Case Report

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Collodion Baby: A Case Report

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Abstract

Collodion baby is a rare congenital defect of skin characterized by the presence of parchment like taut membrane which breaks on stretching which results in fissure formation. It is a very rare clinical condition, with a worldwide occurrence of 1: 300,000 births; being an uncommon dermatosis. We present a one day old collodion baby, our experience with this case taught us that careful attention to skin care, judicious use of skin care products, and meticulous attention to asepsis are of paramount importance when caring for an infant with collodion baby syndrome.

Keywords: Study; Research; Knowledge.

Introduction

Collodion baby is a rare congenital defect of skin characterized by the presence of parchment like taut membrane which breaks on stretching which results in fissure formation. This condition makes the newborn susceptible to dehydration, electrolyte imbalance, sepsis and temperature instability which invariably causes frequent complications and high mortality. Here, we present an newborn baby born with this condition and presented with severe asphyxia, shock and respiratory failure at birth which was successfully managed in a low resource setting with interdepartmental co-ordination and without the above-mentioned complication.

It is a very rare clinical condition, with a worldwide occurrence of 1:300,000 births; being an uncommon dermatosis. After the introduction of the term "collodion", approximately 270 cases of collodion baby have been reported in medical literature until now.

Alterations in cornification that is, ichthyosis are a prominent group of hereditary diseases, which are characterized clinically by desquamation patterns and histopathologically, evidence of hyperkeratosis is seen. In most cases, there is history of the disease in family or history of consanguinity is present. That's why it may present with acquired disorders. They are distinguished by their mode of inheritance, clinical characteristics, associated anomalies and histological findings.[1-6] In neonates, we commonly observe that the collodion are normally born premature, covered in a layer that appears similar to cellophane, most often in association with ectropion and eclabium accompanied by trichothiodystrophy. The collodion membrane tears off between the period of first and 4th week of life which itself is evidencing the definitive phenotype. 60% of the cases correspond to the initial expression of Congenital

Ichthyosiform Erythroderma or Lamellar Ichthyosis, although 10% of cases will have normal skin after resolution of the parchment like collodion membrane.[3,4] Other presentations of disease may also include Ichthyosis Harlequin which is linked to Chromosome X, Erythroderma Ichthyosiform, congenital blister and Nethertan Syndrome.

The important clinical presentations are ectropion (eversion of the eyelids), eclabium (everted lips) and hypoplasia of nasal bones, auricular pavilions, fingers and toes. Dermatological manifestations often exaggerate in cold or dry environments. They often present at birth with severe erythroderma, slight desquamation and bulla or large erosions that are seen mainly present over the trunk and limbs, which evolve to extensive denuded, moist, painful and foul smelling areas.

Case Report

A preterm male newborn at gestational age of 34 weeks, son of a 19-year-old mother who did not attended single prenatal check-ups at the health center, and an obstetrical ultrasound was not performed. As far because the personal history of the oldsters they didn't report exposure to toxic substances or X-rays. This is the mother´s first pregnancy. She began with active labor at the 34 weeks of gestation according to the date of last menstruation. The newborn had an Apgar score of 6 and 8 at the first and fifth minutes, weight 1800 g, cephalic perimeter 31 cm, length 46 cm, chest perimeter 30 cm, heart rate 157 beats per minute, respiratory rate 56 breaths per minute, Silverman Anderson score 1 (barely visible intercostal retractions).

On physical examination, a constrictive, tense membrane resembling an oily parchment or collodion covering the whole body surface was found at birth (Figure 1).

Fig. 1 Ectropion and Eclabium.

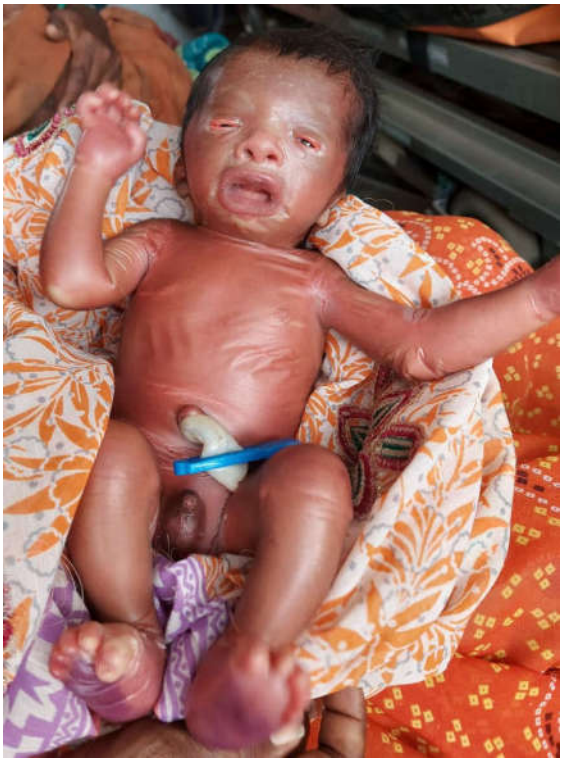

Fig. 2: Parchment like skin (collodion).

