Case Report

Collodion newborn with dehydration fever: A rare case report

Ayush Sopori[1], Mohit Bajaj[2], Seema Sharma[3], Akshit Sapehia[1]

From [1]Junior Resident, [2]Senior Resident, [3]Professor, Department of Pediatrics, Dr Rajendra Prasad Government Medical College, Kangra, Himachal Pradesh, India

ABSTRACT

Ichthyosis is a rare congenital disorder of the skin characterized by the presence of extra covering of tough film-like membrane over the skin leading to fissure formation on stretching. Clinically, newborns present with a membrane with fissures, ectropion, eclabium, and hypoplastic digits. Breakdown of the membrane makes newborns more prone to fluid loss, dehydration, electrolyte imbalance, body temperature instability, and infections leading to high mortality. Here, we report the case of a newborn with this condition presenting with fever, dyselectrolytemia, and dehydration after 48 h of birth managed as dehydration fever after excluding sepsis.

Key words: Newborn, Collodion, Dehydration fever, Ectropion

The term collodion was first used by Hallopeau and Watelet in 1892 which is a Greek word meaning Glutinous (such as glue and sticky) [1]. Ichthyosis is a rare clinical condition with an overall incidence of 1: 300,000 births [2] with around 270 cases reported till today with similar incidence in males and females [3]. Most of the reported cases have a history of consanguinity associated [4]. Collodion membrane is an epidermal developmental malformation that results in tight cellophane-like extra skin covering over the newborn with desquamation gradually over a period of 2 weeks in most of the cases [3]. Ichthyosis is described clinically by desquamation patterns and histopathologically by hyperkeratosis. They are recognized by the method of inheritance, clinical features, related deformities, and histological discoveries [5,6].

Newborn with generalized hyperkeratosis also known as the collodion layer presents with several diverse clinical phenotypes. The severe classic lamellar ichthyosis phenotype is characterized by large earthy colored thick scales with a plate-like appearance over the whole body surface combined with eyelid ectropion. Smaller and lighter scales are seen in milder phenotype with generalized skin redness [7].

Histologically, lamellar ichthyosis shows orthohyperkeratosis while innate ichthyosiform erythroderma shows scattered parakeratosis in thickened corneum. The separation between the two phenotypes in the postnatal period is troublesome until some other time in childhood [7]. This condition makes the newborn more prone to dehydration, electrolyte imbalance, sepsis, and temperature instability leading to frequent complications and high mortality [8]. The management of a collodion baby is challenging, but through timely skincare, good hand hygiene, asepsis maintenance, prevention of insensible water loss in sick newborn care unit (SNCU), and early initiation of breastfeeds most of the complications can be prevented leading to improved outcome. Here, we report a case of a collodion newborn with dehydration fever.

Access this article online	
Received 04 October 2020 Initial Review - 21 October 2020 Accepted - 28 October 2020	Quick Response code
DOI: 10.32677/IJCR.2020.v06.i11.005	

CASE REPORT

A female neonate conceived at full term by vaginal delivery to a 26-year-old primigravida mother, of a non-consanguineous marriage was admitted to SNCU at our tertiary care center with indications of dryness of the skin, scaling, and fissuring at places, and fever for 1 day up to 99.4°F. The mother had no history of any drug intake, radiation exposure, or any significant event during pregnancy.

The baby cried immediately after birth with the APGAR scores of 7 and 9. On physical assessment, the entire body was found to be secured with parchment-like layer (collodion) and was stripping off from the whole body including the face. There was outward turning of the mouth, as illustrated in Figs. 1 and 2.

A possibility of early-onset sepsis was kept. Injection ampicillin/cloxacillin and gentamycin were started on an empirical basis. Liquid paraffin and glycerine were applied over the body thrice daily after consultation with dermatology. For ectropion, eye drops, ciprofloxacin and carboxymethylcellulose sodium, were started, and for constriction bands in bilateral hands, surgical consultation was done and nothing active was done.

