Collodion Baby: A Rare Case Report

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Abstract: The term collodion baby is used for newborns in whom all the body surface is covered by thick skin sheets, so called “collodion membrane”. Ichthyosis is an infrequent clinical entity worldwide (1:300,000 births). When diagnosed in a newborn, two forms can be identified: collodion baby and its most severe form, harlequin fetus or maligna keratoma. In both cases, clinical manifestations are thick and hard skin with deep splits.

A case report of a collodion baby born in a community hospital of Rural Medical College, Pravara Institute Of Medical Sciences (DU), Loni, Maharashtra who was diagnosed and treated.

Collodion baby is a rare congenital disorder characterized by parchment like taught membrane covering the whole body. Other findings in our case include ectropion, eversion of lips, and flattening of nose and ears. Skin biopsy showed features consistent with the diagnosis of collodion baby. The child was treated with supportive measures like antibiotics, intravenous fluids, humidification, and application of emollients.

Keywords: Collodion baby, Ectropion, Eclabium.

I. Introduction

Only ~270 cases of collodion babies have been reported in the literature since 1892, when the term was first introduced by Hallopeau and Wattelet.¹ The first clinical description of collodion membrane by Pérez in 1880 continues to be valid: “The baby’s skin is replaced by a cornified substance of uniform texture, which gives the body a varnished appearance”.² As the name suggests, the term “collodion baby” refers to a phenotype that can be characterized by a yellow, shiny, tight parchment-like membrane stretched over the skin. Observers may sometimes use the descriptor “dipped in hot wax.”³ The most important clinical data concerning collodion baby is the presence of disseminated or generalized ichthyosiform genodermatosis characterized by dry skin, scaling, generalized erythroderma and hypokeratosis, reminiscent of fish scales. This type of dermatosis is also known by the generic name of ichthyosis.⁴

Harlequin ichthyosis, or harlequin fetus, is a severe generalized disorder of epidermal differentiation characterized by massive generalized hyperkeratosis and usually death in the perinatal period. Ultrastructurally, harlequin ichthyosis has an absence of normal lamellar bodies. At birth, the thick stratum corneum results in constrictive deformation, with severe ectropion and eclabium. Vital functions such as respiration and swallowing are compromised. Survival in the neonatal period is rare. Patients with harlequin ichthyosis represent the most severe end of the phenotypic spectrum of autosomal recessive ichthyosis.⁵

The clinical types of ichthyosis depend on the mode of inheritance as well as clinical and anatomic/pathological data.¹ There are several subtypes of each group. Among the true ichthyoses are three subgroups as follows: autosomal dominant ichthyosis (ichthyosis vulgaris, ichthyosis simple, fish skin disease) X-linked recessive ichthyosis (ichthyosis nigricans, ichthyosis of the male, sauroderma) and autosomal recessive ichthyosis (laminar ichthyosis, non bullous congenital ichthyosiform erythroderma).⁶ Rarely there may be an association with bullous congenital ichthyosiform erythroderma, Gaucher’s disease and Sjögren-Larsson syndrome. Furthermore, a new form of the disease with an autosomal recessive inheritance called “self healing collodion baby” has been notified where the newborn completely recovers in a couple of weeks.³³

We report a unique case of a male that was born as a Collodion baby at the University of Pravara Institute Of Medical Sciences (DU), Loni, Maharashtra. Although the impairment of the skin barrier function put the patient at risk for a number of complications, he was treated with emollients and antibiotics.
II. Case Report

A preterm (34 weeks) female child was born to a set of 3rd degree consanguinous parents, weighing 2098 grams. The baby was noticed to be encased in a parchment like membrane at birth along with mild ectropion. The baby was shifted to NICU and was started on iv fluids and antibiotics. Application of emollients was done hourly. Strict temperature control was maintained. Septic screen done for the child was normal. Biochemical parameters - namely liver function tests, renal function tests and serum calcium levels were within normal range.

