Clinical Case Report

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Particularities of the management and the treatment in a rare sepsis with Candida tropicalis of a Collodion baby

Case report

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Abstract

Rationale: Collodion baby is a rare autosomal recessive disorder. It can be the first expression of some forms of ichthyosis.

Patient concerns: The authors present the case of a newborn diagnosed with severe Collodion baby syndrome who required prolonged hospitalization in the intensive care unit because of infectious complications like the fungal sepsis and other bacterial superinfections.

Diagnoses: The case has many diagnostic and therapeutic particularities and management difficulties. Skin culture, dermatological and genetic exam were required.

Interventions: The treatment required multidisciplinary involvement: neonatologist, pediatrician, geneticist, dermatologist, psychologist, ophthalmologist, audiologist.

Outcomes: The evolution during hospitalization was slowly favorable, but later, after a few months, it developed some complications.

Lessons: In our case, skin injuries, total parenteral nutrition, aggressive and prolonged antibiotic therapy, intravenous devices, high hospitalization duration were risk factors for colonization and sepsis with fungi, especially in the neonatal period, sometimes with severe evolution and prognosis.

Abbreviations: ABCA = ATP-binding cassette transporter, ALOX12B = arachidonate 12-lipoxygenase, ALOXE3 = arachidonate lipoxygenase 3, MRSA = methicilin-resistant Staphylococcus aureus, TGM1 = keratinocyte transglutaminase 1 enzyme.

Keywords: Collodion baby, congenital ichthyosis, lamellar ichthyosis, neonatal fungal sepsis

1. Introduction

Lamellar ichthyosis or Collodion baby is a rare autosomal-recessive disorder which belongs to the ichthyosis group and is manifested since birth compared with other types of ichthyosis which manifest after the age of 3 months. Foundation for Ichthyosis and Related Skin Types estimates an incidence of 1 case per 100,000 individuals and the incidence in males and females is equal. The disorder is caused by mutation in the gene of keratinocyte transglutaminase 1 enzyme (TGM1). There are 6 gene for Collodion baby: TGM1(14q11), ABCA12(2q34), 19p12-q12, 19p13, ALOXE3-ALOX12B(17p13), ichthyn (5q33).[1,2] The pathophysiology of this disorder consists in an accelerated epidermal turnover with proliferative hyperkeratosis, in contrast to retention hyperkeratosis.[3] The newborn is encased in a Collodion membrane that sheds within 10 to 14 days. The disorder is not life threatening, but in the neonatal period, there is an increased risk for threatening complications such infections, severe dehydration, impaired thermoregulation, respiratory distress, aspiration pneumonia, and malnutrition. Later in childhood disorders of the normal sweat gland function may occur, also ectropion, hearing loss (due to changes in the external auditory canal and tympanic membrane), orthopedic abnormalities.

2. Case report

We report the case of a newborn male baby of 3 weeks age, transferred from Galati Children Hospital with observation of Sepsis and Congenital ichthyosis. The baby was normally born at term, with 3200 g and was the first child of a young and no consanguineous parents. On physical examination the patient was with altered general state and initially hypothermia then fever (39°–40°C), height=53 cm, weight=3500 g, ponderal index =0.89 (first degree malnutrition), dry skin, glossy, with fissures, covered in thick crusts, general infiltrates aspect, cutaneous membranes, umbilical cord mumified, undetached with the granuloma, axillar pustule, severe ectropion, bilateral conjunctivitis, eclabium,

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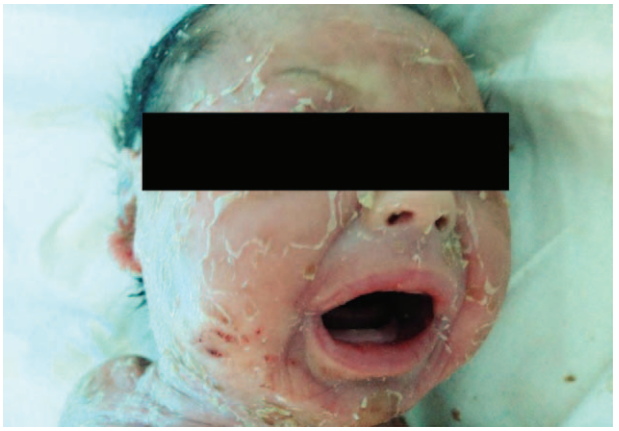

Figure 1. Thick crusts, ectropion, and eclabium on Collodion baby (at admission).