Correspondence to: Dr Mohit Bajaj, Room Number 106, Vivekananda Hostel, Dr Rajendra Prasad Government Medical College, Kangra, Himachal Pradesh, India. E-mail: tandian427@gmail.com

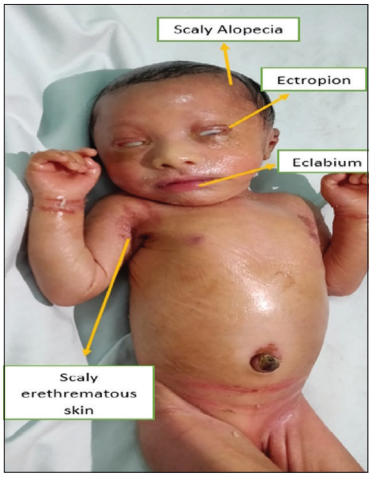

Figure 1: Classical features of collodion baby

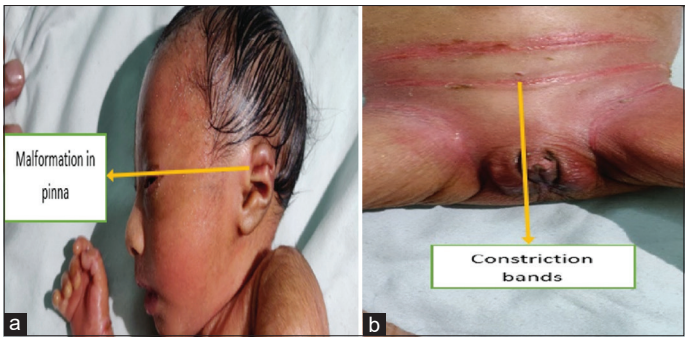

Figure 2: (a) Malformed pinna; (b) constriction bands on the abdomen

The initial sepsis screen of the newborn was negative (total leukocyte count – 15,000 with N53%, L37%) with C-reactive protein (CRP) negative. Renal function tests were suggestive of acute kidney injury with hypernatremic dehydration. Initial reports of kidney function tests revealed raised blood urea nitrogen (BUN) to 54 mg/dl and creatinine to 2.1 mg/dl. Electrolytes revealed hypernatremia with sodium values of 156 mmol/L.

The newborn received Intravenous fluids and antibiotics on day 1 of admission and expressed breast milk by orogastric tube on the 2nd day of admission. Fever subsided on day 2 of admission with renal function test improving (BUN – 43 mg/dl and creatinine 1.6) and sodium levels also coming to normal (150 mmol/l). On the 4th day of life, the skin desquamation was noted. Peeling away off the skin layer at folds and joints resulted in many raw areas. Breastfeeds were gradually increased to 150 ml/kg/day. Repeat sepsis screen on day 4 of admission was negative after 48 h. Acute kidney injury and hypernatremia returned to normal values (Table 1).

With the supportive management and continued skincare, the collodion membrane was limited to a small area of the body on day 5 of life leaving the normal skin behind. The blood culture of the newborn was sterile and antibiotics were omitted on day 5 of admission. Skin desquamation was complete at 2 weeks of life and the baby was discharged on day 14th of life. Follow-up at 4 weeks of age revealed healthy skin covering and adequate weight gain. For ectropion, the child is on eye drops and under follow-up. Hearing examination by BERA was normal. The newborn was given emollients for home application and is currently doing well (Fig. 3).

DISCUSSION

Collodion baby is a term used to describe a newborn covered with a thick tight membrane all over the body with fissures; this membrane subsequently sheds off over a period of time [2]. It is not a single disease entity, but rather a phenotype that later on demonstrates various spectrum of disorders including autosomal recessive congenital ichthyosis, lamellar ichthyosis, and harlequin ichthyosis, or less commonly may heal completely (self-healing), especially in those presenting with a membrane restricted to the extremities [5]. Although we had no capacity to do genetic testing to determine the phenotype, the clinical presentation of this newborn suggested a harlequin or lamellar ichthyosis. This is because of the presence of collodion membrane at birth with eclabium, ectropion, scaly alopecia, edematous feet, and nail hypoplasia with subsequently scaly erythematous skin.

