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Congentinal Bullous Ichthyosiform Erythroderma

IBRAHIM GALADARI, MD AND SUHAIL HADI, M.Phil.

From the Department of Dermatology, Al Ain Hospital, UAE

ABSTRACT-A case of Bullous ichthyosiform erythroderma is reported. This is a rare disease with autosomal dominant inheritance. It is characterized by blister formation, scaliness & erytherma starting after birth, leaving flexural hyperkeratotic areas with rippled appearance later in life, and this is usually persistent.

Case Report:

A 14 yrs old Pakistani boy presented with a history of fluid-filled lesions on joint surfaces and flexures started since birth which by time ended with thickened and pigmented lesions on those sites.

The second son of the family died at the age of two years (unknown cause).

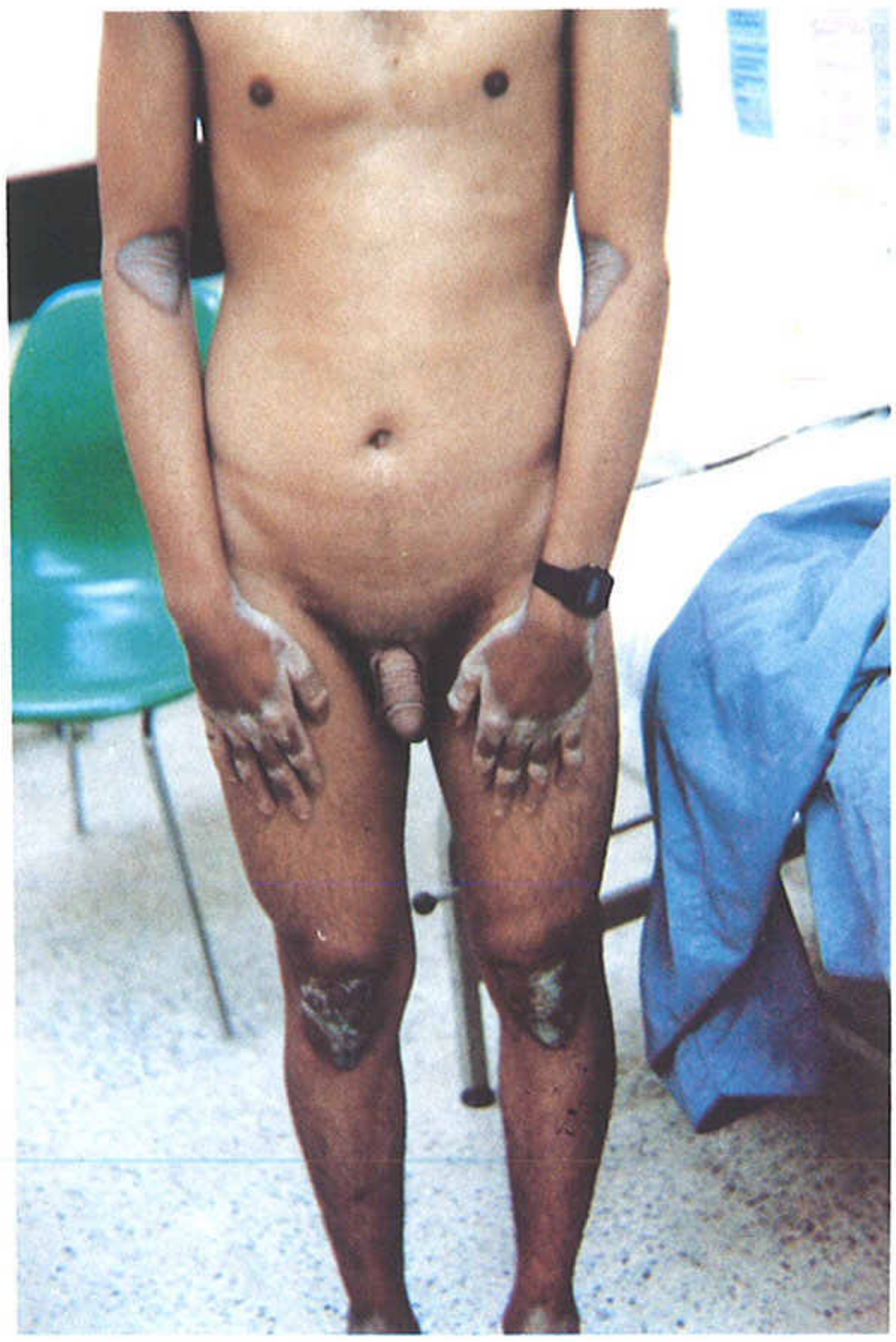
On examination: there were hyperkeratotic, hyperpigmented, well demarcated skin lesions over all the big joints and flexures. The skin over those sites was acanthotic with warty-like projections. The rest of skin showed roughness and dryness. Hairnails and Mucous membranes are free.

