

Case Report

Rare Case Report- Neonatal Lamellar Ichthyosis in Newborn - Collodian Baby

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Abstract

Lamellar ichthyosis, also known as ichthyosis lammellaris and non-bullous congenital ichthyosis, is a rare inherited skin disorder, affecting around 1 in 600,000 people. Affected babies are born in a collodion membrane, a shiny waxy outer layer to the skin. This is shed 10–14 days after birth, revealing the main symptom of the disease, extensive scaling of the skin caused by hyperkeratosis. With increasing age, the scaling tends to be concentrated around joints in areas such as the groin, the armpits, the inside of the elbow and the neck. The scales often cover the skin and may resemble fish scales. In medicine, the term collodion baby applies to newborns that appear to have an extra layer of skin (known as a *collodion membrane*) that has a collodion-like quality. It is a descriptive term, not a specific diagnosis or disorder (as such, it is a syndrome). Clinically, the collodion babies may encounter dehydration, electrolyte imbalance, temperature malfunction and increasing sepsis risk because of relatively severe skin damage. Therefore, morbidity and mortality rates are fairly high in these cases. Conclusively, these newborns should be monitored carefully in intense care units and appropriate and supportive treatment must be undertaken.

Keywords: Lamellar ichthyosis, Newborn, Collodian Baby

1. Introduction

Lamellar ichthyosis, also known as ichthyosis lammellaris and non-bullous congenital ichthyosis, is a rare inherited skin disorder, affecting around 1 in 600,000 people. Affected babies are born in a collodion membrane, a shiny waxy outer layer to the skin. This is shed 10–14 days after birth, revealing the main symptom of the disease, extensive scaling of the skin caused by hyperkeratosis. With increasing age, the scaling tends to be concentrated around joints in areas such as the groin, the armpits, the inside of the elbow and the neck. The scales often cover the skin and may resemble fish scales. In medicine, the term collodion baby applies to newborns that appear to have an extra layer of skin (known as a *collodion membrane*) that has a collodion-like quality. It is a descriptive term, not a specific diagnosis or disorder (as such, it is a syndrome).¹ Clinically, the collodion babies may encounter dehydration, electrolyte imbalance, temperature malfunction and increasing sepsis risk because of a relatively severe skin damage. Therefore, morbidity and mortality rates are fairly high in these cases. Conclusively, these newborns should be monitored carefully in intense care units and appropriate and supportive treatment must be undertaken.²

2. Case Report

Twenty three year un-booked Primigravida, labourer by occupation, presented with history of eight months amenorrhoea and presenting complaints of per vaginal watery discharge since twelve hours and labour pains since five hours before admission. She was not sure about her date of last menstrual cycle. She was married two years back. There was no consanguinity. She had not received antenatal care, haematinics and tetanus toxoid immunization. Obstetric ultrasound was not done during antenatal period. There was no significant past medical or surgical or family history.

General examination revealed mild degree of anaemia. Obstetric examination revealed that the baby was average in size and in cephalic presentation. She was in latent phase of labour. Labour augmentation was done with oxytocin infusion. She had normal vaginal delivery four hours after admission.

Patient delivered a male baby with birth weight of 2098 grams. Baby was appearing to be covered with white parchment. Apgar score at birth was 6, 7 and 8 at 1, 5 and 10 minutes. Baby had features of rare congenital genetic disorder i.e Lamellar Ichthyosis- Collodian baby. (Fig.1 and 2) Baby had parchment paper like skin all over the body, fish like mouth and ectropion of both eyes. Baby was managed by paediatrician in neonatal nursery care unit (NICU)

Investigations of baby revealed normal haematological picture, (Hb-15.8g/dl, Total leucocyte count of 7500/cubic ml, platelet count was 2.55 lacs, Packed cell volume -48.2%, Peripheral blood smear revealed anisocytosis, poikilocytosis and predominantly macrocytes. Renal and liver function tests were within normal limits. Test for C reactive proteins was negative. X ray chest revealed no abnormality. Dermatologists were consulted for skin condition. Diagnosis of Collodian baby was confirmed. Baby was kept in humidifier, Baby was kept in NICU. Baby was given naso-gastric feed, intravenous fluids, Intravenous antibiotics (Ampicillin, Gentamycin, Cephalexin).

Parchment like layer started peeling from second day of birth, exposing the skin underneath. Baby started showing signs of sepsis from third day. Baby's condition deteriorated fast over three day's time. Parents of the baby were counselled about the condition of the baby, possible complications and long term prognosis. For financial reasons relatives were not willing to take the baby to higher centre having better NICU facilities. Baby developed frank septicaemia and disseminated intravascular coagulation and died on fourth day of birth.

Fig.1- Showing fish mouth appearance, ectropion of lids and peeling of skin in neck region



Fig. 2- Showing ectropion of lids and skin changes over abdomen and lower limbs



3. Discussion

Lamellar ichthyosis is an autosomal recessive genetic disorder, which means the defective gene is located on an autosome, and both parents must carry one copy of the defective gene in order to have a child born with the disorder. Carriers of a recessive gene usually do not show any signs or symptoms of the disorder.³

Collodion baby as a term was first used by *Hallopeau* in 1884^{4,5,6}. It was used for newborns in which all the body surface is covered by thick skin sheets, so called "collodion membrane". The collodion membrane is the result of an epidermal developmental dysfunction. The collodion membrane is composed of thick skin sheets which resemble translucent, tight parchment paper. The skin of a Collodian baby has a shiny film that looks like a layer of Vaseline. The eyelids and mouth may have the appearance of being forced open due to the tightness of the skin. There can be associated eversion of the eyelids (ectropion). Collodion baby can have severe medical consequences, mainly because the baby can lose heat and fluid through the abnormal skin. This can lead to hypothermia and dehydration.⁷ Strategies to prevent these problems are the use of emollients or nursing the baby in a humidified incubator.⁸ There is also an increased risk of skin infection and mechanical compression, leading to problems like limb ischemia.^{1,2,8} There is also a risk of intoxication by cutaneous absorption of topical products, for example salicylate intoxication (similar to aspirin overdose) due to keratolytics.⁹ The condition is not thought to be painful or in itself distressing to the child. Nursing usually takes place in a neonatal intensive care unit, and good intensive care seems to have improved the prognosis markedly.¹ The collodion membrane should peel off or "shed" 2 to 4 weeks after birth, revealing the underlying skin disorder. The condition can resemble but is different from harlequin type ichthyosis. In collodion babies fluid and electrolyte balance and body temperature must be carefully monitored. In addition to this the membrane must be lubricated and to achieve elasticity and desquamation an adequate hydration of the skin are the major components of management. Suitable eye care and pain control should be carried out for the collodion babies with ectropion. Humidified incubators and water dressings followed by emollient agents are the essentials of the management. If there is a respiratory failure, ventilatory support for the collodion babies may be needed.

The "Collodian Baby" appearance can be caused by several skin diseases, and it is most often not associated with other birth defects.¹⁰ In most cases, the baby develops an ichthyosis or ichthyosis-like condition or other rare skin disorder. Most cases (approximately 75%) of collodion baby will go on to develop a type of autosomal recessive congenital ichthyosis (either lamellar ichthyosis or congenital ichthyosis form erythroderma).¹⁰ In around 10% of cases the baby sheds this layer of skin and has normal skin for the rest of its life.^{2,5} This is known as *self-healing collodion baby*. The remaining 15% of cases are caused by a variety of diseases involving keratinization disorders.¹⁰ Known causes of collodion baby include ichthyosis vulgaris and trichothiodystrophy.³

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