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Case Report

Obstetrics and Gynaecology Section

Collodion Baby: A Clinical Enigma

SONAM SINGH, SUMITA MEHTA, TARUN K RAVI, ANSHUJA SINGLA, ANKITA MANN

ABSTRACT

Collodion baby is a term used for neonates in whom the body surface is covered by thick skin sheets, appearing like a translucent, tight parchment paper. This membrane is formed due to dysfunction of epidermal development resulting in epidermal hyperkeratosis. It is a rare condition with an incidence of 1 in 50,000-100,000 deliveries. The early neonatal period of a collodion baby can be complicated by hypernatremic dehydration, electrolyte imbalance, hypothermia and skin infections, resulting in high morbidity and mortality. Early diagnosis helps in optimal initial management and prevention of complications and thereby improves prognosis. Here, the present authors report a case of a collodion baby born and managed successfully in the institute.

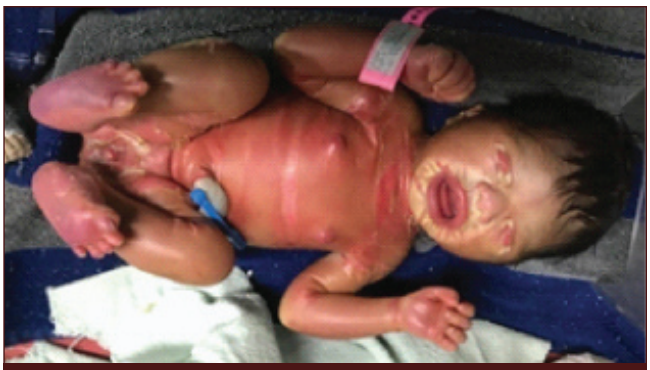
Keywords: Hyperkeratosis, Ichthyosis, Harlequin, Lamellar ichthyosis

CASE REPORT

A 30-year-old patient, delivered an alive term baby girl vaginally. Her antenatal period including ultrasound scan was unremarkable. A positive history of consanguinity was established. There was no history of any drug intake or exposure to radiation during the first trimester. She had no major medical illness in the past. There was no history of any skin or connective tissue disorder or any previous congenitally malformed baby delivered by the patient or in her family. She had a normal four-year-old male baby.

The baby at birth was covered with a thick taut membrane with scaly skin all over the body, along with ectropion and eclabium [Table/Fig-1,2]. There were no contractures of wrist, ankle or fingers and no organomegaly. Apart from these features the baby had a shrill cry, and had mild respiratory distress; APGAR score was 8, 9 and 9 at birth, 5 minutes and 10 minutes respectively.

The mother was stable after delivery and the baby was shifted to neonatal intensive care unit for evaluation and observation. Her vitals were normal and a complete blood count, sepsis screen and serum electrolytes were done. The baby was started on intravenous fluids and antibiotics regimen of cefotaxime and amikacin. A dermatological reference was taken and ointment framycetin was advised for application over the skin. The total leucocyte count of the baby was raised (22,500/cumm) but electrolytes level and platelet count were normal and CRP

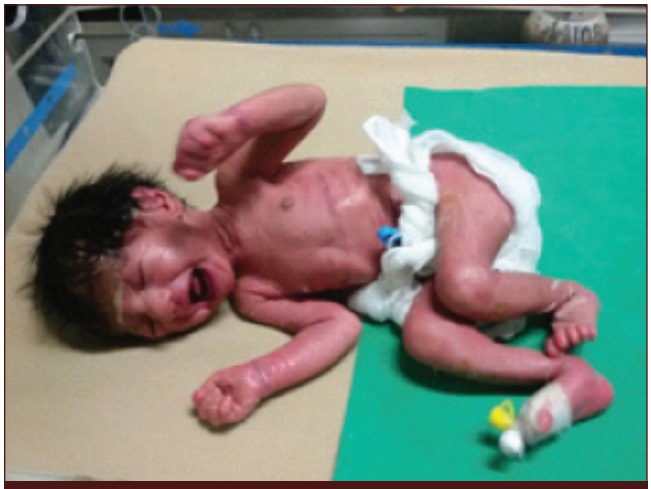


[Table/Fig-1]: Collodion baby with parchment-like membrane, fissuring in the body folds.

