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Address of Correspondence

Executive Editor, TMSS Medical College Journal
TMSS Medical College, Rangpur Road
Thengamara, Bogra, Bangladesh.
Phone: 051-61830, Fax : 051-61830
e-mail: editortmcjournal@gmail.com
Web: www.tmssmedicalcollege.com

Case Reports

Collodion baby : An uncommon clinical case report

Mondal CK[1]

1. Dr. Chanchal Kumar Mondal, Assistant Professor, Department of Paediatrics, TMSS Medical college and RCH , Bogra.

Abstract

Collodion baby describes a highly characteristic clinical entity in newborns encased in a yellowish translucent membrane resembling collodion. The collodion membrane is composed of thick skin sheets which resemble translucent, tight parchment paper. The purpose is to report the rare occurrence of collodion baby among our population. In this report we present a rare case of collodion baby in whom the skin was parchment like, Shiny and thickened with distorted facial feature like ectropion and eclabium with pseudo contracture of digits. In almost all of the collodion membrane cases an autosomal recessive ichthyosiform disease is implicated. Usually these babies are premature. Congenital Lamellar Icthyosis also known as Icthyosis lamellaris or non bullous congenital icthyosis is a rare inherited, phenotype Autosomal Icthyosis (ARCI). Gradually these children will develop signs of one of several types of icthyosis which gives the skin appearance of "Fish Scales". Conclusively, these newborns should be monitored carefully in intense care units and is difficult to diagnose in antenatal period.

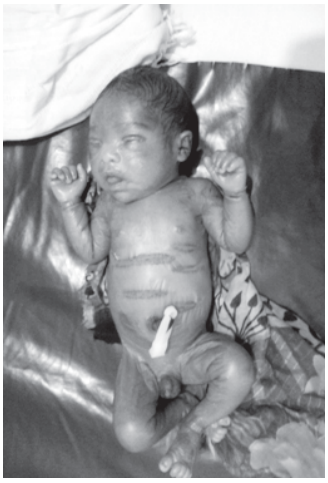
Key Words: Collodion Baby, Neonates, Genetic Disorder.

Introduction

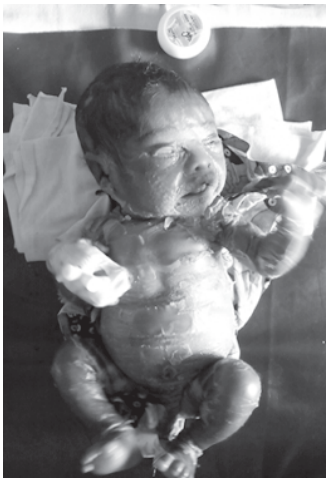
The first clinical description of collodion membrane (Pérez, 1880) continues to be valid: "The baby's skin is replaced by a cornified substance of uniform texture which gives the body a varnished appearance".[1] The term collodion baby refers to a clinic entity used for newborns who are encompassed by a translucent, tight and parchment paper like skin sheets so called collodion membrane, on the entire body surface.[2.3.4] Collodion baby as a term was first used by Hallopeau in 1884.[4,5,6] Although, the pathogenesis of molecular mechanisms apparently lead to an epidermal cornification disorder, keratinocyte protein and lipid metabolism defects resulting from autosomal recessive genetic mutations have also been notified as important cofactors.[3] The cause of both autosomal recessive lamellar ichthyosis and congenital ichthyosiform erythroderma (nonbullous) have been reported to be transglutaminase 1 gene mutation localized on the 14q11.[5, 7.]

About 270 cases of collodion baby have been reported since 1884.[8] It is a congenital disorder, occurring with an incidence of 1:300,000 live birth[9] and both gender are equally affected.[10] Collodion baby is also known as collodion fetus.[11] Collodion baby is characterized by shiny, tight,cellphone-like membrane stretched over the skin.[12]

The collodion membrane is a temporary condition, which desquamates later on, and ultimately these children manifest sign of ichthyosis. About 45% of collodion babies develop some complication due to compromised skin barrier function. It is associated with mortality rate of approximately 11%.[13]