Discussion

The first description of Ichthyosis was made by Rev Oliver Hart in Charleston, South Carolina in 1750. It gets its name from the texture of the skin presented by newborns. It is very well established that they are a group of congenital disorders with cutaneous manifestations, which may or may not be linked with other genetic defects their characteristics features are the alterations of the skin, which are relevant for the recognition of the pathological basis and its appropriate approach. Sepsis is its most severe manifestation, concomitantly severe dehydration secondary to trans-epidermal water loss, which leads the patient to an electrolyte imbalance.[7]

Collodion baby is an underreported condition. According to Chung et al, about 270 cases were reported in the literature since 1892 till now[8]. These babies are usually born preterm. The parchment-like membrane undergoes desquamation with time. Once this layer peels off, underlying skin disease is typically evidenced. At least one-tenth of these patients develops normal underlying skin afterwards. This is one way of disease progression for this heterogeneous disorder and thus can be called 'self-healing' collodion baby. Another way of progression is keratinizing disorder presenting congenitally is Harlequin fetus and these babies have horny plates all over the body. Most of these babies die within first few days to weeks of life due to respiratory difficulty, inability to feed, and severe skin infections. Patients who have lived after infancy have severe ichthyosis and commonly have variable neurologic disabilities. In all cases, barrier function of the skin is hampered once collodion dries up and starts cracking. This leaves the baby vulnerable to dehydration. Thick

membrane can create problems such as infection, temperature instability, and excessive transcutaneous fluid losses which leads to restricted movement leading to difficulties with breast-feeding. If chest wall movements are restricted, it can cause hypoventilation. Inability to shut eyelids may cause corneal damage if proper precautions aren't followed. With advancements in intensive neonatal care the mortality rate in contemporary era has been decreased to 11 %. Mortality rates are high in patients with associated erthroderma and common causes include hypothermia, renal failure, dehydration and electrolyte imbalance, constricting bands of extremities which leads to vascular compromise and edema[9]. Collodion membrane can lead to decrease in breathing movements which can cause respiratory distress and desaturation. Nasal obstruction can cause difficulty in breathing, often necessitating probing.[10] Collodion membrane may result in difficult swallowing, and restriction of joint movements.[10,11] With ectropion are at very high risk of developing keratitis but expert and timely ophthalmic management can prevent the complications[12,13]

Primary pathology in ichthyosis is altered cornification which in turn results in formation of collodion membrane. Two most common types of dermatological disorders presenting as collodion congenitally are non-bullous congenital erythroderma and Lamellar ichthyosis. Other less frequent pathologies which results in this presentation include: Ectodermal dysplasia, Sjögren Larsen syndrome, Comel-Netherton syndrome Gaucher Disease type 2, Hay-Well syndrome, Trichothyodystrophy, Neutral lipid storage disease, etc.[14] Genetically, it is a heterogeneous disorder which usually follows autosomal recessive transmission. Until now, six genes have been linked with mutations with transglutaminase 1 (TGM1) accounting for approximately 40% cases. TGM1 mediates cross-links in the formation of the cell envelope during terminal differentiation of in genes involved in nucleotide excision repair pathway has also been implicated.[17] Management requires the expertise of a dermatologist and therefore the pediatric team. The baby is initially kept during a humidified, neutral temperature environment like an incubator. Other supportive treatments such as intravenous fluid and tube feeding is ensured to maintain hydration and nutrition. One should keep the skin soft and also plan to reduce scaling. The collodion membrane must not be debrided. Some clinicians find use of emollients helpful, although Van Gysel et al. in their case series of 17 babies with similar problem found emollient application to be associated with increased the risk of infection[18]. There is risk of intoxication by cutaneous mode of absorption of topical products and Yamamura et al. have reported a case of salicylate toxicity due to use of keratolytics[19]. Hence, the emollient used should be inert-like petroleum jelly. Cakmak et al. found acitretin in dose of 1 mg/kg/day to be effective and safe in management of congenital ichthyosis[20]. Artificial tears could also be necessary if there's severe ectropion. Treatment can also include analgesic and mild topical steroids to scale back secondary inflammation. The management of the case relies on the timely identification and early institution of medical care therapy. Intensive care therapy includes maintaining adequate oxygen saturation, intensive skin care, thermoregulation of baby and adequate feeding. Molecular diagnosis was not possible because of the cost involved. In our case, a diagnosis was made at the time of birth and quality intensive care was instituted immediately. The baby was monitored for electrolyte abnormality and judicious fluids were given to prevent dehydration. We initiated feeds early so as to prevent infection related complications. The baby improved gradually with supportive care and was discharged without complications.

Conclusion

Collodion baby probably is underreported condition with cases which have mild presentation not getting reported. It requires early diagnosis and multidisciplinary approach to prevent mortality. Our experience with this case taught us that careful attention to skin care, judicious use of skin care products, and meticulous attention to asepsis are of paramount importance when caring for an infant with collodion baby syndrome. Because of rarity and heterogeneous nature of this disease, experience of individual clinician could also be limited; hence, a protocol should be established to manage the dynamic nature of this condition.

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