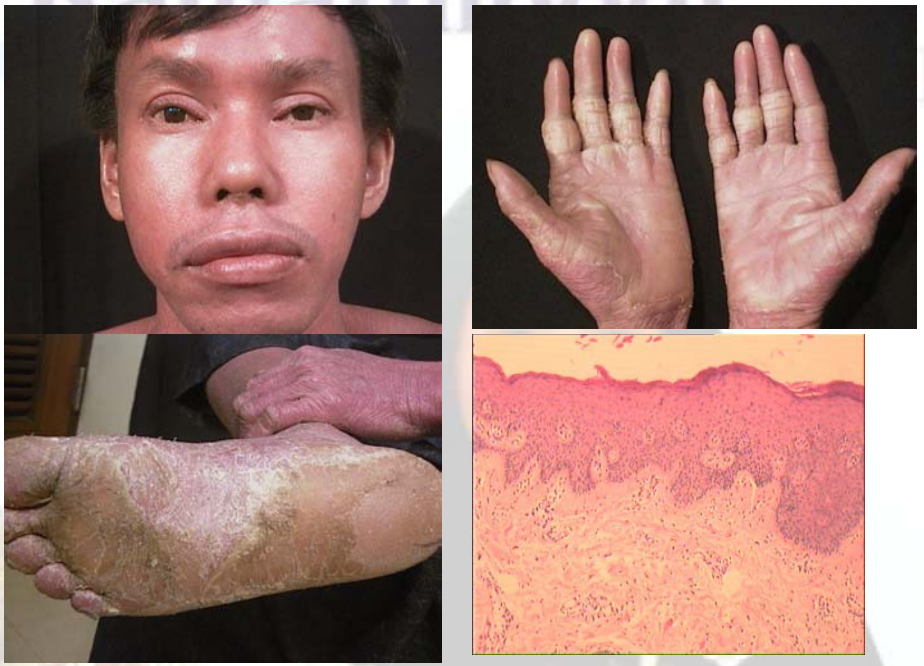
Abdomen Liver and spleen not palpable

Skin exam generalized erythroderma and palmoplantar keratoderma.

Histopathology: S01-6990

There is confluent of parakeratosis with occasionally neutrophils and psoriasiform epidermal hyperplasia in association with a sparse superficial lymphocytic infiltrate

Diagnosis: Congenital, non-bullous, ichthyosiform erythroderma (CIE)



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Comment:

Congenital, non-bullous, ichthyosiform erythroderma is an inheritated disorder, usually transmited by autosomal recessive (rarely AD or sporadic). Clinical manifestation of generalized erythroderma and fine scale, with little or no ectropion, eclabium or alopecia. It was thought to be mild form of lamellar ichthyosis. The symptoms improved with age, but often deteriorated in summertime, hypohidrosis due to sweat duct obstruction by the hyperkeratotic stratum cornium may occur.

Reference

1. Kanitakis J, Hoyo E, Chouvet B, Thivolet J. eratinocyte proliferation in epidermal keratinocyte disorders evaluated

through PCNA/cyclin immunolebelling ang Ag NOR counting. *Acta Derm Venereol. 1993 Oct;73(5)370-5*

2. Coenraads PJ. Congenital non-bulluos ichthyosiform erythroderma. *Br J Dermatol. 1980 Apr; 102(4): 482-3.*
3. Peery TB, Holbrook KA, Hoff MS. Prenatal diagnosis of congenital–non-bullous ichthyosiform erythroderma (lamella ichthyosis). *Prenat Diagn. 1987 Mar;7(3): 145-55*