Case 13

A 3-day-old Thai female infant from Ayutthaya.

Chief complaint: Translucent membrane covered all entire body surfaces since she was born.

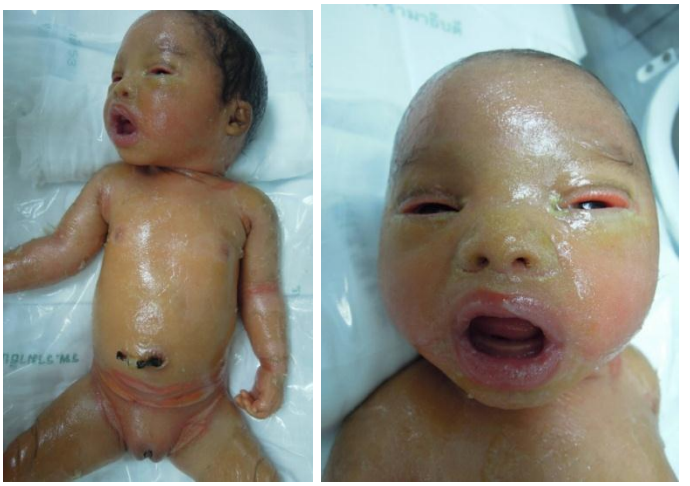

Figure 1: 3-day-old baby

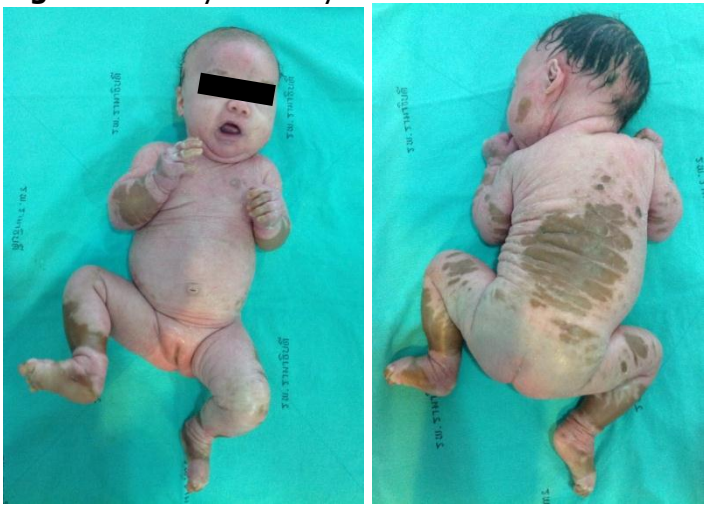

Figure 2: 1-month-old baby

Present illness: An appropriate-for-gestational-age infant girl was born at 37weeks' gestation to a 32-year-old female, gravida 2, para 1, with an unremarkable pregnancy. Elective cesarean section was performed for breech presentation. Apgars were 9 and 9 at 1 and 5minute. Her birth weight was 2800 gram. She was referred from private hospital due to shiny translucent membrane cover all over the body with ectropion of both eyelids and eclabium since she was born, as shown in figure 1.

Family history:

Patient was born to nonconsanguineous parents.
Her brother is a healthy 13 years old boy.
No significant dermatologic disease in family member

Physical examination

A Thai female infant, active
Vital sign: normal
HEENT: not pale, no jaundice, severe ectropion of both upper eyelids, no conjunctivitis, no corneal ulcer, slight eclabium, no cleft lip, no cleft palate
CVS: unremarkable
Lung: clear
Abdomen: soft, no hepatosplenomegaly
Extremity: no limit range of motion
Skin: shiny translucent membrane cover all entire body surfaces with fissure at inguinal area, lower part of abdomen and both wrists

Investigation:

CBC: Hb 15.4g/dL, Hct 46.7%, Plt 264,000
WBC 14,730 (N54%, L33%, M9%, E2%, B2%)
Electrolyte : Na 138, K 4.3, Cl 107, HCO 20.2 mmol/L

Diagnosis: Collodion baby

Treatment: Humidified incubator, Vaseline ointments apply all the entire body surface q.i.d., fusidic acid ointment apply fissure b.i.d, chloramphenical ointment apply both eyes q.i.d

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Discussion

Our patient presented with translucent membrane cover all entire body surfaces with ectropians and eclabium since she was born. The patient was full term infant; born to nonconsanguineous parents. Her parent and their family members have no history of significant skin disease. The clinical presentation was compatible with collodion baby.

