

A Rare Case Report of a Collodion Baby with Severe Hyponatremia

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ABSTRACT

Background: Collodion baby is a rare condition (i.e., 1:300000 birth), which is referred to a neonate covered with a tight and shiny membrane desquamating within two weeks. This condition takes place as a result of the epidermal cornification disorder. Given the impairment of the skin barrier function, these neonates are at the risk of several complications, including hyponatremic dehydration, hypothermia, skin infections, fissures, conjunctivitis, sepsis, dehydration, and constrictive bands of the extremities resulting in vascular compromise and edema. This condition has a high mortality rate; accordingly, the majority of the collodion babies die within the first few weeks of birth due to the secondary complications depending on the type of mutations. Although the collodion membrane is a transient condition, it can lead to a number of complications in 45% of the newborns and result in 11% mortality rate in the first few weeks of the neonatal life. However, in the recent years, the mortality rates have been declined owing to the introduction of systemic retinoids to the clinical practice and the advanced care methods, which are used in the intensive care units. Harlequin ichthyosis has been associated with the mutation in the ABCA12 gene; therefore, genetic counseling and mutation screening of this gene should be considered.

Case report: Herein, we reported a case of a collodion baby with favorable evolution, who developed hyponatremia and suffered from some complications having aroused from the condition.

Conclusion: Infection prevention and supportive care of collodion babies mainly with incubator, serum therapy, and feeding support can be effective in preventing complications.

Keywords: Collodion baby, Harlequin fetus, Hyponatremia, Sepsis

Introduction

Collodion baby is a rare condition (i.e., 1:300000 birth), which occurs due to epidermal cornification disorder (1). This condition is referred to a newborn covered with a tight and shiny membrane that desquamates within two weeks (2). Most of the collodion newborns develop signs and symptoms of several ichthyosis types; however, some autosomal recessive types completely recover following scaling (2). In a study, 43%, 19%, 12%, and 25% of these neonates had erythrodermic autosomal recessive lamellar ichthyosis, non-erythrodermal autosomal recessive ichthyosis, other forms of ichthyosis, and normal skin, respectively (3).

Although the collodion membrane is a transient condition, it can raise some complications in 45% of newborns and result in 11% mortality rate in the first few weeks of neonatal life (2). These

complications include skin abnormalities (e.g., fissuring, bilateral ectropion, autoamputation of digits due to constricting membrane, and limb swelling), as well as hair and nail, ophthalmologic, gastrointestinal (e.g., poor weight gain despite high caloric feeding, constipation, gastroesophageal reflux), respiratory (the most common problem in neonatal period that leads to death), and biochemical (e.g., hyponatremia, hypocalcaemia, and hypoglycemia) problems. In the present report, we presented the case of a collodion baby who developed hyponatremia and uremia (4).

Case report

Our case was a full term male neonate born with Apgar score of 8-9 and birth weight of 2500 g through vaginal delivery. The patient was

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Table 1. Results of laboratory tests during admission

Variable	Post-admission time												
	1 st hour	12 th hour	24 th hour	36 th hour	48 th hour	60 th hour	72 th hour	84 th hour	96 th hour	5 th days	6 th days	7 th days	8 th days
Blood sugar (mg/dL)	801	454	400	381	310	252	220	196		150	154	110	
Urea (mg/dL)	402	400	335	322	254	239	221	186		130	77	33	16
Cr (mg/dL)	3.7	3.3	3.1	2.9	2.5	2.4	2.2	1.8		1.4	1.1	0.8	0.5
Na (mEq/dL)	187	185	181	179	176	174	169	167	164	159	150	148	142
K (mEq/dL)	6.5	5.3	4.4	5	4.6	5.6	4.5	4.1		3.8	3.5	3.7	4.3
Ca (mg/dL)	7.3	8.4			9.6	9.9		10.2				9.8	10.1
Urinary specific gravity				1030				1016		1020	1017	1006	

suspected of ichthyosis due to having dry skin. He was discharged from the hospital with good general condition and breast fed without any problem. The parents were advised to consult with a dermatologist in an outpatient clinic on the second day after birth.

After one week, he was admitted to the Neonatal Intensive Care Unit (NICU) of Imam Reza Hospital, Mashhad, Iran, due to weight loss of about 500 g, poor feeding, and severe generalized skin scaling, oliguria, and cyanotic attacks. The physical examination revealed generalized scaling with erythematous skin and lethargic appearance. His primitive reflexes were reduced, and the extremity pulses were palpable. No other abnormality was detected in the patient.

The initial laboratory data regarding the venous blood gas (VBG) values included: pH: 7.36, PCO₂: 59.6 mmHg, HCO₃: 27 meq/l, BE: 1.8 mmol/L, CBC: Hb: 16.1 g/dL, WBC: 8200 / μ L, CRP: 18 mg/L, ESR: 57 mm/h, Cr: 3.7 mg/dL, Urea: 402 mg/dL, Ca: 7.3 mg/dL, K: 6.5 mEq/dL, Na: 187 mEq/dL, BS: 801 mg/dL, PT: 20 sec, and PTT: 51 sec.

