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Case Report

COLLODION BABY WITH CONGENITAL BILATERAL ECTROPION AND ECLABION- RARE PRESENTATION

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ABSTRACT

Congenital Ichthyosiform Erythroderma- an extremely rare dermatological condition with an estimated incidence of 1 in 50,000 to 100,000 birth. There is defective Stratum corneum barrier associated with collodion membrane when the baby moves from an amniotic fluid to external dry environment during parturition. The membrane dessicates and peels off. This condition is due to mutations in Transglutaminase1, ALOX genes, ABCA12, Ichthyin, ABHD5.

KEY WORDS: Ichthyosiform Erythroderma, Dermatological, Transglutaminase1, ALOX genes, ABCA12, Ichthyin, ABHD5.

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INTRODUCTION

Collodion baby is an old term describing transient appearance of neonate at birth which subsequently develops Congenital Ichthyosiform Erythroderma.

Etiopathogenesis: At birth, due to defective Stratum corneum barrier.

Mutations in Transglutaminase1, ALOX genes, ABCA12, Ichthyin, ABHD5.

Types: Self healing and Ichthyosis form.

CASE REPORT

One day old female child referred from paediatric ward as a case of Congenital Ichthyosis-nursed in neonatal ICU.

Born by normal delivery of non-consanguine ousmarriage with birth weight of 1.8Kg.

General examination-glistening taut plastic skin like collodion membrane stretched all over the body,Eclabion (everted lips) with Fish-mouth appearance, flattened pinnae , Ectropion of both the lids.

Fig. 1: Collodion baby.

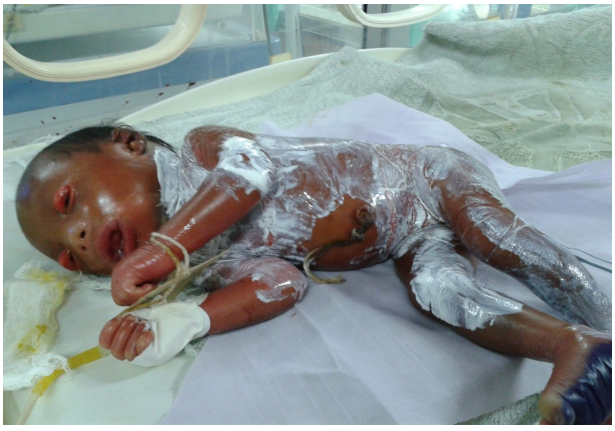

Fig. 2: Ectropion with Eclabion.

DISCUSSION

Hallopeau and Watelet were the first who gave the term Collodion baby (CB) [1]. The skin of the newborn is replaced by a cornified substance, which gives the body a parchment like appearance or a varnished appearance [2]. This condition is inherited primarily as autosomal recessive ichthyosis either LI or NBCIE. CB is an extremely rare dermatological condition with an estimated incidence of 1 in 50,000 to 100,000 birth [3]. A new form of the disease has been notified as 'self-healing collodion syndrome' in these cases newborn completely recovers within few months after birth [4].

Collodion baby is described as a congenital condition characterized by presence of parchment or cellophane like membrane encompassing the whole body. This is secondary to disorder of cornification. These are usually prematurely born and diagnosed at the time of birth only. Due to presence of tight membrane these babies develop many complications like ectropion, eclabium, restricted extremities and digits movements due to pseudocontractures, absence of eyebrows, sparse hairs on head, deformed nose and ears due to hypoplasia of nasal and ear cartilage. The babies has poor sucking, distal limb ischemia and oedema of extremities.

Commonly caused by various genetic mutations including TGM1(commonest), NIPAL4, ALOX12B, ALOXE3, ABCA12, CYP4F22, PNPLA1 (OMIM 615024), (17) and CERS3 (OMIM 615023). TGM1 is located on chromosome 14q11.2 and has 15 exons (Genbank NM- 000359.2) [9].

