Case report

A full term male infant was born by spontaneous vaginal delivery to a 35 y/o, G5P4 woman. Her prenatal course, including prenatal ultrasound, was unremarkable.

Examination at birth: He had normal anthropometry with height, weight and head circumference being between 25th to 75th percentiles.

Skin: Diffuse erythematous skin lesion with collodion membrane covering the chest and back, tight skin on upper and lower extremities, fissures on areas with skin folds, eversion of the eyelids with inability for complete eye examination, eversion of the lips, and malformation of the auricle.

Tone and turgor: within normal limit.

Heent: Anterior fontanelle: soft and flat, normal shape and size.

Eyes: Eversion of the eyelids.

Ears: External malformation, unable to assess if secondary to skin lesions.

Nose: Nares and septum appear within normal limit.

His cardiovascular, abdominal, spine, back, extremities and external gentile examinations were normal.

On further evaluation, family history was not significant for any medical and dermatological conditions. There was no history of consanguinity. The patient has a 2 y/o healthy full sister. Mother was carrier for Cystic Fibrosis. He has two half-brothers (13 y/o and 10 y/o) from maternal side who has eczema, otherwise healthy; two half-sisters and one brother from paternal side - all reportedly healthy (Figure 1 and 2).

Discussion

Collodion baby is a severe form of Congenital Ichthyosis. Congenital Ichthyosis is heterogeneous group of disorders of cornification involving all or most of integumentary system [1] which is characterized by a generalized scaling of the skin of varying severity. Mucosal surface and visceral organs are generally spared. Cornification is process of terminal keratinocyte differentiation. At this stage, a lipid envelope is extruded into the intercellular space from lamellar bodies to form lipid sheets composed of ceramides, cholesterol, and free fatty acids [2].

Normal skin turnover

Skin has dermis and epidermis layers. Epidermis has four layers-stratum basale, spinosum, granulosum, and corneum from base to top. During normal skin turnover of 28 days, site-relevant proteins and lipids are synthesized in different layers of epidermis.

Revised nomenclature and classification of inherited ichthyoses

The First Ichthyosis Consensus Conference in Sorèze 2009 proposed the following pathogenesis of inherited Ichthyosis [1] which is based on disorders involved in different stages of protein and lipid metabolism and on other mechanism.

Figure 1. Collodion membrane with eclabion, tight membrane around joints.
Collodion baby (CB) is a presentation of inherited Ichthyosis—it is not a disease. It is called “inherited” to different from rare condition of “acquired Ichthyosis”, primarily seen adults as a manifestation of systemic disease, including paraneoplastic syndrome. Collodion membrane is transient, transparent membrane present at birth, made of keratinized epithelium [3]. Clinical course of collodion baby varies widely with complete resolution within few weeks in ten percent, lifelong complication of various degrees to most affected individuals and five percent mortality as neonate [4]. Skin peeling occurs from weeks to months of life [5].

CB presents at birth with erythroderma and shiny, tight skin resembling parchment (the so-called collodion membrane) covering the entire body [4]. The mechanical force of membrane causes distortion in different body parts that can result ectropion, eclairon, pseudocontractures, absence of eyebrows, sparse hair, and hypoplasia of nasal and auricular cartilage [4]. Distal limb ischemia and hypohydrosis has been reported. Less common presentation includes poor sucking (due to eclairoon), restricted pulmonary ventilation (decreased chest wall compliance), digital vascular constriction, and edema of the extremities [5]. After birth, the expansion of the chest with breathing may cause tearing in the inelastic membrane, which begins to peel. The membrane is shed within 3 to 4 weeks, usually revealing an ichthyosis phenotype [4,6]. CB is rare, estimated to occur in 1 in 50,000 to 100,000 deliveries. Most CB infants are born at full-term and are appropriate size for gestational age. A slight male predominance has been found in the literature [4] and this is explained by the fact that only males are affected by X-linked forms of inherited ichthyosis. Cases have been reported in twins, and consanguinity of parents has been noted [7]. Vitamin D deficiency rickets has been reported to be associated with Ichthyosis [8].

Newborn management

Fluid and electrolyte management: Increased insensible water loss, more than five times normal trans-epithelial water loss has been reported, which leads to dehydration and hyponatremia [9,10]. To avoid dehydration, newborn are kept in isolette with high humidity and milk/ intravenous fluid intake is adjusted to avoid dehydration. CB also have impaired sweating mechanism so clinical must be vigilant to avoid and aggressively treat hyperthermia and hypohydrosis. Daily weight and electrolytes help in maintaining fluid and electrolyte balance.

Routine use of emollients is controversial as it helps decreasing insensible water loss but increases the risk of infection [11,12]. Defecting skin barrier promotes bacterial and fungal infection. Deep skin crease may delay early diagnosis. Daily skin examination including skin fissures is recommended. Rigorous hand wash cannot be over emphasized. Early diagnosis and aggressive treatment of both local and systemic infection is required for better outcome. Eye care should be coordinated with ophthalmologist and bland lubricant is recommended to prevent development of keratitis and conjunctivitis [6,13]. For severe scaling, oral retinoids, such as acitretin and isotretinoin, are an important therapeutic option [14].

Our case

We kept newborn in tertiary level NICU. He was in Isolette with humidity of 70-80 %. He was given feeds in demand and IV fluid for first 2 days. Intravenous Ampicillin and gentamycin was given for 5 days. We monitored daily weight and electrolytes, strict input and output. There was no significant weight change and no electrolyte abnormalities. We used topical Vaseline twice daily and looked for signs and symptoms of cutaneous and systemic infections daily. He did not have any infection. He got eye lubricants 3-4 times a day to prevent dry eye and its complications. Baby had umbilical venous line was maintained for first 2 days. Intravenous Ampicillin and gentamycin was given for 5 days. We monitored daily weight and electrolytes, strict input and output. There was no significant weight change and no electrolyte abnormalities. We used topical Vaseline twice daily and looked for signs and symptoms of cutaneous and systemic infections daily. He did not have any infection. 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weeks. Ectropion of eyelid and eclabion improved as membrane shed. Genetic testing were negative. He got afebrile, superficial skin infection, treated with systemic antibiotics at 6 weeks of age. Child is doing well in 15 months of life. We discussed the inheritance of congenital Icthyosis. The majority of cases of congenital Icthyosis are inherited in an autosomal recessive manner. This means that for both male and female offspring, the risk of recurrence is 25% with each pregnancy.

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