Sepsis workup was performed, and the antibiotic therapy (adjusted with glomerular filtration rate [GFR]) was initiated. In addition, 20 cc/kg of normal saline was administered three times until the patient urinated. Subsequently, the serum therapy was continued due to insensible water loss plus ongoing loss (fluid loss caused by damaged skin estimated as 20%), and urine output. For hyperglycemia, the patient received a single dose of subcutaneous insulin. The electrolytes and other laboratory tests were repeated every 12 h, and the serum therapy was adjusted considering his condition and laboratory test results (Table 1).

Simultaneously, conservative skin management was continued using vaseline plus urea, etc. The patient showed improvement in terms of the scaling and erythrodermia during admission. Vitamin K was administered, and the antibiotic therapy was changed to vancomycin after the observation of the culture results (i.e., CSF/C: Neg,

Urine/C: Neg, B/C *2: *Staphylococcus epidermis* sensitive to vancomycin). The antibiotic dosage was determined based on the patient's GFR each day. The subsequent cultures were negative, and the result of the brain computed tomography scan was normal.

Finally, after 16 days of hospitalization, the patient was discharged from the hospital with good general health condition and appropriate weight gain (reached to 2,260 g) while he was exclusively breast fed. The parents were thoroughly provided with the necessary information about the respective condition. The patients were followed up outpatiently. The auditory brainstem response test was performed after discharge.

Discussion

In this case report, we presented the case of a collodion baby who developed hyponatremia. Collodion baby is a rare condition (i.e., 1:300000 birth) (1) referring to a baby covered with a tight translucent sheet on the entire body, which desquamates within two weeks (2). This condition has been reported to be caused by the autosomal recessive ichthyosiform disease in almost all the cases.

Collodion baby is associated with specific conditions. Some of these conditions include autosomal recessive congenital ichthyosis, epidermolytic hyperkeratosis (bullous congenital ichthyosiform erythroderma), Gaucher's disease, Sjögren-Larsson syndrome, self-healing collodion baby, neutral lipid storage disease, trichothiodystrophy, annular epidermolytic erythema, Loricrin keratoderma, and hypohidrotic ectodermal dysplasia.

Although the collodion membrane is a transient condition, the complications and mortality rates are 45% and 11%, respectively. The collodion babies are highly exposed to dehydration, electrolyte imbalance, and infections (3). The majority of the newborns with such

condition die within the first few weeks of birth due to the secondary complications depending on the type of mutations (5).

The patients with the compound heterozygote mutation have higher survival rate, compared to those with the homozygote mutation (4). In the recent years, the mortality rates have been declined due to the entrance of systemic retinoids into the clinical practice and the advanced care methods, which are applied in the NICU (1).

The collodion membrane exerts mechanical compression distorting the features of the face and extremities. This gives the newborn a striking appearance that may initially frighten the family members and physicians who may have never confronted such a case. These newborns can have malformed ears as well as everted eyelids (ectropion) and lips, resulting in a fish-mouth appearance (2).

Due to the impairment of the skin barrier function, these neonates are at the risk of different complications, including hypernatremic dehydration, hypothermia, skin infections, fissures, conjunctivitis, sepsis, dehydration, and constriction bands of the extremities resulting in vascular compromise and edema (2, 3). Therefore, it is essential to place the collodion babies in a humidified incubator soon after birth to prevent hypernatremic dehydration and hypothermia (7).

In addition to the provision of the routine care, such as checking vital signs, these patients should be kept in a warm and humid incubator and be hydrated. Since it is difficult to access the peripheral veins, an umbilical venous catheter might be needed. In order to keep the skin soft and accelerate the desquamation, it is essential to shower the newborn twice a day and apply saline compresses as well as gentle emollients.

Furthermore, water and electrolyte disturbances must be managed in these newborns. The environment must be cleaned to prevent infection; therefore, repeated cultures of the skin would be necessary to detect the hazardous microorganisms.

In addition, genetic counseling and molecular investigation of the ABCA12 gene should be considered.

As mentioned above, our patient was admitted to the hospital one week after birth due to weight loss of about 500 g, poor feeding, desquamation, and cyanosis; furthermore, he showed evidence of dehydration. The collodion newborns should be attended in terms of the dehydration and hypernatremia during the hospital stay. With proper attention to the patient, the high blood glucose, acute renal failure, and hypocalcemia after hypernatremic dehydration can be well-managed (5).

Conclusion

Infection prevention and supportive care of collodion babies mainly with incubator, serum therapy, and feeding support can be effective in preventing complications.

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Conflicts of interests

The authors declare no conflicts of interest.

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