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Case Report

Collodion Baby

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Introduction

Collodion baby is a rare congenital disorder characterized by parchment-like taut membrane covering the whole body, often resulting in ectropion and eversion of the lips.¹ We present one such case where diagnosis could not be entirely made out at initial examination (out of the entire spectrum) but essential supportive care helped in tiding through the initial critical phase.

Case report

A 2-day-old male neonate patient was admitted with complaint of membrane over the whole body since birth. The baby was born at term and was the first child of parents with third degree of consanguinity. On examination, the whole body was covered with parchment-like membrane, which was peeling off on the chest and abdomen (Fig. 1). Associated

findings included eversion of lips, ectropion, flattening of nose and ears, claw-like hands, and limitation of joint movements. There were no other congenital anomalies. Hair and nails were normal. Routine blood and urine analysis were within normal limits. Skin biopsy showed epidermal hyperkeratosis and preservation of granular layer. The patient was treated in humidified environment with intravenous fluids and prophylactic antibiotics. Emollient was applied to the whole body. The patient recovered with membrane peeling but erythroderma was persistent (Fig. 2). Later, the patient was discharged with advice for regular emollient application and follow-up.

Discussion

Collodion baby is an underreported condition. According to Chung et al., about 270 cases were reported in the literature since 1892.² Incidence of collodion baby is estimated to be 1 in 300,000¹ although FIRST (Foundation for Ichthyosis and Related Skin Types) estimates it to be relatively more common 1 in 100,000.

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Fig. 1 – At birth.

These babies are usually born before term. The parchment-like membrane undergoes desquamation with time. Once this layer peels off, underlying skin disorder is usually revealed. At least one-tenth of these patients later develop normal underlying skin. This is one end of the spectrum for this heterogeneous disorder and can be called 'self-healing' collodion baby. Another end of spectrum in keratinizing disorder presenting at birth is Harlequin fetus. These babies have horny plates all over the body. Most of these babies die within first few days to weeks of life because of respiratory difficulty, inability to feed, and severe skin infections. Patients who survive beyond infancy have severe ichthyosis and variable neurologic impairment. In all cases, barrier function of the skin is affected once collodion dries up and starts cracking. This leaves the baby susceptible to infection, temperature instability, and excessive trans-cutaneous fluid losses leading to dehydration. Thick membrane can create problems related to free movement leading to difficulties with breast-feeding. If chest wall movements are restricted, it may cause hypoventilation. Inability to close eyelids may cause corneal damage if proper precautions are not followed.

Primary pathology in ichthyosis is disordered cornification, which leads to formation of collodion membrane. Two most common types of skin disorders presenting as collodion at birth are non-bullous congenital erythroderma and Lamellar ichthyosis. Other less common pathologies responsible for this presentation include: Ectodermal dysplasia, Sjögren Larsen syndrome, Comel-Netherton syndrome Gaucher Disease type

2, Hay-Well syndrome, Trichothiodystrophy, Neutral lipid storage disease, etc.³ Genetically, it is a heterogeneous disorder with usually autosomal recessive transmission. So far, six genes have been associated with mutations with transglutaminase 1 (TGM1) accounting for approximately 40% cases.⁴ TGM1 mediates cross-links in the formation of the cell envelope during terminal differentiation of epidermis.^{5,6} Mutation in genes involved in nucleotide excision repair pathway has also been implicated.⁷

Management requires the expertise of a dermatologist and the pediatric team. The baby is initially kept in a humidified, neutral temperature environment like an incubator. Other supportive treatments such as intravenous fluid and tube feeding may also be required to maintain hydration and nutrition. One should keep the skin soft and also attempt to reduce scaling. The collodion membrane must not be debrided. Some clinicians find use of emollients helpful,² although Van Gysel et al. in their case series of 17 babies with similar problem found emollient application to be associated with increased the risk of infection.⁸ There is also a risk of intoxication by cutaneous absorption of topical products, and Yamamura et al. have reported a case of salicylate intoxication due to use of keratolytics.⁹ Hence, the emollient used should be inert-like petroleum jelly. Cakmak et al. found acitretin in dose of 1 mg/kg/day to be effective and safe in management of congenital ichthyosis.¹⁰ Artificial tears may be necessary if there is severe ectropion. Treatment may also include pain relief and mild topical steroids to reduce secondary inflammation.

Fig. 2 – After 1 week.

Conclusion

Collodion baby probably is underreported condition with cases which have mild presentation not getting reported. Our experience with this case taught us that careful attention to skin care, judicious use of skin care products, and meticulous attention to asepsis are of paramount importance when caring for an infant with collodion baby syndrome. Because of rarity and heterogeneous nature of this disease, experience of individual clinician may be limited; hence, a protocol should be established to manage the dynamic nature of this condition.

Conflicts of interest

The authors have none to declare.

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