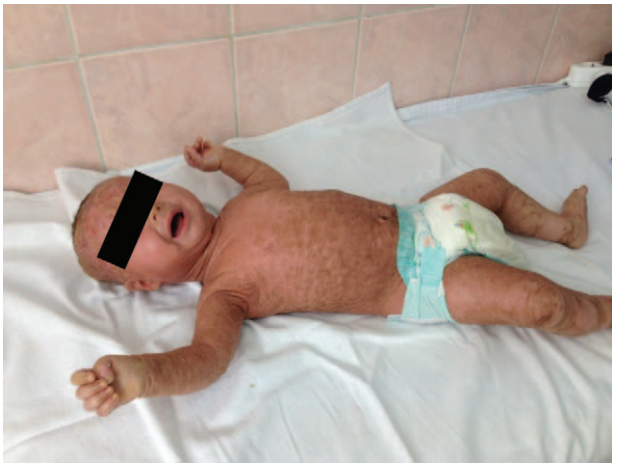

Figure 3. Erythrodermic lamellar ichthyosis at 5 months age.

difficulty in chest mobility and dyspnea with the respiratory rate 60/min, Sat O2–93%, hyporeactivity to external stimuli, but reflexes present (Figs. 1 and 2). Routine blood reveals moderate anemia, high erythrocyte sedimentation rate, leukocytosis, hepatic cytolysis, the altered functional kidney tests, dehydration, hypoproteinemia but no serum immune deficiency. Skin cultures were positive for Enteroccocus spp. sensitive to Ciprofloxacin, Gentamicin, Norfloxacin, Levofloxacin, Tetraciclin, Vancomycin; Candida spp. in the axillar pustule sensitive to Amfotericin B, Itraconazole, Fluconazole, Voriconazole, Clotrimazole, Econazole, Flucytosine, Nystatin, Miconazole; stafilococcus aureus MRSA (umbilical granuloma, skin, external ear canal) sensitive to Oxaciline, Cloramfenicol, Ciprofloxacin, Gentamicin, Novobiocin, Norfloxacin, Tobramycin. Hemoculture-positive for Candida tropicalis, sensitive to Fluconazole, Voriconazole, Caspofungin, Micafungin, Flucytosine. Dermatological and genetic examination confirmed the diagnosis of Collodion baby. The baby associates the interstitial pneumonia and the cardiac congenital anomalies: permeable foramen ovale and arterial canal persistence.

The treatment required multidisciplinary involvement (neonatologist, pediatrician, geneticist, dermatologist, psychologist, ophthalmologist, audiologist). Humid atmosphere and constant temperature in the incubator were required, topic cutaneous emolients after bath with alpha hydroxiacids (glycolic, lactic, citric acids). The collodion membrane should not be debrided. It was a necessary treatment of the dehydration and adequate caloric intake by the parenteral nutrition subsequent enteral nutrition on nasogastric tube, plasma transfusions, and administration of the human albumin. To combat cutaneous inflammation and pain we used topical Methylprednisolone acetonate. Antifungal and antistaphylococcal treatment of sepsis involved the use of the Fluconazole associated with the Vancomycin. Artificial tears were also needed. Evolution was slow favorable with the detachment of crusts and satisfactory weight gain and the patient was discharged after 21 days. In time, however, it developed sensitivity of cow's milk protein with scratching

Figure 2. Collodion baby-dry skin, thick crusts, umbilical cord mumified, undetached with the granuloma.

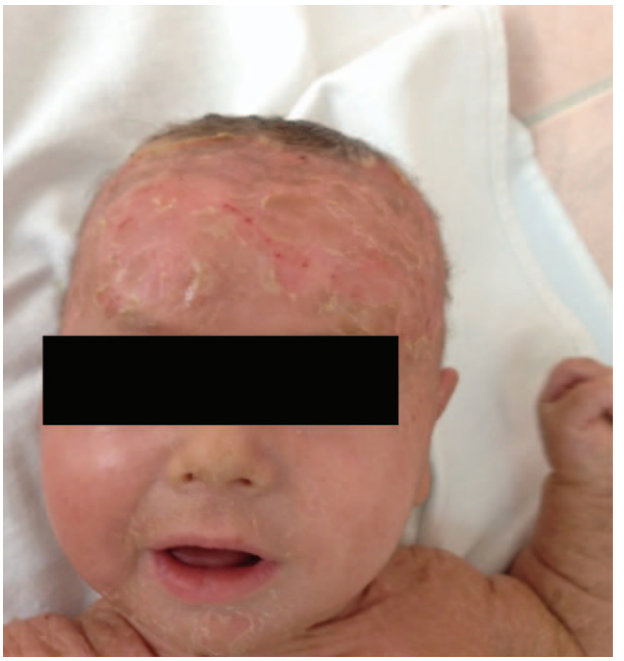

Figure 4. Erythrodermic lamellar ichthyosis at 5 months age, scratching damage on the skin secondary of the milk allergy.

damage on the skin, which required the passage from the hypoallergenic formula to extensive protein hydrolyzate. At the age of 5 months, the skin appearance was erythrodermic lamellar ichthyosis (Figs. 3 and 4).