Collodion baby is rare disease, estimated to occur in 1 in 50,000 to 100,000 deliveries, with a slightly male predominate[1,2]. Most cases are born at term and are not small for gestational age. At birth, the neonate is covered with a taut, shiny, transparent membrane formed by the thickened stratum corneum, which resembles a plastic wrap. The tightening of the skin may leads to ectropion, eclabium, absence of eyebrows,sparse hair, pseudocontractures, limitation range of movement, hypoplasia of nasal and auricular cartilage[1,3,4]. Systemic complication can occur including restricted pulmonary ventilation, dehydration, infection, sepsis, percutaneous toxicity from topical medication[5,6].

Collodion baby is a prodromal stage resulting from different congenital disorders of keratinization. The patient will spontaneously desquamate within 2-4 weeks, and reveal the true underlying skin disease. Most affected individuals later have development of autosomal recessive congenital ichthyosis encompassing, the clinical spectrum of classic lamellar ichthyosis to congenital ichthyosiform erythroderma. However several disorder of keratinization had been described as seen in table 1.

Histopathology of collodion membrane is nonspecific and reveals mostly an excessively thickened, orthokeratotic stratum corneum. However, a skin biopsy and genetic testing should be consider after transition to the underlying disease phenotype[7,8].

Histopathology of ichthyosis disease is generally non specific with exception for epidermolytic hyperkeratosis, neutral lipid storage disease, Refsum disease, and acquired ichthyosis associated with sarcoidosis.

Table 1

Disorder associated with collodian baby
Nonsyndromic icthyosis • Lamellar ichthyosis • Congenital ichthyosiform erythroderma • Ichthyosis valgaris • Recessive X-linked ichthyosis • Epidermolyticichthyosis • Bathing suit ichthyosis • Self-healing collodion baby
Syndromicichthyosis • Neutral lipid storage disease with ichthyosis • Trichothiodystrophy with ichthyosis • Conradi-Hunermann-Happle syndrome • KID syndrome • Loricrin keratoderma • ARC syndrome • KLICK syndrome
Metabolic disease • Halocarboxylase synthetase deficiency • Gaucher disease type 2
Other • Hypohidrotic ectodermal dysplasia • Congenital hypothyroidism

Treatment of collodion baby is to keep appropriated environment and to closely monitor for fluid and electrolyte imbalances. The exposed eyes in ectropian condition should apply with lubricating ointment[9]. Prevention for secondary pneumonia due to restricted ventilation and aspiration of amniotic fluid containing scales is essential. Systemic retinoid is infrequently used in collodion baby, due to spontaneous clinical improvement.

Our patient was placed in humidified incubator and the skin was regularly applied with vaseline ointment. Both eyes were applied with Chloramphenical ointment for prevention of corneal scratch. After 1 month of admission, the almost skin surface is desquamated and reveal generalized erythematous, fine scaly patches at trunk, back and all extremities, as shown in figure 2. Skin biopsy and genetic testing is our future plan of treatment in this patient.

References

1. Van Gysel D, Lijnen RL, Moekti SS, de Laat PC, Oranje AP. Collodion baby: a follow-up study of 17 cases. J Eur Acad Dermatol Venereol 2002;16:472-5.
2. Kurtoglu S, Ozturk MA, Koklu E, Gunes T, Akcakus M, Hatipoglu N. Serum insulin-like growth factor-I (IGF-I), IGF-binding protein-3, and growth hormone levels in collodion babies: a case-control study. J Pediatr Endocrinol Metab 2008;21:689-94.
3. Akcakus M, Gunes T, Kurtoglu S, Ozturk A. Collodion baby associated with asymmetric crying facies: a case report. Pediatr Dermatol 2003;20:134-6.
4. Harting M, Brunetti-Pierri N, Chan CS, Kirby J, Dishop MK, Richard G, et al. Self-healing collodion membrane and mild nonbullous congenital ichthyosiform erythroderma due to 2 novel mutations in the ALOX12B gene. Arch Dermatol 2008;144: 351-6.
5. Barrett DA, Rutter N. Transdermal delivery and the premature neonate. Crit Rev Ther Drug Carrier Syst 1994;11:1-30. Mancini AJ. Skin. Pediatrics 2004;113:1114-9. DiGiovanna JJ, Robinson-Bostom L. Ichthyosis: etiology, diagnosis,
6. and management. Am J Clin Dermatol 2003;4:81-95. Scheimberg I, Harper JI, Malone M, Lake BD. Inherited ichthyoses: a review of the histology of the skin. Pediatr Pathol Lab Med 1996;16:359-78.
7. Taieb A, Labreze C. Collodion baby: what's new. J Eur Acad Dermatol Venereol 2002;16:436-7.