Generally the child looks healthy, active and well oriented.

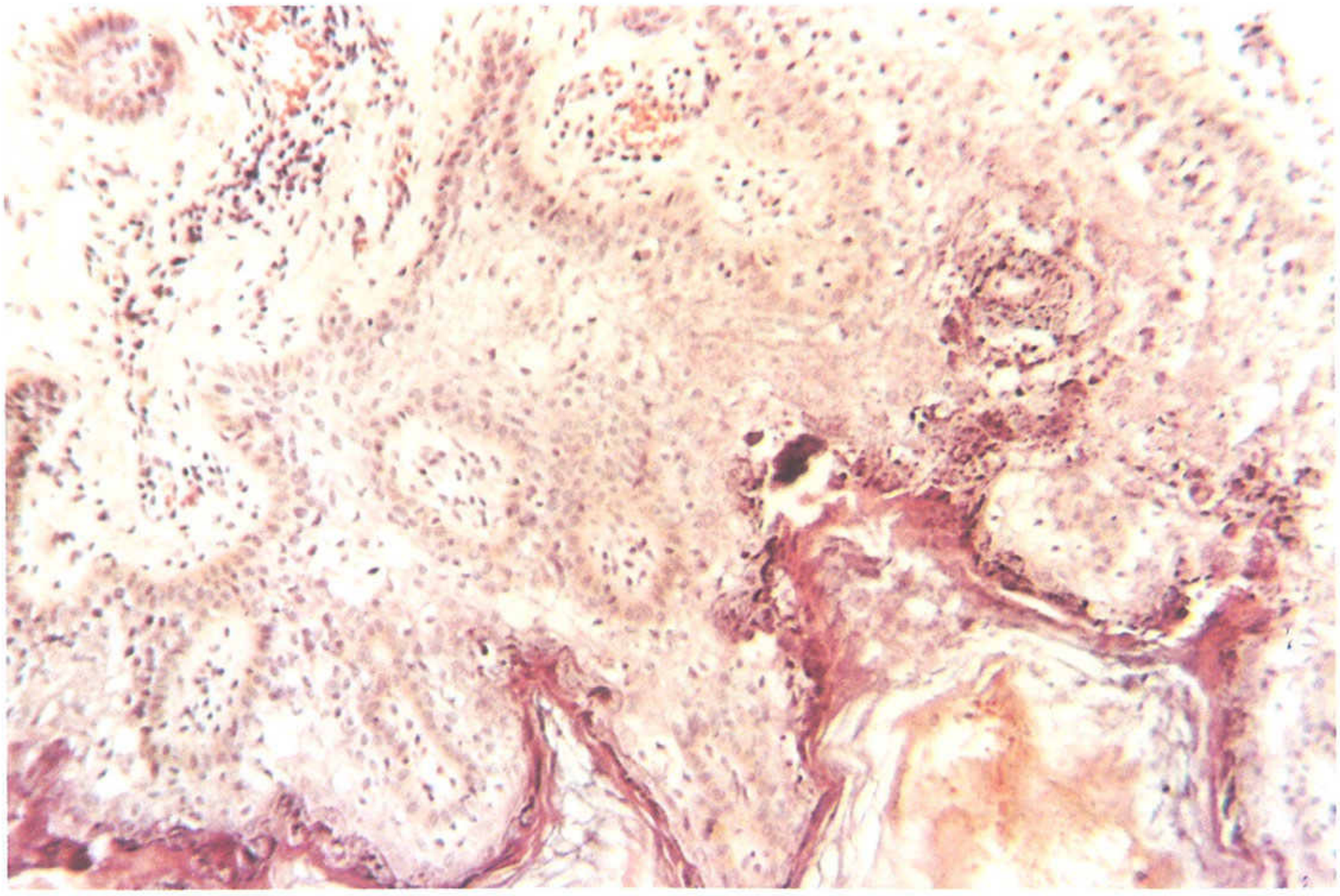
Systemic examination: not significant.