The authors declare that they have permission from children's parents to publish photos with their child and the case. The ethical approval from The Institutional Review Board was obtained.

3. Discussion

The disease is present at birth and continues throughout life. Diagnosis is suggested by the clinical appearance of the newborn, but skin biopsy correlated with the molecular genetic test is useful in the diagnosis of the lamellar ichthyosis, in the detection of transglutaminase-1 expression, and also in determining the genetic risk for the disease in the family. In our case the family refused this investigation. Collodium baby is an initial presentation of several genetic conditions and long-term evolution depends on underlying condition. Possible outcomes of Collodium baby are nonsyndromic ichthyosis (like congenital ichthyosiform erythroderma, ichthyosis vulgaris, epidermolytic ichthyosis, recessive-X-linked ichthyosis), syndromic ichthyosis (neutral lipid storage disease with ichthyosis, Conradi-Hunermann-Happle syndrome, KID syndrome, ARC syndrome), metabolic disease (Holocarboxylase synthetase deficiency, Gaucher disease type 2), and other (Hypohidrotic ectodermal dysplasia, congenital hypothyroidism). The management of this condition is difficult and requires a team of clinicians.[4] The coexistence of risk factors such as high-risk prematurely, immune deficits, wide-spectrum antibiotics, venous catheter, total parenteral nutrition can induce severe bacterial and fungal infections in neonate.[5–7] In our case although the newborn was in term, with normal weight and without associating the immune deficiency the probability that some risk conditions such as total parenteral nutrition, extended and prolonged antibiotherapy, intravenous devices would favor sepsis with Candida spp., is high. The particularity of our case, however, is the rare neonatal fungemia with Candida tropicalis in conditions where no colonization with this pathogen. Although neonatal fungemia is most commonly caused by candida albicans, in the last years are more frequent reported cases of sepsis with noncandida albicans spp like candida parapsilosis and candida tropicalis, especially in the colonized versus noncolonized patients.[8] In our case, documented bacterial and fungal infection required aggressive treatment according to antibiogram and fungigram. Candida tropicalis spp had azole sensitivity (fluconazole) although some studies reported total or dose-dependent resistance to fluconazole and itraconazole, with the choice of amphotericin B or flucytosine.[8] The duration of the antifungal treatment was 21 days with careful follow-up of liver and kidney function. In 2014, a Canadian study has gone through a severity score that includes the following parameters: generalized involvement, ectropion, eclabium ("fish mouth"), abnormal ears/nares, contractures (fingers), contractures (toes), contractures (upper extremities), contractures (lower extremities), respiratory impairment secondary to skin restriction, erythroderma, edema (hands), edema (feet), edema (upper extremities), edema (lower extremities), hypernatremia; presence=1; absence=0; the maximum score is 15; low severity score=0 to 5; intermediate severity score=6 to 10; high severity score=11 to 15.[9] The final score was 9 which places in an intermediate severity score the baby, which means additional complex of the drug treatment and care measures. The environment in which these children are kept is extremely important, requiring a humid atmosphere, and constant temperature as in an incubator because they can easily dehydrate. For the same reason of risk of dehydration, monitoring of water and electrolyte balance is necessary. Also nutrition and hydration are done parenterally and enterally (nasogastric tube); parenteral administration of human albumin is required as this baby loses protein in large amounts by cutaneous. The collodion membrane must not be debride; they are detaching with time. Although most clinicians find the use of skin emollients useful,[10] some studies show that their use increases the risk of infections; there is also a risk of intoxication with topical products like salicylates and keratolytics with high concentration of urea, by cutaneous absorption through skin alteration.[11,12] In current practice petrolatum-based topical emollients are recommended several times a day which we have also used. Pain management is also necessary and encouraging parents to get involved in child care, psychological support. The association of ectropion requires ophthalmic consultation, because it is very important to protect the exposed eyes with artificial tears. In the case presented the evolution of the disease was complicated by the association of milk allergy that caused pruritus and increased skin erythema increasing the risk of local superinfection, child agitation, capricious appetite, and the diet of the hydrolyzed milk formula was imposed.

4. Conclusion

The Collodion baby is the first expression of some forms of ichthyosis. In these children, skin injuries, total parenteral nutrition, aggressive and prolonged antibiotic therapy, intravenous devices, high hospitalization duration are risk factors for colonization and sepsis with fungi, especially in the neonatal period, sometimes with severe evolution and prognosis.

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