The first line of management is moisturizers and topical keratolytic agents, they enhances skin barrier and facilitate desquamation. Sodium chloride, urea, vitamin E acetate, glycerol and petroleum jelly are various agents available as moisturizers and lubricants. These infants are at increased risk of intoxication by absorption of topical products, like salicylates or keratolytics due to impaired skin. In severe cases with marked hyperkeratosis keratolytical agents like lactic acid, glycolic acid, salicyclic acid, N- acetyl- cystine, glycol can be used. Ectropion is managed by application of artificial tears and eye lubricants. In severe cases surgical correction is done. Administration of retinoids have keratolytic effect and help in elimination of scales and prevent hyperkeratosis of skin. Isotretinoin and retinoids both have proven to be effective in these cases.

CONCLUSION

Collodion syndrome is a rare disease so it is essential to have a protocol for the treatment of these patients, the instructions to follow in treatment and proper management of the complications that can arise. In long-term course the etiological cause of the disease should be established so that appropriate measures can be given to the patient.

Conflicts of Interests: **None**

REFERENCES

[1]. an Gysel D, Lijnen RLP, Moekti SS, De Laat PCJ, Oranje AP. Collodion baby: a follow-up study of 17 cases. Journal of the European Academy of Dermatology and Venereology. 2002;16(5):472–75.

[2]. Cortina WJ, Cruz MJ, Villalobos OA, Espinoza MA. Ictiosis congenital (fetoarlequín). Bol Med Hosp Infant Mex. 1975;32:699-702.

[3]. Dyer JA, Sparker M, William M. Care of newborn with ichthyosis. Dermatol Ther. 2013;26(1):1-15.

[4]. Shwayder T, Akland T. Neonatal skin barrier: structure, function and disorders. Dermatol Therapy. 2005;18:87-103.

[5]. Srivastava P, Srivastava A, Srivastava P, Betigeri AV, Verma M. Congenital Ichthyosis-Collodion Baby Case Report. Journal of clinical and diagnostic research: JCDR. 2016 Jun;10(6):SJ01.

[6]. DiGiovanna Robinson-Bostom L. Ichthyosis: etiology, diagnosis, and management. Am J Clin Dermatol. 2003;4(2):81-95.

[7]. Offiah M, Brodell RT, Campbell LR, Wyatt JP. Collodion-like membrane in a newborn exposed to infliximab. J Am Acad Dermatol. 2014;71(1):e22-23.

[8]. Dermatology at the Millennium. By Delwyn Dyall-Smith, Robin Marks, Page 586, Published by Informa Health Care, 1999,

[9]. Moslehi R, Signore C, Tamura D, Mill JL, DiGiovanna JJ, Tucker MA, et al. Adverse effects of trichothiodystrophy DNA repair and transcription gene disorder on human fetal development. Clin Genet. 2010;77:365–73.

[10]. Fachal L, Rodriguez-Pazos L, Ginarte M, et al. Charaterization of TGM1 c.984+1G>A mutation identified in a homozygous carrier of Lamellar ichthyosis. Int J Dermatol. 2012;51(4):427-30.

[11]. Taïeb A, Labrèze C. Collodion baby: what's new. J Eur Acad Dermatol Venereol. 2002;16(5):436–37.

[12]. Schmuth M, Martin V, Janecke AR, et al. Inherited ichthyosis/generalized Mendelian disorders of cornification. Eur J Hum Genet. 2013;21(2):123-33.

[13]. Rubio- Gomez GA, et al. Development of a disease severity score for newborns with collodion membrane. J Am Acad Dermatol. 2014;70(3):506-11.

[14]. Roberts JB, Adelson D. Case report: prolonged collodion membrane causing constrictive bands of the digits and treatment. Dermatology Online Journal. 2010;16(1):15.

[15]. Harvey HB, Shaw MG, Morrell DS. Perinatal management of harlequin ichthyosis and literature review. J Perinatol. 2010;30(1):66-72.

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