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Journal of Skin and Sexually Transmitted Diseases

Case Report

Management of a collodion baby – Our experience

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Received : 12 May 19
Accepted : 25 May 19
Published : 02 December 19

DOI
10.25259/JSSTD_31_2019

Quick Response Code:

ABSTRACT

Collodion baby is a transient condition in newborns where they are covered in a taut, shiny membrane. Later, it can evolve into any form of autosomal recessive congenital ichthyosis or other ichthyosiform syndromes. Retinoids are one of the most effective therapeutic modalities for ichthyosis and have been found to be safe in neonates. Hence, early and judicious use of retinoids can significantly improve the quality of life in severe ichthyosis. Herein, we report a case of congenital ichthyosis which showed an excellent response to acitretin therapy.

Keywords: Ichthyosis, Collodion baby, Oral retinoid, 3 months on and off therapy

INTRODUCTION

Ichthyosis is a heterogeneous group of keratinization disorder with congenital and acquired forms, characterized by dryness and scaling of the skin. Abnormalities in the structure of stratum corneum may lead to increased transepidermal water loss resulting in excessive dryness.[1] Collodion baby is a phenotype common to several disorders where the skin is dry at birth and is similar to parchment. Around 60%–80% of collodion babies eventually develop into non - bullous congenital ichthyosiform erythroderma (NBCIE) or lamellar ichthyosis and around 10%–20% evolve into normal skin as self-healing collodion baby or other ichthyosiform syndromes.[2] The primary treatment consists of moisturizing the skin, reducing fluid loss, and preventing infection. Successful treatment with oral retinoids has been reported.[3] Herein, we report a case of NBCIE which showed an excellent response to acitretin therapy.

CASE PRESENTATION

A new born female was referred to our hospital on the 4th post-natal day due to abnormal physical appearance. She was the third child of a consanguineous (second degree) marriage, delivered by lower segment cesarean section at term. On examination, the baby was encased in a collodion membrane [Figure 1]. There were ectropion, eclabium, restricted limb movements, and suckling difficulty. Her siblings were normal at birth. She was admitted in the neonatal intensive care unit (NICU) and kept under reverse barrier nursing. Parents were reluctant to nurse the baby and abandoned the child subsequently. Through the legal action, the parents were coerced to come back. Routine investigations were within normal limits except for hypernatremia which was corrected by giving calculated amount of water through the nasogastric tube over a period of 48 h. Expressed breast milk was also given. In the

incubator she was laid on a plastic sheet instead of cotton clothes, in order to avoid peeling of skin. Since the NICU is air conditioned perspiration will be minimal and the plastic sheet won't cause any discomfort to baby. Also, skin was protected with liberal application of emollients and topical antibiotics. Special care was given to eyes and other mucosae. Parenteral antibiotics were given to prevent any secondary infection. Fluid correction was done according to daily weight and electrolyte requirement.

After obtaining written informed consent from the parents, she was started on capsule acitretin (1 mg/kg/day). The capsule was refrigerated and one-third was administered after dissolving it in breast milk. Treatment plan was 3 months on and 3 months off therapy. At 1 month of therapy, clinical improvement was observed and at 3 months, the child became near normal including amelioration of ectropion and eclabium [Figure 2]. The child showed an excellent response to acitretin and tolerated therapy well. She was off treatment for the next 3 months and at the end of 6 months, she developed scaling and erythema over the face and extremities suggestive of NBCIE [Figure 3]. At the end of 9 months of treatment with acitretin, there is marked resolution of dryness of the skin [Figure 4]. Specific management with retinoid has boosted the quality of life of the child and her family. Hence, we plan to pursue the same.

DISCUSSION

The term collodion baby was given by Hallopeau and Watelet.[4,5] It is an extremely rare dermatological emergency with an estimated incidence of 1 in 50,000 to 1 in 100,000 birth.[6] The collodion baby is described as a congenital

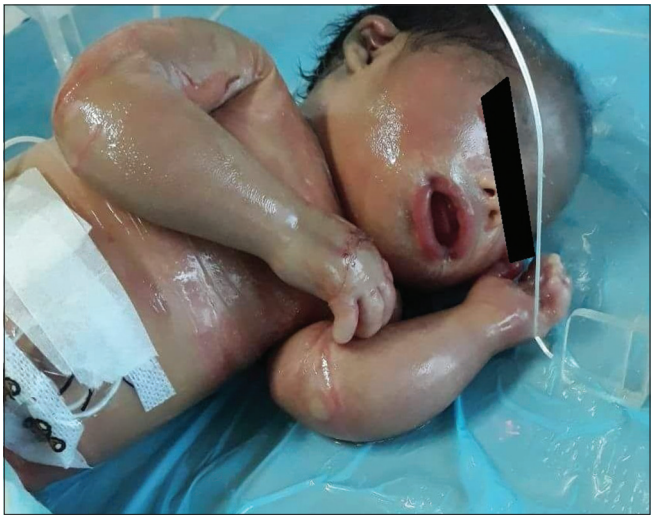

Figure 1: Baby encased in a collodion membrane at birth.