Picture-1



Picture-2

Case Report

A 38 weeks preterm girl was born to 22 years old primiparous mother by spontaneous vaginal delivery at RCH Bogra. She was the first live born of the non-consanguineous parents (picture-1,2). There was no family history of the similar condition and Antenatal history also uneventful. The Karyotypes done were normal. On examination notable findings include baby was encased in a tight, transparent, shiny membrane with restricted movements, eyelids were turned outwards, ectropion, Lips were turned outwards, eclabium, Fissuring were over trunk, limbs. Exfoliation of skin over the hands and feet. The baby's birth weight was 2.6 kg, and head circumference of 34 cm. A clinical diagnosis of collodion baby was made based on clinical findings as noted above. Treated with local application of liquid paraffin, lacrimal eye ointment, with artificial teardrops and prescribed one intravenous antibacterial agent to combat infection. Infant was managed conservatively and started accepting breast feeds by 4th day of postnatal life. She was discharged on 15th day when her clinical condition was stable. They were also counseled regarding prolonged period of observation.

Discussion

The collodion baby is a clinical entity, referring to newborns who have extra sheets of skin, termed as collodion membrane**.**[18] Collodion baby is a rare condition needs more care and attention in the neonatal period.**[17]** The newborns are encased in glistening, taut, parchment-like membrane.[19] Furthermore, described as dipped in hot wax.[18] The neonates are usually born prematurely.[17] The tight collodion membrane over the face leads to eclabium (eversion of lips), ectropion, and deformed pinna. Collodion membrane can restrict breathing, swallowing, and movement at joints.[16,20] Nasal obstruction can cause diffculty in breathing, which may require probing.[16] Sometimes hairs are absent.[19] The collodion membrane begins to dry early, and cracks in 48 hours usually shed off completely in 2 or more weeks. Finally, collodion membrane is replaced by normal appearing skin. The desquamation causes impairment of skin barrier function and fissure formation.[14, 15]. In this study the newborn presented with clinical features include baby was encased in a tight, transparent, shiny membrane with restricted movements, eyelids were turned outwards, ectropion, Lips were turned outwards, eclabium, Fissuring were over trunk, limbs. Exfoliation of skin noted over the hands and feet.

It is the early presentation of various congenital ichthyosis.[17] Nearly, 75% of collodion babies progress to autosomal recessive congenital ichthyosis such as lamellar ichthyosis and congenital ichthyosiform erythroderma. About 10% of collodion fetus will have normal skin after the membrane is shed off. They are termed as self-healing collodion babies. Remaining 15% of collodion babies develop other keratinization defects such as Netherton syndrome, Gaucher disease type[15], ichthyosis vulgaris, trichothiodystrophy, and Sjogren-Larsson syndrome.[21]

Diagnosis based on the evolution of the cutaneous findings, associated abnormalities and family history.[22] In the immediate neonatal period skin biopsy may be non-specific. Histologic evolution may be postponed until after the age of 3-6 months[23].

Complications include temperature instability, defective barrier function, increased insensible water loss predisposing to hypernatremic dehydration.[24] (Buyse et al 1993). Pneumonia secondary to aspiration of squamous material in the amniotic fluid and cutaneous infection from gram-positive organisms and candida albicans.

Treatment consists of aggressive supportive care. Infants must be placed in a highly humidified isolette. Fluid and electrolyte balance must be closely monitored. A high index of suspicion must be maintained for signs of cutaneous or systemic infection. Overzealous administration of antibiotics how ever may lead to gram-negative infections and subsequent septicemia. Topical skin care should include application of a bland occlusive ointment. Emollient every 6-8 hours until the hyperkeratosis has resolved. Potentially toxic topical agents should be avoided because of the increased risk of percutaneous absorption. Manual debriment is not indicated. The eyes should be protected with a bland lubricating ointment. Aggressive surgical management of ectropion is almost never necessary. Systemic retenoids have not been useful[25] (Waisman et al, 1989). With optimal supportive care, the thickened stratum cornium usually resolve in 2-4 weeks, but can persist, especially in infants with Lamellar Ichthyosis. Several out comes have been reported including complete healing without sequelae [26, 27]

The newborn in this case repot was treated conservatively with local application of liquid paraffin, lacrimal eye ointment, with artificial teardrops and one intravenous antibacterial agent. She was discharged after two weeks of postnatal period on well condition with proper counseling.

Conclusion

Collodion baby is a rare condition, which requires proper supportive care. A prolonged period of observation is necessary to determine the precise diagnosis and prognosis. As soon as a definite diagnosis has been made genetic counseling should be provided.

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