Case Report

A rare case report on colloidion baby syndrome with severe ectropion in neonates

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Abstract

Colloidion baby is a name given to baby born yellow, scaly wax like covering the baby. It is also known as lamellar icthyosis and is an inherited disorder present through birth. The disorder is a social stigma to the society. As the child ages, the hyperkeratosis can alter with normal body perspiration mechanismand lead to heat intolerance and possible heat shock.

Keywords: Lamellar ichthyosis, Ectropion, Eclabium.

Introduction

The first clinical description of collodion membrane was given by Perez [1] in 1880. The baby's skin is replaced by excessive scaling of the body present through birth giving a wax like appearance.

Ichthyosis is an infrequent clinical entity worldwide (1:300,000 births) [2] in newborns. The clinical manifestations are thick broad scales over whole body and bright red erythematous skin at birth. Some of the severely affected infants may appear like colloidion baby as if a thick cellotape has been firmly applied over the whole skin. The skin is so tightly stretched that it can cause respiratory distress with eversion of the eyelids (ectropion) and lips (eclabium). The split scales and waxy appearance are more common in flexor surface of the body [2].

Case Report

A day 3 day old female outborn term vaginal delivery by "dai" and cried immediately after birth was referred to Subharti Medical college casualty department of pediatric with complain of peeling of skin, some abnormality in both eyes and difficulty in accepting feed. According to parents the child had a whitish covering over his body. There was no similar history in the family. The marriage was non consanguineous and first birth order. The child was consulted in Ophthalmology OPD for ocular examination. On examination the child was having a covering over his whole body, with scaly skin and peeling more on the nose area, joint area and flexor surface of the body. The child was having severe ectropion in both eyes and mouth was fish type (eclabium). The child was also having temperature (100.4 F). After Ophthalmology consultation the child was diagnosed having severe ectropion with no other complications. Fundus examination of the child was normal. Cornea was not involved and pupil was of normal size and normal reaction. The child was admitted in NICU and started managmement conservatively. The child was given lubricating eye drops and topical antibiotics from ophthalmology side. We started on IV fluids, antibiotics and antipyretics (as child was having temperature) as per NICU protocols. Emollients were also applied. Strict temperature and vitals charting was maintained. Septic screen and other required investigations were sent. The child was kept in close observation for 10 days. After 10 days child was referred to ophthalmology side for management of severe ectropion. We wanted to evaluate the child after 7 days but we lost follow up as attendant took baby against medical advice.

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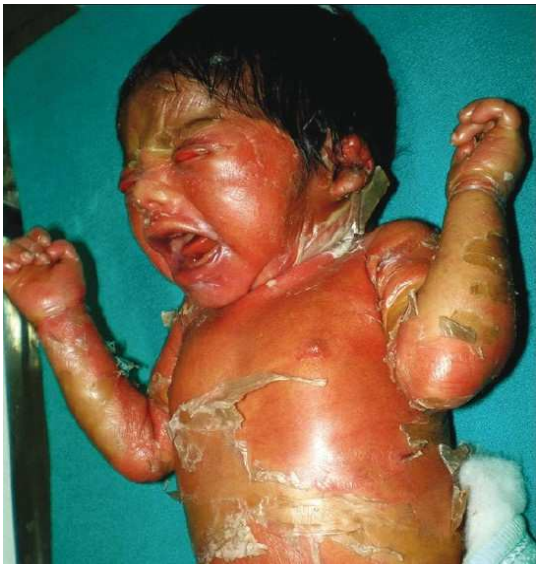

Fig 1: Showing Colloidion baby with severe ectropion and eclabium

Discussion

The term collodion baby includes all newborns born with an extra layer of skin. It is a descriptive term, not a specific diagnosis or disorder (as such, it is a syndrome) [3]. This is similar to our case in which there has an extra layer of skin. The collodion membrane is peeled off or shed 2 - 4 weeks after birth, revealing the underlying skin disorder [4]. The cause of collodion baby syndrome is not well known but mutation in the gene tranglutaminase 1 is considered [5]. There are four forms of ichthyosis, that is, ichthyosis vulgaris, X linked ichthyosis, lamellar ichthyosis and epidermolytic hyperkeratosis. Rarer forms of congenital ichthyosis including trichothiodystrophy, Gauchers disease, neutral lipid storage disease, Conradi-Hunermann disease, Sjogren- Larsson syndrome, and isolated palmoplantar keratoderma may be preceded by a collodion membrane. The collodion baby are often premature and at risk of losing heat due to abnormality of the skin. Therefore various steps to be taken to prevent heat loss (hypothermia, dehydrtaion, and keratitis) such as application of emollients, maintaining room temperature and lubricating eye drops. Despite this, systemic infections were reported [2]. In our case child developed septicemia so antibiotics were upgraded accordingly and managed conservatively. During the hospitalization period the child was active and calm so we considered the condition may be painless.

Conclusion

The diagnosis of the colloidion baby is clinical and management is conservative. Detailed history of the patient should be obtained for better management. Colloidion baby is susceptible to infections so care must be taken and minimal handling should be done. NG feed was given in our case to prevent aspiration. Along with NG feed, IV fluids, proper antibiotics, were given. One should not hesitate in upgrading the antibiotics depending on septic screening. Management of colloidion baby requires a team comprising of pediatrician, ophthalmologist and dermatologist.

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Refernces

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