A New Born with Lamellar ichthyosis (Collodion Baby)

Zahid Gul, Gauhar Ali Khan, Fahad Liaqat and Kashif Muqarrab

ABSTRACT

Ichthyosis is an infrequent clinical entity worldwide with an incidence of 1:600,000 births. It can be one of the two types: collodion baby and Harlequin fetus or malignant keratoma (most severe form). The clinical manifestations in either form are thick and hard skin with deep splits. Affected babies are born in a collodion membrane, a shiny waxy outer layer to the skin that is shed 10 - 14 days after birth, revealing the main symptom of the disease. The reported case is of a neonate, born to primigravida mother at seven and a half month's gestation with a birth weight of 2160 grams and Apgar score of 6/10 and 8/10 at 1 and 5 minutes respectively. Conclusively, early diagnosis of this condition can help cope and prevent serious morbidity or even mortality at time. These newborns should be monitored carefully in intensive care units by a multi-disciplinary team.

Key Words: Lamellar ichthyosis. Newborn. Collodion baby.

INTRODUCTION

The word ichthyosis is derived from the Greek 'ichthys' meaning 'Fish' due to the similarity in appearance to fish-like scales. It is a very rare inherited skin disorder, affecting around 1 in 600,000 live births. The affected infant has a collodion type membrane, a shiny waxy outer layer to the skin. The severity of symptoms can vary enormously, from the mildest types such as Ichthyosis vulgaris which may be mistaken for normal dry skin up to life-threatening conditions such as Harlequin ichthyosis. The most common type is Ichthyosis vulgaris, accounting for more than 95% of cases. The babies with such condition shed the skin 10 - 14 days after birth. 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. Clinically, the collodion babies may encounter serious morbidities or even mortality due to dehydration leading to electrolyte imbalance, hypothermia due to altered perfusion, recurrent pneumonia or respiratory problems and increased sepsis risk because of severe skin damage.

CASE REPORT

A 23 years old un-booked primigravida, housewife, presented with history of seven and a half month’s gestational amenorrhea and labour pains for 5 hours before admission. She was married since 18 months. There was no consanguinity. She had not received any antenatal care and obstetric ultrasound was not done during antenatal period. There was no significant past medical/surgical history.

General physical examination revealed mild anemia. Obstetric examination revealed the baby was average sized with cephalic presentation and normal CTG. She was in the latent phase of labour. Labour augmentation was done with oxytocin infusion. She had normal vaginal delivery 2 hours after admission.

Patient delivered a male baby with birth weight of 2160 grams (below 5th Centile weight for age). Baby was covered with white parchment. Apgar score was 6/10 and 8/10 at 1 and 5 minutes respectively. Baby had features of rare congenital genetic disorder i.e. Lamellar ichthyosis/Collodion baby (Figure 1 a,b,c,d). Baby was kept in neonatal intensive care unit (NICU) for proper management. A multi-disciplinary team was involved and the diagnosis was confirmed after review by a dermatologist. The baby was kept in incubator for prevention of dehydration, intravenous IV line was maintained and was started on intravenous (IV) fluid and

Figure 1 (a,b,c,d): Parchment like skin with cracks and peel, fish like mouth and ectropion.
injectable antibiotic. The parents were counseled regarding the disease and its future prognosis and outcome.

**DISCUSSION**

*Lamellar ichthyosis* is known to be an autosomal recessive genetic disorder, the carriers do not show any sign or symptoms of the disorder. The term collodion baby was first used in 1884 by Hallopeau. The term was used for newborns in whom the entire body surface is covered with thick skin sheets, called collodion membrane. The collodion membrane is the result of an epidermal developmental dysfunction. It is composed of thick skin sheets which resemble translucent, tight parchment paper. The skin of a collodion baby has a shiny film that looks like a layer of vaseline. The taut membrane may impair respiration, sucking and mechanical compression, leading to problems like limb ischemia. There are fissures with impairment of barrier to infection and water loss leading to difficulties in thermal regulation and hypernatremic dehydration. Neonatal morbidity and deaths may be due to cutaneous infection (gram-positive and *Candida* spp.), aspiration (squamous material) and hypothermia. The eyelids and mouth may have the appearance of being forcibly open due to the tightness of the skin. There can be associated eversion of the eyelids (ectropion).

Treatment with a high-humidity environment and application of non-occlusive lubricants may facilitate shedding of membrane. Long baths, using bath oils or application of lubricants before drying can prolong the hydration and softening. The collodion membrane is peeled off or shed 2 - 4 weeks after birth, revealing the underlying skin disorder. In addition to this, the membrane must be lubricated, to achieve elasticity and desquamation, an adequate hydration of the skin is 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, ventilator support for the collodion babies may be needed.

In this case, a multi-disciplinary team was involved from the day first to be ready for anticipated complications. However, we did not document any electrolyte imbalance. The C-reactive protein was normal and blood culture showed no growth which ruled out sepsis. The baby was kept in humidified incubator and was given IV fluids, IV antibiotics, and hourly application of emollient and frequent oiling of the skin with minimal and aseptic handling. The patient was discharged on day 9 of admission with advice to come for regular follow-up.

**REFERENCES**


