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Lamellar ichthyosis (Collodion Baby) - A Rare Case

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Abstract

A "Collodion baby" is a relatively rare and transient condition in which a newborn is encased in a yellow, thick, shiny, tight, parchment-like membrane; called a 'collodion membrane'. This condition usually precedes the development of one of the various types of ichthyosis- a disorder of cornification. To date, approximately 270 cases of collodion babies have been reported since the disorder was originally described. We report a unique case of a one day old female neonate who presented to our hospital with the presence of this disorder.

Keywords: Collodion, Icthyosis, Ectropion, Eclabion

Introduction

The word ichthyosis; is derived from a Greek word 'ikthus' meaning fish. Ichthyosiform dermatosis, are a group of hereditary disorders characterised by dryness and roughness of the skin with excessive accumulation of epidermal scales. Lamellar ichthyosis, is one of the rare congenital ichthyosiform dermatoses [1]. The characteristic feature of the disease is a thin, dry, shining, brownish-yellow parchment-like membrane which completely envelopes the newborn. This gives a collodion or "backed apple" look to the newborn and such children are called "collodion babies". The collodion baby is not a disease entity but phenotype common to several disorders. A total of 60%-80% of children born with a collodion membrane eventually develop ichthyosis either nonbullous ichthyosiform erythroderma or lamellar ichthyosis. Around 10%-20% eventually develop normal skin, referred to as self-healing collodion baby. [2] Here, we report an uncommon case who had classical clinical features of Collodion baby.

Case Report

A singleton preterm (32+2 weeks) female baby weighing 1.5 kg (Appropriate for gestational age) was delivered by Caesarean Section of non-consanguineous parents in Tata Main Hospital. The whole body of the baby was covered with a brown, dry, shining membrane. Due to tautness of the membrane, the eyes presented with marked ectropion

of the lids and the mouth was fixed in an open position with everted lips (eclabion).

On Local cutaneous examination, skin peeling was present at some sites over upper and lower limbs. Hairs and nails were normal.

Pictures of patient

Figure 1

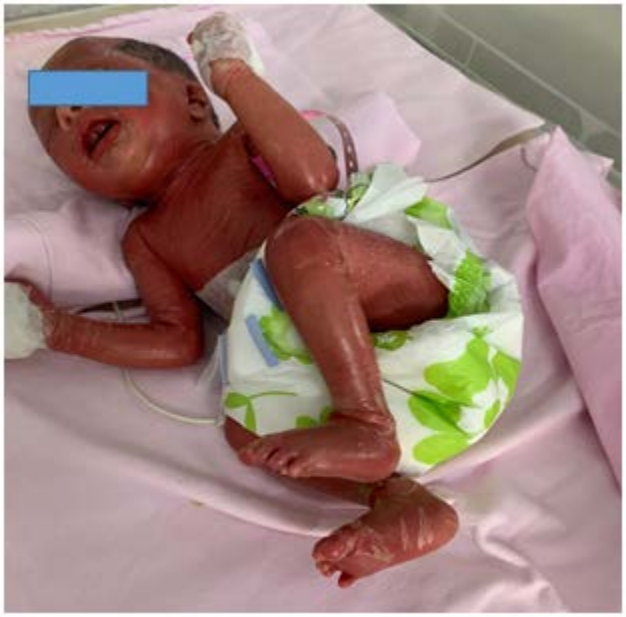

Figure 2

Systemic Examination

On examination

Baby's weight was 1500 gram (Just below the 50th percentile), length was 41 cm (50th percentile) and occipito-frontal circumference was 27 cm (50th percentile) on fenton chart.

General condition of the baby was fair on admission, Capillary Refill Time (CRT) was less than 3 sec, Heart Rate was145/min, Respiratory Rate was 42/min, Spo2 was 96% on room air, extremity was warm, pulses well felt. Systemic examination was within normal limit.

Based on the above clinical findings, diagnosis of Collodion baby was made because parents did not give consent for skin biopsy.

During hospital stay, baby remained hemodynamically stable and was on katoori spoon feed. 1st line antibiotic as per Nursery protocol were started empirically in view of suspected sepsis. Baby was kept in a humidified environment and frequent coconut oil application was done. Baby was treated for neonatal hyperbilirubinemia during hospital stay. Antibiotic was stopped after getting sterile blood culture. Baby was clinically stable, accepting feed well, passing stools and urine and was discharged in a healthy state from hospital.

Discussion

Collodion baby is a rare genodermatosis of unspecified inheritance, affecting both sexes equally. The neonate is born often prematurely, encased completely in a yellowish, taut, glistening, parchment-like membrane that restricts its movements minimally. The skin markings are obliterated and mild ectropion, eclabion, and flattened pinnae are usual. Nasal obstruction, when present, needs probing, as otherwise it will restrict breathing. The collodion membrane starts drying early and cracks / sheds completely within two weeks of life, exposing the infant

to the risk of hypothermia, skin and lung infections, septicemia, and electrolyte imbalance, with a propensity for renal and neurological damage. About 90% of these cases evolve into non-bullous ichthyosiform erythroderma or less often lamellar ichthyosis. The management is primarily supportive and nursing in humidified incubators at birth will prevent complications. Bland emollients are preferred to topical steroids lest their increased absorption, due to the compromised skin barrier, lead to systemic toxicity. Skin can be made soft and moist by applying emollients. The point to be remembered is to avoid urea containing agents because of chances of developing high plasma urea levels. Other drugs that are toxic like salicylates and retinoids should be avoided. [3] sub There is no need to operate immediately for ectropion as it can be corrected by local application of clobetasol in older children. [4]

Conclusion

Collodion baby be a manifestation of various conditions, [5] but often it is a manifestation of lamellar ichthyosis. Collodion baby represents difficult treatment challenge, because of prematurity, dehydration, temperature instability, and infection. Therefore **supportive care is most important to prevent mortality.**

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent form. In the form, the patient's parents have given their consent for the images and other clinical information to be reported in the journal. They 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.

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