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Research Article

SELF HEALING COLLODION BABY: A RARE PRESENTATION

Prashant Abusaria*, Shital Kolhe, Swati Singh and Chinmay Choudhari

Department of Pediatrics, D.Y.Pati Medical College, Navi Mumbai, Maharashtra, India

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ABSTRACT

Collodion baby is phenotype resulting from different congenital disorders of keratinization characterized by the presence of a tight, translucent membrane that covers the entire skin at birth. It is a congenital disorder, occurring with an incidence of 1: 300,000 live birth and both gender are equally affected. Collodion baby is a descriptive term used for transient appearance of Neonates born with some form of congenital Ichthyosis . This membrane usually sheds around 10 to 14 days and reveals the underlying disease. It is associated with a mortality rate of approximately 11%. A rare variant of this phenotype (10% to 20%) clear spontaneously or later show only very mild ichthyosis, being referred to as self-healing collodion baby (SHCB).[1,2,7,9.] The condition resolved spontaneously within a few weeks and patient does not developed any other manifestation. Although no molecular analysis was performed, we contribute to the knowledge of the clinical features of this extremely uncommon and benign entity.

INTRODUCTION

Congenital Ichthyosis also known as Collodion baby is an uncommon clinical condition, with a worldwide occurrence of 1: 300,000 births. The exact cause of the Collodion Baby syndrome is not known but in most of the cases autosomal recessive inheritance pattern is seen and they are very rare and may be associated with consanguinity.[1,4] Alterations incornification (ichthyosis) are a prominent group of hereditary diseases presented clinically by desquamation patterns and histopathologically by hyperkeratosis. Family history of similar illness is usually present. The appearance is often described as shiny film looking like a layer of cellophane. The eyelids and mouth may have the appearance of being forced open due to the tightness of the skin. Collodion babies are normally bornpremature, in association with ectropion (eversion of the eyelids) and eclabium (everted lips).[7] The collodion membrane breaks off between the 2nd to 3rd week of life. 75% of the cases correspond to the initial expressionof Congenital Ichthyosiform Erythroderma or Lamellar Ichthyosis. 15% cases association with various entities is seen like ichthyosis vulgaris, trichothidystrophy, metabolic and endocrinal disorders which involve keratinization disorders. In just 10% of these cases the membrane sheds off and underlying skin is normal for rest of the life, notified as 'Self-Healing Collodion Baby syndrome'.[2,7,8.]

Prenatal diagnosis can be done by amniocentesis, villus biopsy, fetal skin biopsy, blood or urine analytical studies if there is a family history with a genetic pattern that makes transmission possible.[2]

The treatment consists of maintaining the body, skin care and above all pain treatment. Incubator with humidifier, hydration and electrolyte balance control. Topical application of urea, alphahydroxy acids, lactic acid and propylene glycol, vegetableoils such as petroleum jelly and topical retinoid may be used as supportive treatment. pyoderma, sepsis from respiratory origin or due to an interventional approach. uncontrollable hypothermia, hypernatremia dehydration due to excessive evaporation and transepidermal loss of water should be prevented. Also, these patients might develop atopic dermatitis, asthma and allergic rhinitis, as the alteration of the skin barrier predisposes them to a greater allergic sensitization.

CASE REPORT

A full term female baby born to a 22-year-old primigravida mother.[fig-1] Mode of delivery was spontanous vaginal. No abnormality were detected in ante-natal scans. No history of consanguinity was present. There was no history of similar condition in the family. The baby's birth weight was 2.8kg with normal APGAR score at time of birth. On physical examination, a tense membrane resembling an oily parchment or collodion was present, covering the whole body surface.

Corresponding author:* **Prashant Abusaria
Department of Pediatrics, D.Y.Pati Medical College, Navi Mumbai, Maharashtra, India

