Case report

Colloidon baby – Rare case with preventable complications

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Abstract
Colloidon baby is a rare congenital disorder characterized clinically by parchment like taught membrane covering the whole body at the time of birth, which subsequently develops Non bullous ichthyosiform erythroderma or Lamellar ichthyosis in most cases and in few cases other ichthyosiform disorders. The colloidon membrane spontaneously desquamates within 2 weeks or up to 3 months in few cases. Herein, we present 2 cases of colloidon babies born to consanguineously married couples of which the first baby was born at term by normal vaginal delivery and second baby born prematurely by caesarean section. Both 1st & 2nd baby were delivered in different private hospitals in villages of Nizamabad district, Telangana state and reported to tertiary level children’s hospital in Hyderabad city on 4nd and 6th day of life respectively with complaints of colloidon membrane and maceration of skin in diaper area, was admitted in Neonatal intensive care unit (NICU) in humidified incubator, treated with emollients, intravenous fluids and prophylactic antibiotics to avoid complications. Nursing care is of prime importance. This presentation was aimed at stressing not only the importance of early recognition by pediatrician & timely referral to dermatologist and ophthalmologist for saving life of affected baby but also equal importance to proper nursing care.

Key words: Colloidon baby, Colloidon membrane, Clingfilm wrap, sausage skin, plastic skin, skin dipped in hot wax.

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Colloidon baby is a rare congenital disorder characterized clinically by parchment like taught membrane covering the whole body at the time of birth, which subsequently develops Non bullous ichthyosiform erythroderma or Lamellar ichthyosis. Colloidon is a syrupy solution of pyroxylin (nitrocellulose) in ether and alcohol which has medicinal and commercial uses. So far only 267 cases were reported since 1892 in the literature, familial self-healing cases & localized forms have been noted.

Case report

Case 1: A male baby born to primi, 1st degree consanguineous married couple with colloidon membrane at time of birth. Baby delivered by normal vaginal route at 37 weeks of gestation in private hospital with birth weight of 2.10kgs and was referred to our hospital on 4th day of life (Fig 1 & 2).

Case 2: A male baby born to primi, 1st degree consanguineous married couple with colloidon mem-
bran at time of birth. Baby delivered by caesarean section at 35 weeks of gestation in private hospital with birth weight of 1.90kgs and was referred to our hospital on 6th day of life (Fig 3).

Both babies were brought by their parents with complaints of tough yellowish covering all over body, eversion of eyelids and lips since birth and macerated skin around diaper area and high grade fever since 2 days & 3 days respectively. Nursed in hospital just like other normal babies, causing additional problem which could have been preventable. Relevant investigations were done. Managed by pediatrician, dermatologist and ophthalmologist in neonatal intensive care unit (NICU).

Discussion
The collodion membrane is thought to develop during the third trimester, the exact cause of formation of collodion membrane is unknown, but placental insufficiency and post maturity are implicated. At birth the defective stratum corneum barrier, associated with the collodion membrane, shifts from an aquatic to a dry environment, and the membrane desiccates and peels off within 2 weeks. The association between periderm and formation of collodion membrane is unknown. The collodion membrane shares the same etiopathogenesis as the subsequent ichthyosiform erythroderma, which evolves in the majority of collodion babies. Functional mutations in transglutaminase 1, ALOXgenes, ABCA12, ichthyin, ABHD5and other genes involved in epidermal lipid and protein homeostasis have been identified in ichthyosiform erythrodermas preceded by collodion baby presentation. Self-healing collodion baby is caused by compound heterozygous transglutaminase 1 mutations which in utero render the enzyme inactive in its cisform. In the extra uterine environment the enzyme isomerizes back to its partially active transform, allowing a normal phenotype. The rare type 2 variant of the lysosomal (sphingolipid) storage disorder, Gaucher disease is due to β-lucocerebroside mutations. It presents with collodion membrane and evolves to ichthyosis. Ultra structural abnormalities in lamellar body and membrane formation and defects in ceramide production underlie the epidermal dysfunction. Transglutaminase deficiency, keratin-associated-protein defects (e.g. loricrin mutations) and other lipid metabolic disorders also cause collodion membrane and ichthyosis by disrupting epidermal differentiation and function.

The histopathology of light microscopy of skin in early neonatal phase of a collodion membrane shows a compact hyperkeratosis with a thick eosinophilic periodic acid–Schiff stain (PAS)-positive stratum corneum. However, the epidermis is attenuated because of a reduced granular layer. The mid- and lower epidermis and the dermis are usually unremarkable. Electron microscopy features in the early neonatal phase vary. In one case, cells of the upper two-thirds of the stratum corneum were convoluted and irregular in shape, with prominent nuclear debris and dense intracytoplasmic granules. Lamellar bodies were numerous in the intercellular spaces and desmosomes were well preserved. Repeat skin biopsy at 16 months showed that, in spite of apparent self-healing, the ultra structural changes persisted. However, in a further two cases, the ultra structural features on the 15th day of life were predictive of the outcome, with a self healing case showing relatively normal epidermal ultra structure, while an infant who developed erythrodermic ichthyosis showed continuing changes. In our cases skin biopsy was deferred as parents were scared.

