CASE REPORT

COLLOIDION BABY – A CASE REPORT

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ABSTRACT
A female newborn baby delivered by LSCS to a primi mother with second degree consanguinity with typical features of colloidion baby is being presented. If this condition being recognized early and with early institution of therapy morbidity and mortality can be significantly reduced.
Key-Words: Colloidion Baby, Icthyosis, Neonate

Introduction

Colloidion baby is one of the variety of icthyosis and is inherited as autosomal recessive manner.[1] The condition is usually a manifestation of Congenital Icthyosiform Erythroderma (CIE) or Lamellar Icthyosis (LI) and is responsible for about 2/3 of colloidion babies.[2] The above two lesions transmitted as autosomal dominant trait in about 10% of cases. Sex linked Icthyosis never begins with a colloidion baby syndrome.[2] Colloidion baby born with encasement of shiny tight inelastic scale resembling oiled parchment is designated as having colloidion membrane which will subsequently shed and is a self-limiting condition. Approximately 11% of these babies will die during first few weeks of life[2,3], most babies later develop severe type of icthyosis, skin of these babies appears like scales of fish.[4]

Case Reports

In our hospital a female newborn baby delivered by lower segment cesarean section (indication being premature rupture of membrane and non-progression of labour) to a second degree consanguineous couple with uneventful antenatal and natal history. Because of the abnormal appearance, baby was referred to neonatal intensive care unit. No similar history in the family. On examination it was a term female baby weighed about 2.6 kg, length 48 cm, head circumference 33cms, chest circumference 31 cm. Vitals were stable. There was an ectropian (figure 1), absence of eye lashes and eyebrows. Fish like mouth (figure 1), flat nose and small flat ears, skin was parchment like feel with varying degrees of fissures at groin and axilla. Fish like scales over abdomen and back (figure 1), there was joint stiffness with flexed skin at all joints of hands and legs and swollen dorsum of feet and hands, nails were hypo plastic, neonatal reflexes could not be elicited because of joint stiffness.

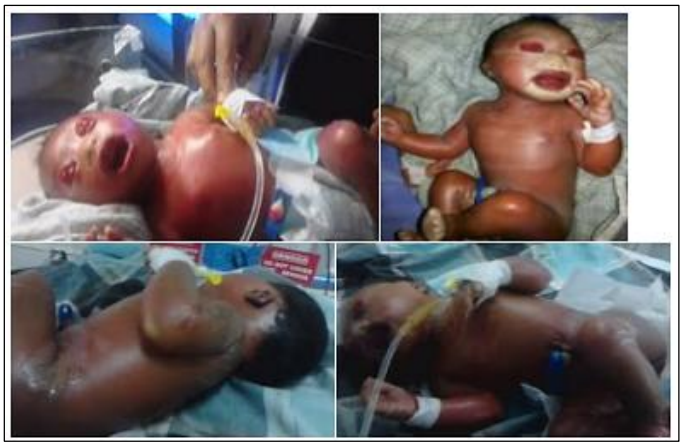

Figure-1: Colloidion Baby

Baby was given warm care, oronasal suction was done, hydration was maintained with intravenous fluids and empirical antibiotics were started because of premature rupture of membrane (>12 hours). The routine blood investigations were normal with no elevation of septic markers, hydration status well maintained and baby skin smeared with coconut oil as a lubricant to facilitate desquamation of skin. After 24 hours of life baby was looking better without any respiratory distress. Since suckling of the baby was weak due to tight skin, nasogastric feed was started with expressed breast milk and full feed was established by third day of life. It was also noticed skin scales were started peeling. The further management was discontinued as the parents were not willing stay back in hospital and later did not turn up for follow up. Maintenance of hydration nutrition skin care was explained up to 4 to 6 weeks as there will be spontaneous improvement in some type of colloidion babies.

Discussion

Most colloidion babies do have a form of icthyosis and majority of them develop features of lamellar icthyosis, bullous icthyosis, X–linked icthyosis, netherton's syndrome or Goucher's disease.[5] These babies are usually at birth covered by thick tight membrane resembling oiled parchment membrane or colloidion which is subsequently shed, often resembling ectropion and eclabium Figure 1. Within few hours of life membrane dries cracks and peels off but may reform several times. Infrequently an affected infant has normal skin after the membrane is shed and affected neonates have ectropion, flattening of ears and nose, fixation of lips in an O shaped configuration. Our case also had similar features with decreased activity because of tight skin and stiff joints, sucking was poor because of tight lips. The skin shortly after birth begins desquamation in large sheets and complete shed may take several weeks and a new membrane may form in a localized areas.

The colloidion membrane is greatly composed of thickened stratum corneum that has been saturated with water. As the water content evaporates in extra uterine life large fissures appear in membrane and membrane is shed revealing red skin underneath.[6-10] The taught membrane may impair respiration and sucking as in our case, fissured skin leads to water loss and prone for infection leading to difficulty in thermal regulation and hypernatremic dehydration. Complications include cutaneous infection, aspiration pneumonia, and hypothermia or hypernatremic dehydration. It may be associated with medical problems like, overheating; eye problems; constriction bands (tight bands of skin around toe and finger tips; hair loss; patchy loss which is rare but permanent.[11-12] The outcome is uncertain and accurate prognostification is impossible with subsequent development of icthyosis.[11-12]

Treatment initially consists of high fluid intake to avoid dehydration and prevent transepidermal fluid loss and use of heated humidified incubators and emulsifying ointment and retinoids, vitamin D preparations, bathing using bath oils or application of lubricant before drying can prolong hydration and skin softening. Keratolytic agents (urea salicysilic acid and alpha hydroxylic acid) are used to enhance corneocyte disadhesion.[13] Parents should be explained to avoid hot environment as they are deficient in sweating and intolerant to heat, to carry spray water bottles to moisten the skin. A spontaneous healing colloidion baby has been reported in 10 to 25% cases.[14] The skin biopsy picture differs from those evolving into lamellar icthyosis and helps in early diagnosis and prognosis.[14,15] Most cases 75% will go on to develop autosomal recessive congenital icthyosis. Known cases of colloidon baby include icthyosis vulgaris and trichothiodoystrophy, less well documented causes include sjogren -lorsen syndrome, netherton syndrome, gaucher disease type 2, congenital hypothyroidism, conradi syndrome, dorfmann-chanonin syndrome, ketoadipiaciduria, icthyosis variegata and palmoplantar keratoderma with anogenital leukokeratosis.[5] Many of the above conditions are autosomal recessive and inheritance was associated with consanguineous marriage which can be diagnosed at 20 to 22 weeks of gestation by fetal skin biopsy. Transglutaminase-1 (TGM1) is the only gene identified to date to be mutated in patients with lamellar icthyosis (LI). In harlequin icthyosis prenatal diagnosis by fetal skin biopsy and ultrasonography helps for precise diagnosis by taking chorionic villous sampling.[16]

Conclusion

None of the clinical features in a colloidion baby can be used to predict the final diagnosis or prognosis of underlying icthyosis phenotype. Histopathological features of skin biopsy in first few weeks of life will not be helpful in differentiating different types of icthyosis1 but early recognition of this entity and early institution of supportive treatment definitely reduce morbidity and mortality in neonates. A protocol must be established so that appropriate measures can be taken months or years following the shedding of the colloidion membrane.

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