

The Dry Baby - (Non-Bullous Congenital Ichthyosiform Erythroderma) - A Case Report

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1. Introduction

The ichthyosis are typically inherited conditions exhibiting disordered keratinization secondary to excessive transepidermal water loss.¹ Non-bullous congenital ichthyosiform (NBCIE) erythroderma is a rare chronic autosomal recessive form of inherited ichthyosis. Clinically NBCIE appears as generalised erythroderma with fine white scales that gradually replace the collodion membrane. NBCIE is estimated to occur in 1:300,000 newborns of all races, more frequently seen in consanguinity.^{2,3} The genes involved in mutation are TGM1, ALOX12B, ALOXE3 and ichthyin. Here we report one such rare case of NBCIE in a 10 month old child with consistent clinical and skin biopsy findings.

2. Case Report

A 10-month old female child, born of second degree consanguineous marriage, was brought to a tertiary care hospital with complaints of dryness, peeling of the skin, fissuring all over the body since 9 months which was aggravated past 1 week. It was associated with redness, itching and low-grade fever. Earlier the child had redness confined to her neck region alone, which eventually spread to involve the whole body and was associated with scales. On enquiry parents also informed that the baby was born enclosed in a constricting parchment like shiny membrane (collodion baby) that had gradually shed off leaving behind diffuse erythema, dryness and scaling.

General physical examination on first day of admission showed body temperature of 98F, heart rate 142/min, respiratory rate 44/min, peripheral pulses were well felt in all the four limbs with mild pallor.

Dermatological examination revealed diffuse erythema with fine white scaling involving almost the whole body; scales being larger on the legs and finer at other places. There were no bullae, vesicles, ectropion, eclabium, blister, or cicatricial alopecia. Scalp showed few large thick scales. The mucous membranes were spared.

Complete hemogram revealed haemoglobin of 10.2g/dl, white blood count $18 \times 10^3/\mu\text{l}$, platelet count $350 \times 10^3/\mu\text{L}$. Thyroid and lipid profiles were within normal limits. Liver function tests, renal function tests and urine routine were normal. Albumin was 3.1g/dl.

Histological examination showed epidermis displaying parakeratosis and pseudoepitheliomatous hyperplasia. The parakeratotic corneocytes had enlarged nuclei. The

superficial dermis revealed mild edema and telangectasia. A diagnosis of NBCIE was made.

Symptomatic management in the form of aseptic handling, hydration of the skin with lubricating creams and application of non-occlusive emollients have reduced the scaling and pruritis. Child is now clinically better and is on regular follow up.

3. Discussion & Conclusion

The diagnosis of NBCIE is established from history, clinical features and histological examination. The close differential diagnosis is lamellar ichthyosis where there are large dark scales compared to the fine white scales of NBCIE.⁴ Collodion babies should be cared for in the neonatal intensive care unit due to increased risk of mortality. Impairment of temperature regulation, increase insensible water loss, hypernatremic dehydration, sepsis, acute renal failure are possible complications.

Ichthyosis management requires a multimodal approach, including topical and oral agents in addition to lifestyle modifications. Treatment should incorporate hydration and lubrication with the addition of keratolytics depending on scale severity.⁵ Hydration can be accomplished with lubricating creams and ointments which act by increasing the water-binding capacity of the stratum corneum. In severe cases topical keratolytics cause desquamation while retinoids and calcipotriol limit epidermal proliferation.⁵ Low dose oral retinoids are a mainstay in the systemic management of severe disease.⁵

Life style modifications like daily bathing with mild cleanser, application of plain emollients frequently, help to seal in moisture, keep the skin hydrated, improving the overall skin appearance.^{6,7} Genetic counselling, an integral aspect of disease management assists parents in consideration of future pregnancies. Careful monitoring and management of these patients may improve the outcome and quality of life in children suffering physical discomfort and social stigmata due to their condition.



Figure 1



Figure 2:



Figure 3

Figure 1, 2, 3: Clinical photographs

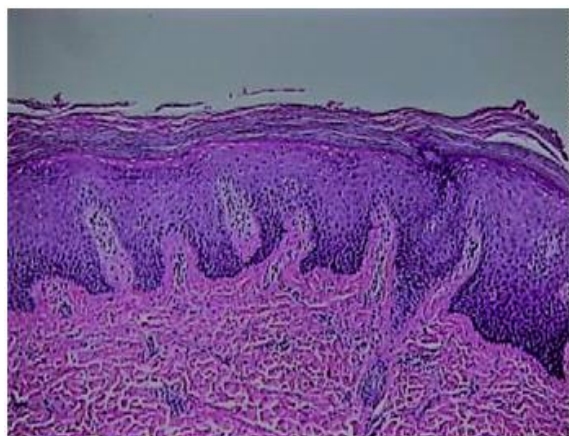


Figure 4: Histology of NBCIE

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