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Collodion Baby –A Rare Case

Navina Singh[1*], R. P. Patange[2], Manisha M. Laddad[3]

{[1]Resident, [2]Professor and HOD, [3]Assistent Professor} Department of Obstetrics and Gynecology, Krishna Institute of Medical Sciences, Karad, Maharashtra, INDIA.

[*]Corresponding Address:

navinasingh24@gmail.com

Case Report

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". The collodion membrane is the result of an epidermal developmental dysfunction. The collodion membrane is composed of thick skin sheets which resemble translucent, tight parchment paper. In almost all of the collodion membrane cases an autosomal recessive ichthyosiform disease is implicated. Especially, in cases of lamellar ichthyosis, congenital ichthyosiform erythroderma and harlequin ichthyosis frequent association with collodion baby formation has been well documented. Clinically, the collodion babies may encounter dehydration, electrolyte imbalance, temperature malfunction and increasing sepsis risk because of relatively severe skin damage. Therefore, morbidity and mortality rates are fairly high in these cases. Conclusively, these newborns should be monitored carefully in intense care units and is difficult to diagnose in antenatal period.

Keywords: collodion baby, new born.

Case

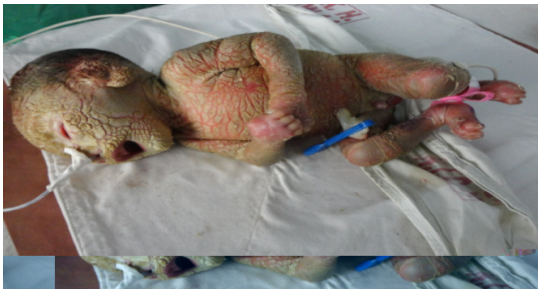
A 20 yrs primigravida with 30 weeks pregnancy got admitted in KIMS on 01/6/12 with chief complain of pain in abdomen. The mother was a registered case with us and had prenatal care from the first trimester with h/o consanguineous marriage. No h/o any congenital anomalies in family .No history of drug intake or exposure to radiation during 1st trimester. She had no major major medical illness or major surgery in past. She was admitted in labor and was given tocolytics and steroids. On examination her vitals were stable. On her abdomen examination uterus was 28-30 weeks size, cephalic presentation with mild contraction and FHS present. On speculum examination cervix and vagina healthy with no demonstrable leaking. On pervaginal examination cervix 1cm dilated, soft, minimally effaced and presenting part vertex with station high up. All blood and biochemical investigations were normal.

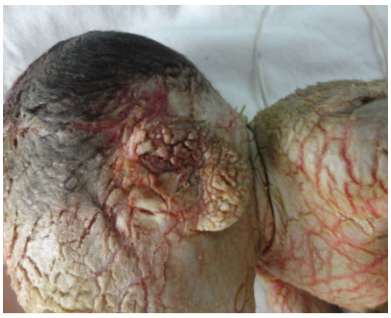
Ultrasonography

Single life intrauterine fetus of 27 weeks with breech presentation. Placenta is located anteriorly. Fetal weight 1000kg. Liquor adequate.
No congenital anomalies.

Management

It was a preterm vaginal delivery, male child, cried immediately after birth. The newborn had an Apgar score of 7 & 8, birth weight 1.308kg with maturity of 30-32 weeks. Upon initial physical examination hard whitish covering with scaly skin present all over the body, eyelids with ectropion with contractures of wrist, ankle fingers present. Cry tone and activity was depressed .Pulmonary field showed adequate air movement without evidence of wheezing or rales. Rhythmic heart sounds had good intensity without aggrevated phenomenon. No organomegally present.





Post delivery mother was stable and baby was shifted to NICU. In NICU various laboratory studies were performed in order to rule out congenital infections. Vitals of baby were normal but dehydration was present. All blood investigations were performed and reported with platelet count 60,000/cumm, PCV-48. Based on investigations baby was started with an antibiotic regimen and IV fluids was given. Liquid paraffin was applied all over the body. Baby was started with direct tube T feeding on 3 post natal day .The baby started deteriorating thereafter. On 6post natal day baby went into apnoea and expired. Baby was not sent for autopsy as relatives were not willing.

Discussion

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.(1,2,3) Collodion baby as a term was first used by Hallopeau in 1884(3,4,5). Since then approximately 270 cases were reported. Although, some other diseases and situations may lead to collodion membrane formation in almost all the cases the cause is an autosomal recessive ichthyosiform disease. The clinical types of ichthyosis depend on the mode of inheritance as well as clinical and anatomo-pathological data. Ichthyosis can be classified into three groups: 1) true ichthyosis, 2) ichthyosiformstates and 3) epidemolytic hyperkeratosis. There several subtypes of each group.