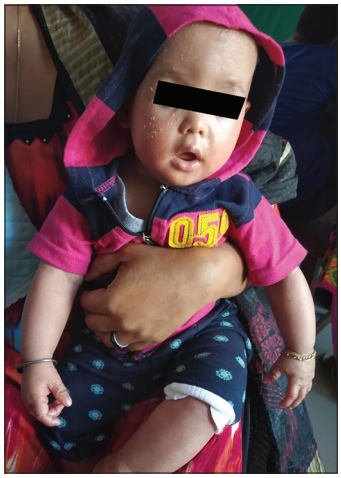

Figure 3: Mild erythema and scaling of extremities at the end of "off period" of acitretin therapy.

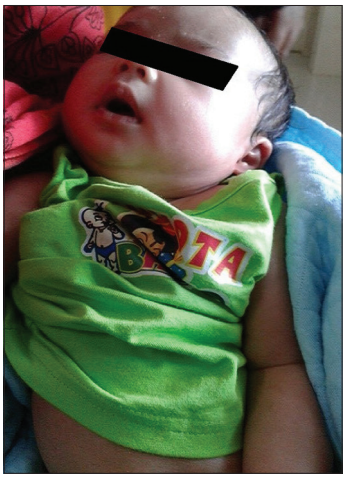

Figure 2: Near normal looking baby after 3 months of treatment with acitretin.

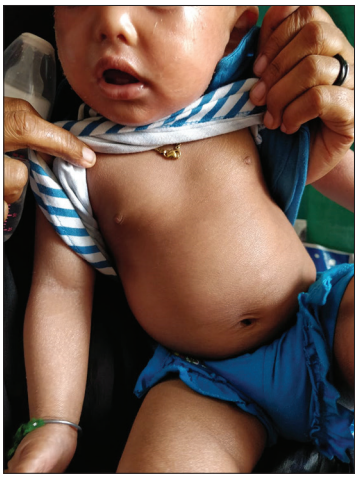

Figure 4: Marked resolution of dryness of skin at the end of 9 months of treatment with acitretin.

condition characterized by the presence of parchment-like or cellophane membrane covering the whole body which cracks and peels off within 2–4 weeks.[7] Due to thickened skin structure and pulling of soft tissues around the lips and conjunctivae, ectropion and eclabium develop. Loss of skin integrity may cause hypothermia, increased insensible water loss and electrolyte disorders, skin infections, and sepsis.[8] Management of this devastating condition requires multidisciplinary approach with the combined effort of dermatologist, neonatologist, and ophthalmologist. Approximately 75% of collodion baby develop an autosomal recessive (AR) congenital ichthyosis as in our case. NBCIE is an AR disorder characterized by erythema and scaling all over the body and is due to mutation in genes such as ALOX12B, ALOXE3, TGM1, ABCA12, and NIPAL4.[9] In this scenario, AR inheritance may be due to parental consanguinity. Acitretin is a second-generation retinoid which controls proliferation and modifies differentiation of keratinocytes. It can cause serious adverse effects such as premature closure of epiphysis, metabolic side effects, xerosis of skin, and mucosae. Because of the side effect profile, dermatologists are reluctant to give retinoids. The risk and benefits of giving retinoids should be assessed in each scenario, and careful monitoring during the course of treatment brings about a better outcome. Previous reports from Turkey also support the same.[8] Because of the potential toxicity of the drug and chronicity of the disease, intermittent therapy is preferred.[10] Treatment is preferably administered as on and off therapy helping the clearance of the drug in the interim period. In the background of our case, we recommend early use of retinoids in collodion babies as it is a good armamentarium which can bring a dramatic change in the life of these babies.

CONCLUSION

Improvement in the physical appearance of the baby brought tremendous change in the attitude of parents who are now caring the baby very well. The timely and judicious use of acitretin may help the collodion babies to have a near normal appearance.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Anjana S, Sobhanakumari K, Mathew R, Mathew R. Management of a collodion baby – Our experience. J Skin Sex Transm Dis 2019;1(2):101-3.