[Table/Fig-2]: Mild Ectropion in both eyes and eclabium.

was negative. Except for a mild respiratory distress, for which the baby was given oxygen by hood, her neonatal period was uneventful. She was started on Nasogastric (NG) tube feeding and breastfeeding on 5th and 8th postnatal day respectively.

The moulting of her skin started by day 5 and was almost completed by day 10 when she was discharged in satisfactory condition [Table/Fig-3].



[Table/Fig-3]: Moulting of skin complete by 8th postnatal day.

The child came for follow-up at three and six months and was doing fine. Due to low resource settings, further chromosomal or cytogenetic studies could not be done.

DISCUSSION

The term Collodion baby was first used by Hallopeau in 1884. Since then approximately 270 cases have been reported and the incidence of collodion baby varies between 1 in 50,000 to 300,000 deliveries [1]. Approximately, 75% of collodion babies develop a type of Autosomal Recessive Congenital Ichthyosis (ARCI); however, 10% of these babies have a milder form of the disease with normal underlying skin. ARCI is genetically highly heterogeneous and is associated with 6 main genes: *TGM1*(most common), *ALOXE3, ALOX12B, NIPAL4, CYP4F22* and *ABCA 12*; about 15% of affected families do not have pathogenic variants in any of the known genes [2]. Prognosis depends on the associated genetic mutation, severity of the initial manifestation, the duration of desquamation, as well as underlying ichthyosis. Neonatal complications can be seen in 45% of all collodion babies, leading to a mortality rate of 11% in first few weeks of life.

In a collodion neonate, a clinical diagnosis based on phenotype alone is often sufficient and a definitive diagnosis at this point is not essential, as the management is typically supportive for all such babies. During the months of infancy, the diagnosis becomes clearer as the patients display characteristic cutaneous phenotype, the most common being lamellar or non-bullous erythroderma. Other conditions associated are lipid storage disorders and Sjogren-Larsson syndrome. Skin biopsy and hair mount, though not routinely indicated can be done after 3-4 weeks when the membrane sheds off revealing the underlying skin disorder. Prenatal diagnosis is not routinely indicated but can be offered to parents with a previous history of a collodion baby either in themselves or their families. A woman who has had a collodion baby previously has a 25% risk of having a collodion baby in the next pregnancy and so ante-natal counselling and pre-natal testing should be offered to her.

The Collodion membrane which is made up of keratinised epithelium causes distortion in different body parts resulting in ectropion, eclabium (giving an "O" shaped configuration), pseudo-contractures, sparse hair, and hypoplasia of nasal and auricular cartilages [3-5]. The baby born in the present institute also had ectropion and eclabium, though she had no contractures. After birth, with chest expansion, the membrane starts tearing up and cracking and sheds off in 3-4 weeks. It is during this time that the baby is exposed to electrolyte imbalance, hypothermia, infections and aspiration pneumonia.

The management of such neonates is mainly conservative in the form of nursing in humidified incubators, prophylactic antibiotics and application of bland emollients to the skin [6]. In the present case also, the neonate was managed conservatively with intravenous antibiotics and emollient application. The moulting of her skin was almost complete by 8th postnatal day [Table/Fig-3] and the baby was discharged in satisfactory condition on 10th postnatal day.

CONCLUSION

In conclusion, a clinical diagnosis based on phenotype alone is sufficient for management of collodion babies. As the collodion membrane starts to tear and moult in the first post-natal week, only conservative management thus prevent babies from hypothermia, electrolyte imbalance and infections improve prognosis.

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AUTHOR(S):

1. Sonam Singh
2. Sumita Mehta
3. Tarun K Ravi
4. Anshuja Singla
5. Ankita Mann

PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Obstetrics and Gynaecology, Babu Jagjivan Ram Memorial Hospital, New Delhi, India.
2. Specialist and Head, Department of Obstetrics and Gynaecology, Babu Jagjivan Ram Memorial Hospital, New Delhi, India.
3. Specialist and Head, Department of Paediatrics, Babu Jagjivan Ram Memorial Hospital, New Delhi, India.
4. Specialist, Department of Obstetrics and Gynaecology, UCMS and GTB Hospital, New Delhi, India.
5. Senior Resident, Department of Obstetrics and Gynaecology, Babu Jagjivan Ram Memorial Hospital, New Delhi, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Ankita Mann,
F-49, Prashant Vihar, Rohini, New Delhi, India.
E-mail: ankitamann1988@gmail.com

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