Among the true ichthyosis are three groups as follows: autosomal dominant ichthyosis (ichthyosis vulgaris, ichthyosis simple, fish skin disease), X-linked recessive ichthyosis (ichthyosis nigricans, ichthyosis of the male, saurodermia) and autosomal recessive ichthyosis (laminar ichthyosis, nonbullous congenital ichthyosiform erythroderma). The frequently reported clinical subtypes: congenital erythrodermic ichthyosis, 48%, lamellar ichthyosis, 12% and ichthyosis vulgaris dominant in 10% of patients. In 10% of cases, the skin eventually developed normally.

Pathogenesis

The collodion membrane occurs due to an epidermal cornification disorder just like all the ichthyosiform diseases. 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 [4, 7]. Moreover, both varied molecular pathogenesis mechanisms and 5 different gene localizations and more than 50 gene mutations in these genes have been detected.

Table 1: Diseases Associated with Collodion Baby

- Autosomal recessive congenital ichthyoses [lamellar ichthyosis, congenital ichthyosiform erythroderma (nonbullous form), harlequin ichthyosis]
- Epidermolytic hyperkeratose (bullous congenital ichthyosiform erythroderma)
- Gaucher's disease
- Sjögren-Larsson syndrome
- Self- healing collodion baby
- Neutral lipid storage disease
- Trichothiodystrophy
- Annular epidermolytic erythema
- Loricrin keratoderma
- X-linked to hypohydrotic ectodermal dysplasia

Clinical Features

The collodion babies may be born with a collodion membrane covering the entire skin surface just like an armor .This situation limits both the baby's respirationand sucking function. The collodion membrane peels off in two or more weeks frequently leaving behind fissures and skin barrier dysfunctions. As a result serious complications like risk of infection, fluid loss, hypernatraemic dehydration, electrolyte imbalance and thermal instability may be encountered. The collodion babies are usually premature at birth. Therefore, these babies should be monitored very carefully [2, 6]. The eyelids and the lips may be everted and tethered (ectropion and eclabion). In such cases loss of proper management can result with keratitis due to

xerophthalmia and eventually blindness.While the skin peels off, the residual skin becomes dry and tough. Particularly, tight membrane on limbs may lead to construction and loss of function [3, 6]. While the child grows up, also the symptoms and findings of the premier diseases which have caused the collodion baby appearance begin to arise (Table 1). However, some cases of collodion babies that have healed spontaneously in a few weeks and no associated disease have ever been determined are also

Treatment

In collodion babies' fluid and electrolyte balance and body temperature must be carefully monitored. In addition to this the membrane must be lubricated and to achieve elasticity and desquamation an adequate hydration of the skin are the major components of management. Suitable eye care and pain control should be carried out for the collodion babies with ectropion. Humidified incubators and water dressings followed by emollient agents are the essentials of the management. If there is a respiratory failure ventilative support for the collodion babies may surely be needed. In cases of epidermolytic hyperkeratose (bullous congenital ichthyosiform erythroderma) which show generalized erythema, bullae and erosions an antibacterial will be needed along with the standard therapy. The collodion babies with large areas of skin erosions are always under the risk of heavy infections an even sepsis therefore suitable local and systemic antibacterial agents must be cautiously determined and preferred [6]. The drugs such as salicylic acid, lactic acid and propylene glycol may be applied in order to remove the hyperkeratotic sheets from the skin. But in such cases with generalize lesions, particularly in newborns it must not be forgotten that the application of salicylic acid locally in extreme doses may cause salicylic acid toxemia. Therefore, local remedy in these cases should be cautiously monitorized and carried out in this way [4].However, in the collodion babies with localized lesions local retinoic acid and calsipotriol treatments have been reported to be successful. Systemic retinoids which are currently a preferred treatment method giving impressive results in cases with generalized lesions have been notified as a perfect alternative. When systemic retinoids came into the practice the mortality rates decreased expressively. In a study, acitretin has been used at the dose of 0,5-0,75 mg/kg/day and the mortality rate of collodion babies has decreased to 11% (1986) compared to the numbers of 1960 (50%).

In cases of lamellar ichthyosis, systemic retinoids have been begun at doses of 0,5 mg/kg/day and later on the doses have eventually been increased to 2 mg/kg/day. It is notified by the authors that the greatly thick scales have improved expressively.

Systemic retinoids have also shown to been effective for cases with congenital ichthyosiform erythroderma.

Although, harlequin fetus is rare among all the ichthyosiform diseases, without any dispute it is the severest form. Hence, high mortality rates have been observed. Nonetheless, in recent years, in these rates have also been declined because of the entrance of systemic retinoids into the clinical practice and the advanced care methods which are used in intensive care units [4].

Conclusion

Mortality of collodion baby has decreased as a result of multidisciplinary care and opportune diagnosis and management but still its diagnosis in antenatal period is difficult.The outcome of next pregnancy is not predictable and can be diagnosed only after conception on amniocentesis with karyotyping.

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