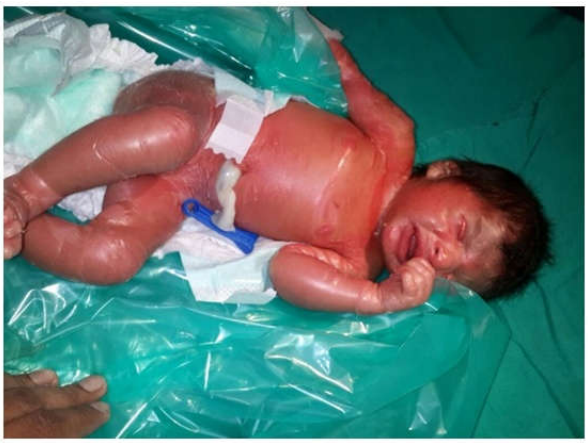

Fig 1

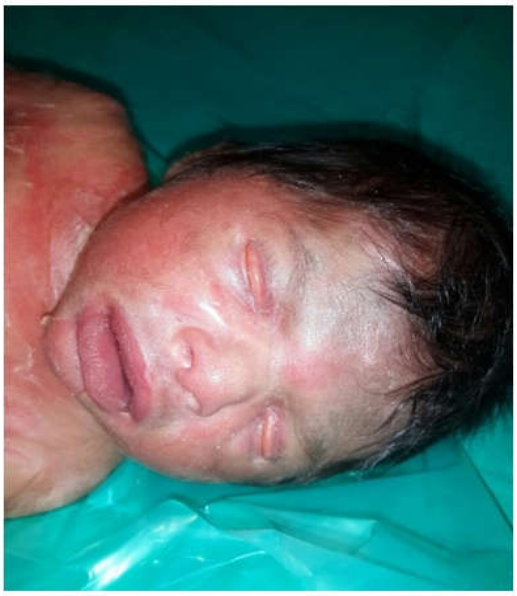

Fig 2

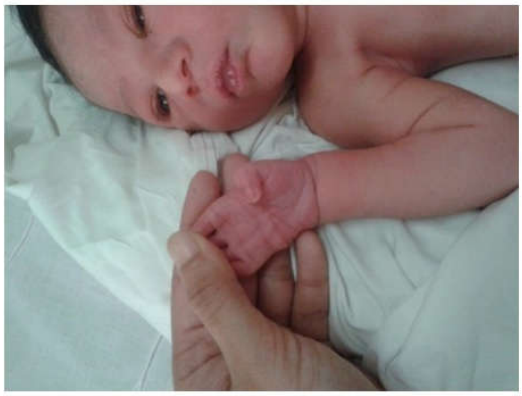

Fig 3

Ectropion (eversion of the eyelids) and eclabium (everted lips) was present. Fissured skin was present on neck, trunk and limbs with flat pinna, effacement of normal skin creases and sausage shaped digits. Systemic examination was normal. The patient was admitted in neonatal intensive care unit. On blood investigation CBC was with in normal range with leukocytes 156 × 10^9/L, neutrophils 54.3%and lymphocytes 37.5%, haemoglobin 13.4 mg/dl, haematocrit 40.5% and platelets 262,000. Patient was stared on broad spectrum antibiotics, injectable Cefotaxim and amikacin. Intravenous hydration was maintained in radiant heat warmer cradle at a suitable temperature in a sterile environment to avoid hypothermia and infections. Topical application of paraffin oil was done as demulsent.[2,7] Patient was hemodynamically stable hence was discharge and was asked to follow up after 2 weeks. At follow up partial resoloution of skin was seen and after 4 weeks complete resolution of skin was there.

DISCUSSION

Collodion baby is a congenital condition characterized by presence of parchment or cellophane like membrane encompassing the whole body. Collodion membrane is a result of disturbed epidermal lipid and protein homeostasis.[6,8] The exact cause of collodion baby syndrome is not known but most of collodion babies have autosomal recessive congenital ichthyosiform erythrodermas due to functional mutations in transglutaminase 1 - TGM1, ALOXE3 or ALOX12B genes, ABCA12, HIPAL4/ ichthyin**,** ABHD5 or other genes .Only 10% of cases eventually develop normal skin, known as self-healing collodion baby.[4] Self-healing collodion baby is caused by mutation of transglutaminase 1 inactive *cis* form; after delivery, a normal phenotype develops, since in the extrauterine environment the enzyme isomerizes back to its active *trans* form.

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 have poor sucking, distal limb ischemia and oedema of extremities. Management requires combined effort of dermatologist, neonatologist and in some cases ophthalmologist and ENT specialist. The aim of treatment is to remove the scaly skin and reduce the excessive irritation.[2,3,10] The parents should be counselled and regular follow-up of the patient is needed.

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