Premature delivery is more common, as noticed in our 2nd case. At birth, the typical collodion baby presents a striking and characteristic clinical picture with a generalized glistening, taut, yellowish film stretched over the skin, which was seen in our cases. It resembles a Clingfilm wrap or sausage skin, described as ‘plastic skin’, ‘parchment-like’ or
‘as if dipped in hot wax’. Normal skin markings are obliterated, the eyelids and lips are tethered and everted respectively (ectropion and eclabion), and these features were more prominent in 2nd case. Pinnae may be flattened and the nostrils obstructed. Sausage-shaped swelling of digits may result from acral extension of the membrane and, rarely, constricting bands encircle the limbs or digits. General examination is usually normal and movement is minimally restricted.

The collodion membrane desiccates and cracks around flexures during the first few days of life, and is usually completely shed within the 1st two weeks of life. It may be incomplete or localized in some infants; it may peel away only to partially reform, this pattern being repeated over a period of up to 12 weeks. In most cases, shedding reveals an erythrodermic ichthyosis. Baby must be admitted in neonatal intensive care unit (NICU) in humidified incubators and treated with emollients, nasogastric feeding, prophylactic intravenous antibiotics & eye care. The common and potentially lethal complications (45%) of a collodion membrane, especially when associated with erythroderma, were hypothermia, renal failure, neurological sequelae of hypernatraemic dehydration, constricting bands of extremities, which leads to vascular compromise and edema skin infections. bacterial sepsis, electrolyte imbalance and dehydration. Increased absorption of topical agents through the skin may lead to systemic toxicity which must be avoided like steroids, urea, salicylates and retinoids. One study showed high plasma urea levels. Improved neonatal care has resulted in a reduction in neonatal mortality from 33% in the 1970s to less than 11% in one review and in uncomplicated cases, mortality is now negligible. In 60–80% of these infants, the features of non-bullous ichthyosiform erythroderma or lamellar ichthyosis become apparent as the collodion membrane is shed. The severity of the subsequent ichthyosis cannot be predicted.

The rare autosomal dominant form of lamellar ichthyosis also presented with collodion membrane. Some patients with lamellar ichthyosis or non-bullous ichthyosiform erythroderma appear not to have a collodion membrane at birth. Certain ichthyosiform syndromes, including trichoiodystrophy (IBIDS), neutral lipid storage disease, Sjögren–Larsson syndrome, Conradi–Hünermann disease and chime syndrome, and the autosomal dominant loricrin keratoderma may present with collodion membrane. Up to 20% of collodion babies subsequently have normal or near-normal skin, so-called self-healing collodion baby or ‘lamellar ichthyosis of the newborn’. Mutation of the ALOX12B and ALOXE3 genes underlie most of the Scandinavian cases of ‘self-improving collodion ichthyosis’. An acral self-healing collodion baby phenotype has also been reported.

Collodion membrane has also been reported in anhidrotic ectodermal dysplasia. Gaucher disease - type 2 and Neu–Laxova syndrome. An adult with isolated palmoplantar keratoderma who was a collodion baby, was observed by the author and a similar case was included in a review of 10 collodion babies in the Thai literature. A further case of evolution of collodion baby to palmoplantar keratoderma also had anogenital leukokeratosis.

A collodion baby is generally easily distinguished from the more severe harlequin fetus presentation, but occasional cases with intermediate features have been recorded named as ‘chrysalis babies’. Restrictive dermopathy, or ‘stiff baby syndrome’, produces a generalized taut, thick, tethered and unyielding skin at birth, which does not desiccate in the neonatal period. Its persistence causes respiratory failure and early neonatal death. Collodion-like presentations have been described in Gaucher type 2 and Neu–Laxova syndromes. Infective causes of desquamation such as staphylococcal scalded skin should be included. Skin biopsy is not usually performed at this stage, but hair analysis, blood film microscopy can be done. Collodion babies should be nursed in the neonatal intensive care unit (NICU) in a humidified incubator. Cleansing with warm water and sterile aqueous cream or antiseptics daily and barrier nursing reduces the risk of infection. An emollient, such as sterile white soft paraffin in single-dose containers, is usually applied 4-hourly to keep the membrane pliable and reduce transepidermal water loss. In these cases emollient consisting of mix-
ture of petroleum jelly and paraffin oil was used & intravenous antibiotics started for infection, as skin barrier was impaired more so in diaper area. Signs of skin or systemic sepsis including Candida and Pseudomonas require investigation and prompt treatment, which was negative in our cases.

Poor sucking and weight loss may necessitate nasogastric tube feeding, and a higher than normal fluid and calorie requirement is anticipated. The risk of exposure keratitis due to ectropion can be reduced by the regular use of lubricating drops and ointments, and antibiotic if necessary. Early ophthalmological advice should be sought. Nasal obstruction can be improved by gentle probing and constricting bands on the limbs should be divided if causing acral pressure effects. Every effort must be made to allow the parents to nurse and bond with their baby and their involvement from the beginning will enhance their confidence in managing at home23. The need for continuing intensive care once the membrane is shed depends on the baby’s condition, can be discharged with a skin care plan and home nursing support. Accurate information on the condition should be made available to family and carers.

Conclusion: Colloidon baby is a rare condition, spontaneously heals in 2 weeks. Early recognition and prompt referral to the specialists / tertiary level hospitals can prevent complications and save lives of babies with colloidon membrane. This presentation was aimed at stressing importance of judicious nursing care in addition to medical care, so that complications can be prevented to large extent.

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References