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Case Report

Collodion Baby - A Rare Clinical Entity

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ABSTRACT

AIM OF THE STUDY:-To report a rare case of dermatological Anomaly that came across in an outpatient department of skin-.Namely –Collodion baby.
PLACE AND PERIOD OF STUDY:-In outpatient Department of Dermatology in the Hospital of Kolar Districts during the year 2007 to 2010.
MATERIAL AND METHODS: - A case of Collodoin baby constituted the material for the present study. This case was examined in detail with .proper antenatal history of mother was taken. There was no history of drug intake during first trimester. No exposure to Radiation, or similar episodes in the past. She was hypertensive but no other systemic diseases.
OBSERVATION:-Newly born child was male weighing 3.0 kg only was covered by transparent tough skin all over the body .At some places skin had peeled off exposing raw surface of skin. There was eclabium, with ectropion, but skin did not show any cracks and fissures like Harlequin baby.
CONCLUSION:-This rare genetic disorder has caused high rate of morbidity and mortality rate. So, complete knowledge and awareness of disease as well as its prevention has to be given to patients. Since it has profound genetic importance, hence it was studied and reported
Keywords:-Lamellar Icthyosis, Collodion baby, Ectropion, Eclabion, Self healing, Collodion membrane.

INTRODUCTION

Collodion baby is rare clinical entity seen in new born where children are born with translucent, parchment and tight paper like skin called collodion membranes which are tightly covering all over the body. [(1, 2, 3)] It was Hallopeau who used term collodion baby in 1884. [(3, 4, 5)] In the world since then approximately 270 cases have been reported. [(2, 4)]

CASE REPORT

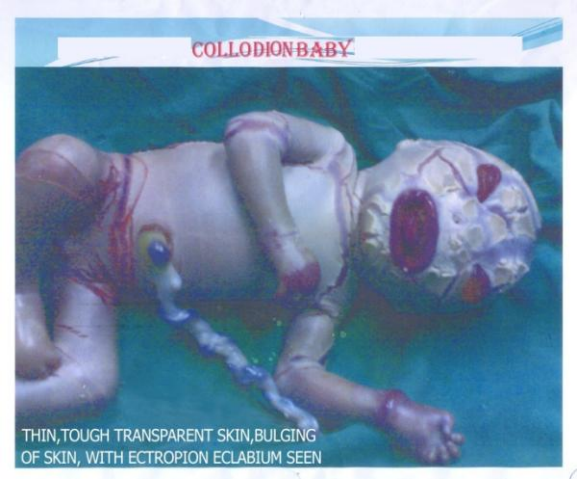
A new born male child was found to have transparent thin tough skin covering all over the body in the Hospital of Kolar district Karnataka. At some place skin had peeled off while in most of the places it was tightly adherent to the body. The baby was cried in shrill voice and had difficulty in breathing due transparent tough skin encircling the body.

Family History

Showed history of anomaly other than skin condition. There was no history of drug intake history or exposure to radiation during first trimester of pregnancy. She was hypertensive but not diabetic. No other Anomalies were found in the body.

On Examination of the Child

The new born child was undernourished. There was transparent skin covering all over the body. At places skin had peeled off near the joints and there were bulging of transparent skin. In the most of the places, the skin was adherent to the body. There was fish like mouth called Eclabium with bulging of eyes – Exopthalmos (like frog eyes). Since the baby was unable to suck the breast milk, he was kept in the incubator in neonatal ICU for a day and milk was fed through riles tube every 4 hours. Mother refused to see the child and give the breast milk even when child cried in shrill voice. The next day, child expired due to respiratory failure.



Collodion Baby

Photograph showing a newly born baby covered by thin transparent tough skin with intact long umbilical cord. At some places it is torn and bulged exposing raw surfaces especially at joints.Eclabium (Oshaped mouth)and ectropion(evertion of upper eye lid) are also seen in the child.

DISCUSSION

Collodion baby is an autosomal recessive ichthyosiform disease where there is formation of collodion membrane. This collodion membrane thin transparent tight paper like skin sheets encircling the entire body surface. [1, 2, 3, 9]

Causes: The causes of congenital both autosomal Recessive Lamellar Icthyosis & congenital icthyosiform erythroderma have been reported to be due to mutation of Transglutaminase 1gene localised on 14 q11 (Shwayder & Jeon et al). [4, 14]

When the collodion membrane peels off in 2-3 weeks time, then fissures of skin are exposed giving rise to serious complications like infection, loss of fluid, malfunctioning of the temperature, and electrolyte imbalance. Such babies are born premature. [2, 6] A study on 10 (TEN) harlequin babies, has revealed that the pathology of disease is due to default in keratin, flaggerin and the lamellar body both structurally and functionally. [3, 7] Another study has been done which revealed that deficiency of serin treonin protein phosphsatase enzyme related to protein phosphatise gene mutation which is present on 11th chromosome is said to be the another cause of this clinical entity. [3, 8] Seventeen collodion babies have been reported. Out of them, 7 (41%) were congenital Icthyosiform erythroderma. Three cases (18%) were Lamellar Icthyosis. One was Sjogren-Larsson Syndrome, another case was epidermolytic hyperkeratosis and other case was Gaucher disease. While rest of patients (4. Cases ... 24%) did show collodion disease. [9] Collodion baby has been reported in a one month old female child, with tight cracked and hard skin. The child had ectropion, o shaped lip, flattening of nose and ears. The weight of the baby was just 3.5kg.There was sparse distribution of hairs over the scalp, associated bilateral ectropion. Baby was born after full term normal

delivery and daughter of nonconsanguinous couples. [10] Ectropion if not treated may lead to dryness of eyes, known as Xerophthalmia followed by keratitis and finally death. [4] Digovanna and Bale have classified Congenital Bullous icthyosiform erythroderma in to six groups. Out of them three showed palmoplantar involvement while other three did show palmoplantar involvement. [11] Localised lesions in collodion babies have been treated by retinoic acid and calpotriol which are supposed to be a succesful treatment. [12, 13] There are variations in the mechanism of molecular pathogenesis and localizations of 5 different genes and detections of mutations of 50 genes in these genes. [15, 16]

Present study: In the present study, newly born male baby showed thin transparent tough (3.T) skin covering all over the body. At some places skin had peeled off and bulged especially at joints. There were raw surfaces over the back and were infected. There was evertion of the eye lid and fish like mouth known as Eclabium. Baby had thick hairs and no flattening of nose. Baby did not have any palmar plantar involvement.

CONCLUSION

These icthyosiform diseases which are of rare anomalies which have high rate of morbidity and mortality. In present days introduction of systemic retinoids as well as by best nursing care in intensive care units, has brought down mortality rate drastically (as cited by Yalquin). Awareness to be given to the general public regarding consanguineous marriages which should totally avoided. Hence studied and reported.

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