CASE REPORT

Colloidion Baby

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Abstarct

A two day old male baby, third issue of a second degree consanguineously married couple with clinical features of colloidion baby is being presented. Early recognition of this clinical entity and early institution of appropriate therapy can definitely reduce morbidity and mortality in neonates.

Key Words

Colloidion Baby, Neonates, Genetic Disorder

Introduction

Colloidion baby is a characteristic clinical entity which may precede the development of one of a variety of ichthyoses or occur as an isolated and self-limiting condition. The condition is usually a manifestation of congenital ichthyosiform erythroderma (CIE) or lamellar ichthyosis (LI) and is responsible for about 2/3 of colloidion babies. Like a harlequin fetus, a colloidion baby appears to one phenotype for several genotypes. These two lesions may be the first signs of an ichthyosis transmitted as an autosomal dominant trait in about 10% of the cases. Colloidion baby is inherited in an autosomal recessive manner. Sex linked ichthyosis never begins with a colloidion baby syndrome (1-5)

Case Report

We Present a two day old male baby, third issue of a second degree consanguineously married couple, born at home by local traditional birth attendant with uneventful antenatal history presented as abnormal appearance of the baby. There was no such history in previous two siblings. Both the two females are alive and healthy. On examination the baby weighed 2500grams, with a length of 48 cm and had head circumference of 34 cm.The vitals were maintained. There was ectropion, absence of eyelashes and eye brows; mouth was fishlike (eclabium). The skin was semitransparent, parchment like feel with varying degrees of fissures at groin and axilla.The skin at places looked to be yellowish brown glistening and film like membrane resembling scales of fish (fig. 1-3). Nails were hypoplastic and there was joint stiffness. Systemic examination did not reveal anything abnormal. The neonatal reflexes could not be adequately elicited due to eclabium and joint stiffness.The baby was monitored regarding the temperature, hydration status and electrolyte balance. Bland lubricants were applied to the skin. The skin was kept lubricated to keep the peeling skin soft to facilitate desquamation. The baby was managed with prophylactic antibiotics. Adequate nutrition and multivitamin preparations were given to the baby.There was slight improvement symptomatically. The baby was regularly followed up.

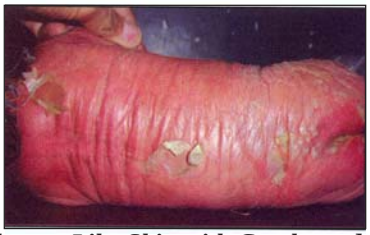

Fig 1 Parchment Like Skin with Cracks and Peels

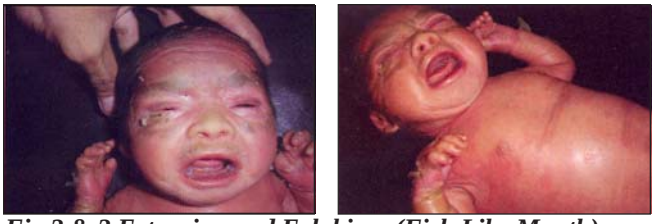

Fig 2 & 3 Ectropion and Eclabium (Fish Like Mouth)

Discussion

Colloidion baby is a common phenotype to several diseases including to variety of icthyoses or as an isolated and self limiting condition. The babies are usually covered at birth by a thick taut membrane resembling oiled parchment or colloidion, which is subsequently shed, often resulting in ectropion and eclabium . Within few hours this membrane dries, cracks and peels off but may reform several times. Infrequently, an affected infant has normal skin after the membrane is shed. Affected neonates have ectropion, flattening of the ears and nose, fixation of the lips in an O- shaped configuration. The case discussed in the text also has similar features. Hair may be absent or may perforate the horny covering. The membrane

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cracks with initial respiratory efforts and shortly after birth, begins to desquamate in large sheets. Complete shedding may take several weeks, and a new membrane may occasionally form in localized areas. The collodion membrane is composed of greatly thickened stratum corneium that has been saturated with water. As the water content evaporates in extrauterine life, large fissures appear in the membrane and the membrane is shed, revealing red skin underneath (1-5).

The taut membrane may impair respiration and sucking, sometimes leaving fissures with impairment of barrier to infection and water loss leading to difficulties in thermal regulation and hypernatremic dehydration. Neonatal morbidity and deaths may be due to cutaneous infection, aspiration (squamous material) and hypothermia. The outcome is uncertain. Treatment with a high - humidity environment and application of non occlusive lubricants may facilitate shedding of membrane. Long baths using bath oils or application of lubricants before drying can prolong the hydration and softening. Keratolyic agents (creams and lotions containing urea, salicylic acid and alpha hydroxyl acids like lactic and glycolic acids) are used to enhance corneoyte disadhesion. Topical retinoids or vitamin D preparations may also be effective but can be irritatig in some patients. Decreased sweating and heat intolerance should be explained to the parents and they should be made aware of signs of heat intolerance such as flushing and lethargy. Avoiding hot environments and carrying spray bottles with water to moisten the skin and cool it through evaporation. A spontaneously healing colloidion baby has been reported in 10-25% cases (6). The skin biopsies from such babies on first and 15th day of life when examined in microscope, features observed were different from those known to occur in colloidion babies evolving into lamellar ichthyosis and may contribute to a more precise, early diagnosis and prognosis of this heterogenous neonatal entity (6, 7). Approximately 1:225 individuals are a carrier. The risk to future pregnancies is 25%. Unaffected siblings of an affected individual have a 2/3 risk of being a disease gene carrier (7). Missense mutations spice -site mutations and deletions occur throughout the gene. Transglutaminase -1 (TGM1) is the only gene identified to date to be mutated in patients with lamellar ichthyosis (LI). Fetal skin biopsy at approximately 20-22 weeks gestation has been accomplished in lamellar ichthyosis , congenital ichthyosiform erythroderma,epidermolytic hyperkeratosis and Sjogren -Larsson syndrome. In Harlequin ichthyosis prenatal diagnosis has been successfully performed by fetal skin biopsy and ultrasonography. This can be done with chorionic villous sampling (8).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. With light microscopic examination of the skin specimens of a newborn baby with a collodion membrane in the early periods an eosinophilic, PAS positive stratum corneum accompanied by hyperkeratosis can be observed. However, the epidermis is weakened due to the thinning of the granular layer. But electron microscopic examination reveals dense intracytoplasmic granules and convoluted corneocytes that can be seen in the upper portion of stratum corneum. Lamellar bodies are numerous but intercellular space and the desmosomes are well preserved. The thinned granular layer is structurally normal. The collodion babies may be born with a collodion membrane covering the entire skin surface just like armor. Particularly, tight membrane on limbs may lead to constriction and loss of function (8,9).The presence of collodion membrane doesn't necessarily predict that the baby will develop ichthyosis and spontaneous healing may occur. Skin biopsy of collodion membrane is usually not diagnostic. Most collodion babies do have a form of ichthyosis and majority of them develop features of lamellars, bullous & X-linked ichthyosis, Netherton's syndrome or Gauchers disease (10).

Conclusion

Early recognition of this clinical entity and early institution of appropriate therapy can definitely reduce morbidity and mortality in neonates.

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