

Case 10

A 10-year-old Thai boy

Chief complaint: Fine white scaly erythematous skin on the face, scalp, trunk and extremities since birth.

Past history He was preterm baby with 1800 gm birth weight

Family history He is the only child in his family

No history of the congenital skin disorder in his families

Physical examination A young boy with normal growth and development

Skin examination revealed erythematous fine white scaly erythematous skin distributed over scalp, face, trunk and extremities. He had mild ectropion.



Fig 1



Fig 2

Diagnosis: Congenital ichthyosiform erythroderma

Presenter : Pitchaya Somburanasin

Consultant : Somyot Charuwichitratana

Discussion

Congenital ichthyosiform erythroderma is one form of severe congenital ichthyosis. Most of the patients inherit the disease in an autosomal recessive pattern. This form of ichthyosis has markedly increased epidermal turnover rate.

The newborn usually presents with a constricted collodion membrane. After shedding of the membrane underlying erythema with a fine white generalized scale are apparent. The classic CIE has little or no ectropion, eclabium or alopecia, palms and soles involvement is variable. The patients may have minimal sweating with severe heat intolerance.

Histopathologic examination shows hyperkeratosis with acanthosis but not specific for this disease.

The aim of therapy is symptomatic, correct the epidermal function and prevent complications, by focus on hydration, lubrication and keratolysis. Topical treatment with retinoids and vitamin D derivatives may be effective but can be irritating in some patients. Systemic retinoids therapy can induce dramatic improvement in many cases but the decision to initiate systemic retinoids should be weighed carefully.

References

1. John J. DiGiovanna. Ichthyosiform dermatitis. Fitzpatrick, Dermatology in general medicine, vol. 1. 5th ed. New York McGraw-Hill 1999: 581-601
2. Hofmann B, Stege H, Ruzicka T, Lehmann P. Effect of topical tazarotene in the treatment of congenital ichthyoses. Br J Dermatol 1999 Oct;141(4):642-6
3. Akiyama M. The pathogenesis of severe congenital ichthyosis of the neonate. J Dermatol Sci 1999 Sep;21(2):96-104

4. Luckner GP, van de Kerkhof PC, van Dijk MR, Steijlen PM. Effect of topical calcipotriol on congenital ichthyoses. *Br J Dermatol* 1994 Oct;131(4):546-50
5. Saracoglu ZN, Tekin N, Urer SM, Sabuncu I, Aksit A. Oral acitretin treatment in severe congenital ichthyosis of the neonate. *Turk J Pediatr* 2002 Jan-Mar;44(1):61-4

Hospital

August 16, 2002



Interhospital Dermatology Conference