Harlequin ichthyosis is the most severe form of autosomal recessive congenital ichthyosis. It was previously considered to be associated with poor survival, but with good postnatal care, survival beyond 7 years of age has been reported [9]. Complications associated with increased risk of mortality are infection, fluid loss, dehydration, electrolyte imbalance, and body temperature instability [8]. Our newborn presented with mild hypernatremia and high serum creatinine, these investigation results were before administration of any intravenously administered fluids, which resolved within 2 days after adequate hydration. Severe hypernatremia with uremia which responded well to fluid correction has been reported [7,8]. Thus, prompt correction of dehydration is essential as these babies are at increased risk of fluid loss through bare skin and loss to surroundings if they are not kept in a humidified room [8].

In an ideal setting, these newborns must be admitted to SNCU and cared for in high humidity incubators [8]. However, this is challenging in resource-limited settings where the newborn unit may not be able to fulfill these requirements. However, with close monitoring, proper hygiene, and isolation to reduce the risk of infection, the survival of these babies can be improved. In our case, there were no signs of sepsis. A literature review has revealed that there are babies who died within a few days due to sepsis and septic shock [10,11]. Hence, we considered the possible risk of infections in this baby because of the impaired barrier function of her skin and increased susceptibility to *Staphylococcus aureus*, *Streptococcus pyogenes*, and *Klebsiella* species infections [12,13]. Thus, we initiated the baby on antibiotics by intravenously administering Ampiclox (ampicillin and cloxacillin) and gentamycin until sepsis was excluded by the laboratory results of CRP, complete blood count, and blood culture. Our index newborn had hyperthermia at admission. Complication like temperature deregulations [13,11] is very commonly observed in these newborns.

Table 1: Investigation sheet of the patient

Investigations	Day 1	Day 2	Day 3	Day 4	Day 5
Hb (g/dl)	14.2		12.7	12.5	12
Total leukocyte count (/mm^3)	15,000		13,250		12,100
Differential leukocyte count	N53%, L37%		N48%, L42%		N42% L55%
Platelets (cells/mm^3)	354,000		322,000		314,000
C-reactive protein (mg/L)	<6		<6		<6
Blood culture					Negative
Blood urea nitrogen (mg/dl)	54	43	32		26
Creatinine (mg/dl)	2.1	1.6	1.2		0.9
Sodium (mmol/L)	156	150	148		142
Potassium (mmol/L)	5.2	4.8	4.8		4.5
T3/T4/TSH			113.9/18.1/3		
Total serum bilirubin (mg/dl)			9.6		8.5
Neurosonogram			Normal		
Ultrasound kidney and urinary bladder			Normal		

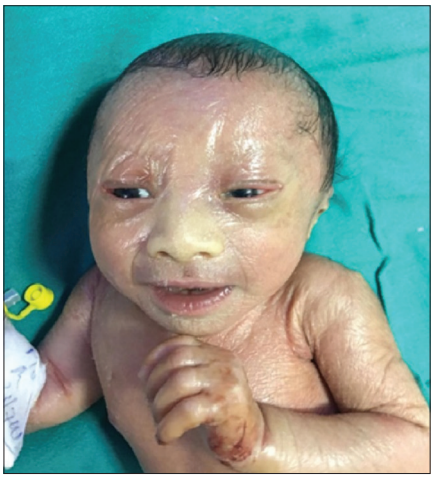

Figure 3: Child on day 8 of admission with improvement in scaling and desquamation after proper emollient application

Another important aspect of care of these babies is eye care because the tight membrane does not allow eyes to close normally and conjunctiva is exposed, leading to exposure keratitis [13]. The long-term complications include hyperopia and anisometropia which may necessitate the wearing of glasses [12]. Persistence of ectropion by the age of 6 months may need a surgical correction [8]. In the case presented here, the ectropion had some improvement in the 1st month of life; the baby was closely monitored by ophthalmologists, who provided regular follow-up eye care.