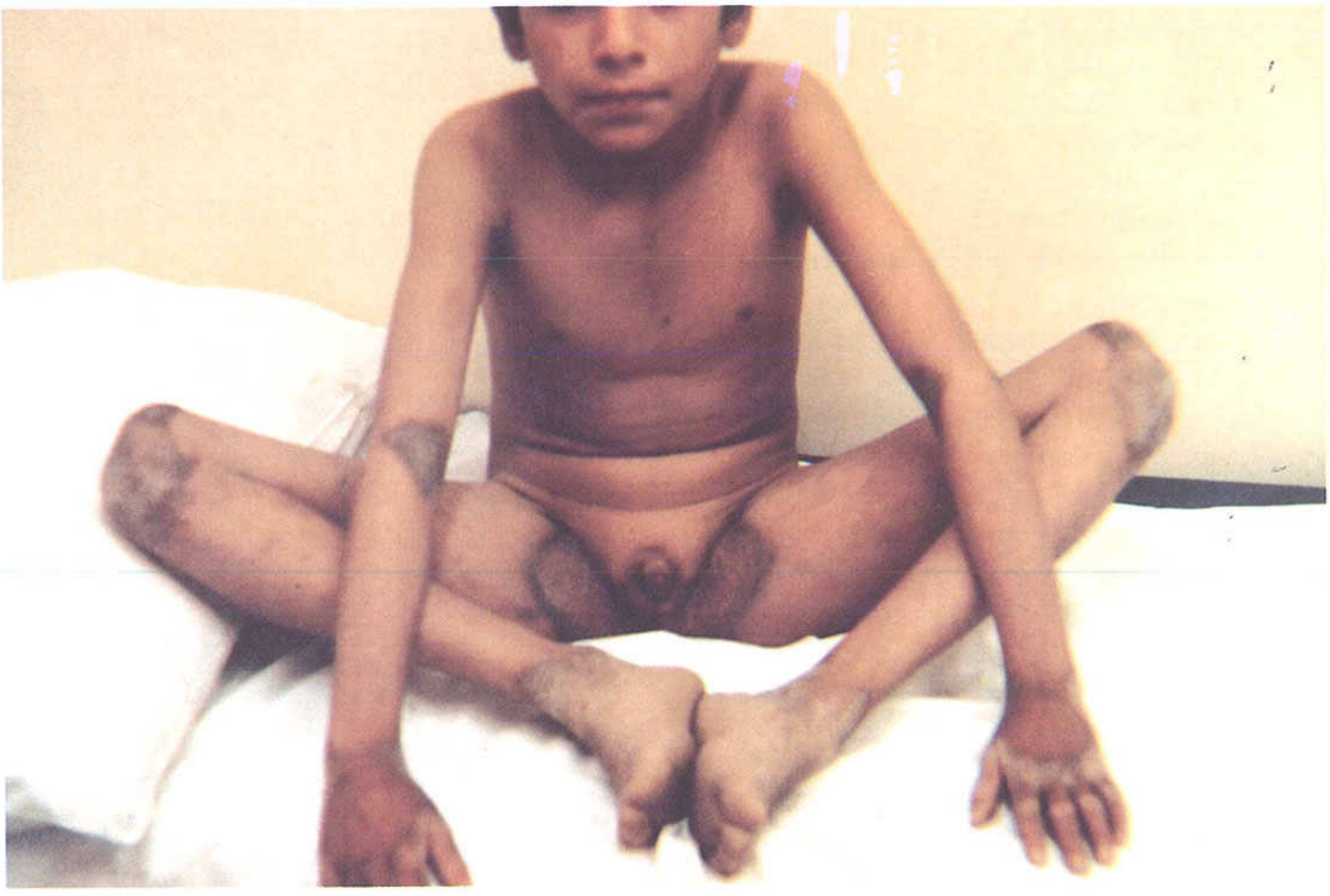
Complete blood picture, ESR: was within normal limits. Liver function tests, cholesterol, Triglycerides, showed elevated AP and TG during tigason therapy which returned to normal after stopping Tigason.

Skin biopsy showed irregular papillomatosis and hyperkeratosis, with perinuclear vacuolization and execessive formation of large irregularly shaped Keratohyaline granules.



The patient improved with keratolytics, emolients hydrating agents and tigason (lmg/kg/day) as starting dose which was reduced to 0.5 mg/kg/day after 6 weeks, and then stopped (by the patient) within few months.





Now the patient was left with hyperkeratotic lesions on the same sites.

Comment:

Bullous Ichthyosiform erythroderma or epidermolytic hyperkeratosis is a rare condition (incidence 1:300,000) which becomes manifest after birth (as in our case). Gene mutation might explain the appearance of sporadic cases(1). Blisters usually appear at sites of trauma with frequent pyogenic infections. Etythema and blistering usually subside with time to leave hyperkeratotic lesions with brownish, verruciform thick scales (rippled appearance) involving mainly the flexures and intertriginous regions(2) these lesions are usually persistent as we saw in our patient where lesions were mainly on the knees, elbows and ankles. Nail, hair sweat glands and mucous membranes are not involved in this condition (as our case), but carcinomata might develop occasionally(3).

Histologically; the changes seen in our case are in agreement with those seen in classical cases namely: Thickened epidermis with vacuolization around the nuclei in the upper stratum spinosum and in the granular layer and formation of abnormal keratolyaline granules(4).

The same pathological changes are seen in systematized linear epidermal nevus so these two conditions might be considered as different expression of a pleiotropic dominant gene. The disease usually responds to treatment with etretinate(5), our case showed moderate response to tigason therapy.

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