The membrane started peeling from day one itself, revealing red coloured skin underneath. Ectropion started increasing progressively along with manifestation of eclabion. Artificial tears were applied regularly for the eye care. Strict asepsis was maintained whenever dealing with the baby.

Baby started showing signs of respiratory distress from 3rd day onwards and was no longer maintaining saturation at room air. Baby was given oxygen via hood box. The septic workup sent on the 3rd day revealed signs of severe septicemia – TLC count was as low as 1132 cells with platelet count of 32000. Renal function tests showed slight uraemia with blood urea being 53.2 and slightly increased serum sodium levels at 149. Antibiotics were stepped up for the baby.

On fifth day the condition of the child started deteriorating further and child was no longer maintaining saturation at room air. Child was put on ventilator support but the child still could not maintain saturation and finally succumbed to death on 5th PND due to septicemia.

COLLODION BABY: Pravara Institute Of Medical Sciences (DU), Loni, Maharashtra, India

III. Discussion

The term collodion baby applies to newborns who appear to have an extra layer of skin (known as a collodion membrane) that has a collodion-like quality. It is a descriptive term, not a specific diagnosis or disorder (as such, it is a syndrome). This is exactly noted in our patients who had extra layers of transparent skin.

The natural course of collodion membrane is intriguing. For instance, approximately 75% of collodion baby will go on to develop a type of autosomal recessive congenital ichthyosis, either lamellar ichthyosis or congenital ichthyosiform erythroderma. Another 10% of cases the baby sheds this layer of skin and has normal skin for the rest of its life. This is known as self-healing collodion baby. This is akin to our series. The remaining 15% of cases could stem from variety of diseases involving keratinization disorders. Known causes of collodion baby include ichthyosis vulgaris and trichothiodystrophy.

The major cause of collodion baby syndrome is not well known. However it has been known to be inherited in an autosomal recessive fashion. Placental insufficiency and post maturity have also been implicated in some forms of collodion membrane formation. This could be due to the effects of DNA repair and transcription gene abnormalities in human pre-natal life. Trichothiodystrophy (TTD), a rare (affected frequency of 10−6) recessive disorder caused by mutations in genes involved in nucle
otide excision repair (NER) pathway has also been implicated.\textsuperscript{12}

Due to the impairment of the skin barrier function, collodion babies are at risk for a number of complications, including hypernatremic dehydration, hypothermia, skin infections, fissures, conjunctivitis, sepsis, dehydration, and constrictive bands of the extremities resulting in vascular compromise and edema.\textsuperscript{1,13} The edema in the patient described here was thought to be due to either hypoproteinemia or mechanical compression by the collodion membrane. One study showed that the transepidermal water loss in collodion babies can be six to seven times higher than that through normal skin.\textsuperscript{14} Skin impairment gives rise to transepidermal water loss (TEWL), percutaneous infection, and toxicity. Accurate intake and output is essential, as is the accurate monitoring of electrolytes for hypernatremia and the appropriate adjustments of fluid intake. Measurement of TEWL with an evaporimeter should be considered. Therefore, it is essential that collodion babies be placed in a humidified incubator soon after birth to prevent hypernatremic dehydration and hypothermia. The patient described here was managed accordingly with the use of emollients and prophylactic antibiotics.

\section*{IV. Conclusion}

Because this is a rare disease it is indispensable to have very clear and precise information on the steps to follow and the complications that may arise. In order to determine the etiologic cause for the collodion membrane, a protocol must be established so that appropriate measures can be taken months or years following the shedding of the collodion membrane.

An incubator provides a humidified, neutral temperature environment. Other supportive treatments such as intravenous fluid and tube feeding are often necessary. The aim is to keep the skin soft and attempt to reduce scaling. The collodion membrane should not be debrided (pulled off).

In our experience through this case study, we recommend careful attention to skin care, minimal use of skin care products, and meticulous attention to preventing infection when caring for an infant with collodion baby syndrome.

\section*{References}

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