These babies are also at increased risk of developing conductive hearing loss [1,8]. A case of a baby with ichthyosis who was diagnosed as having a conductive hearing loss on the 83rd day of life has been reported; the problem persisted for the whole duration of the 2-year follow-up [13]. We cannot predict the hearing outcome of the baby due to the presence of excessive glue-like material in her ears and on recommendations of ENT specialist, daily application of normal saline was done. On follow-up visits , a hearing assessment was done with BERA and it was within normal values.

The patient presented here developed a constricted band, which impaired blood flow to her distal extremities, which caused swelling of her limbs, necessitating surgical release. However on day 4 of life, the band gradually eased off with membrane breakdown. A delay in the release of the band could have caused acute compartment syndrome and gangrene to the distal limbs [8], which could result in loss of the limb. We could not make a definitive diagnosis to determine the phenotype, since it needs genetic testing of both parents and the baby, which is currently unavailable in our hospital.

CONCLUSION

Collodion newborns are rarely born and have multiple organ involvement. Thus, it is essential to have a set protocol for the treatment of these newborns. Early admission to SNCU is a must for the proper management of the complications. Survival of these newborns depends on improved basic care which includes proper hydration; infection control, proper skin, eye, and ear care.

CONSENT

Duly taken from parents regarding the clinical images and case history.

REFERENCES

1. Stojanović V, Doronjski A, Spasojević S, Stašuk N, Radulović A. Collodion baby-a case report. Serbian J Dermatol Venereol 2014;6:186-90.
2. Pehlivan Ö. Collodion baby. Turk Acad Dermatol 2008;2:1689.
3. Shrikhande DY, Chatterjee R, Kumar A, Kochar I, Mandade K, Vasoya M. Collodion baby: A rare case report. IOSR J Dent Med Sci 2014;13:85-7.
4. Sharma S, Mahajan VK. Collodion baby. Indian Dermatol Online J 2011;2:133.
5. Ferrari B, Martínez JP, Luna PC, Larralde M. Acral self-healing collodion baby: A case series. Int J Womens Dermatol 2016;2:140-2.
6. Aradhya SS, Srinivas SM, Hiremagalore R, Shanmukappa AG. Clinical outcome of collodion baby: A retrospective review. Indian J Dermatol Venereol Leprol 2013;79:553.

7. Srivastava P. Congenital ichthyosis-collodion baby case report. J Clin Diagn Res 2016;10:SJ01-2.
8. Glick JB, Craiglow BG, Choate KA, Kato H, Fleming RE, Siegfried E, *et al.* Improved management of harlequin ichthyosis with advances in neonatal intensive care. Pediatrics 2017;139:e20161003.
9. Mithwani AA, Hashmi A, Shahnawaz S, Al Ghamdi Y. Harlequin ichthyosis: A case report of prolonged survival. BMJ Case Rep 2016;114:113-4.
10. Obu HA, Adimora G, Obumneme-Anyim IN, Ndu I, Asinobi I. Collodion baby: A report of 4 cases. Niger J Paediatr 2013;40:307-10.
11. Ramteke S, Agrawal A, Shrivastava J. Harlequin icthyosis: A rare disorder. Indian J Case Rep 2016;2:69-71.
12. Chang LM, Reyes M. A case of harlequin ichthyosis treated with isotretinoin. Dermatol Online J 2014;20:doj_21540.
13. Harvey HB, Shaw MG, Morrell DS. Perinatal management of harlequin ichthyosis: A case report and literature review. J Perinatol 2010;30:66-72.

Funding: None; Conflict of Interest: None Stated

How to cite this article: Sopori A, Bajaj M, Sharma S, Sapehia A. Collodion newborn with dehydration fever: A rare case report. Indian J Case Reports. 2020;6(11):621-624.