

CLINICAL CASE

Collodion baby

Arturo G. Estrada-López¹, Gloria A. García-Miranda², Dulce M. Meza-Zamora¹

Abstract

Background. Ichthyosis is an infrequent clinical entity worldwide (1:300,000 births). When diagnosed in a newborn, two forms can be identified: collodion baby and its most severe form, harlequin fetus or maligna keratoma. In both cases, clinical manifestations are thick and hard skin with deep splits. The splits are more prominent in flexion areas. Moreover, we can observe ectropion in both eyes and deformities in the ears and nose, as well as inverted lips that resemble the mouth of a fish, among others signs.

Case report. We report a case of a collodion baby who was kept in strict isolation to avoid any infection. However, due to purulent ocular secretions due to coagulase-negative *Staphylococcus aureus*, antibiotic treatment was indicated. The evolution of the patient was favorable.

Conclusions. Mortality of collodion baby has decreased as a result of multidisciplinary care and opportune diagnosis and management.

Key words: collodion baby, harlequin fetus, maligna keratoma, autosomal recessive ichthyosis.

Introduction

The first clinical description of collodion membrane (Pérez, 1880) continues to be valid: "The baby's skin is replaced by a cornified substance of uniform texture, which gives the body a varnished appearance".¹ However, a non-medical description that manages to convey the severity of congenital ichthyosis was found by Waring in 1932 in a diary kept by the Rev. Oliver Hart, pastor at the Baptist church in Charleston, South Carolina, in 1896. The Preface says "a Thursday, April 5, 1750, I went to see the most deplorable condition of a child who was born the night before to Mary Evans, in Charleston. It was a surprise to those who had him and barely knew how to describe him. The skin was dry and hard and seemed to be cracked in many places, in

parts resembled the scales of a fish. His mouth was large and open. He had no external nose, but two holes where the nose should be. His eyes looked like two lumps of coagulated blood, protruding and approximately larger than a "lead", with a cadaveric aspect. He had no outer ears but had holes where the ears should be. The hands and feet appeared to be swollen, were bent and were completely hard. The back of the head was more open. He made a strange kind of sound that cannot be described. He lived ~8-40 hours and was alive when I saw him." (J.L. Waring, 1932, quoted in Cortina et al., 1975).² Since the introduction of the term collodion baby, ~270 cases have been reported in the medical literature.

The most important clinical data concerning collodion baby is the presence of disseminated or generalized ichthyosiform genodermatosis characterized by dry skin, scaling, generalized erythroderma and hyperkeratosis, reminiscent of fish scales. This type of dermatosis is also known by the generic name of ichthyosis.³⁻⁵

¹ Servicio de Neonatología, Hospital General de Tlalnepantla, Edo. de México, México; ² Facultad de Estudios Superiores (FES) Iztacala, UNAM, México D.F., México

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When the infant also has a collodion membrane, the tension that it exerts distorts the features of the face and fingers. Rarely, the shedding of the membrane results in a normal integument because shedding of the membrane results in erythema of varying intensity.⁶

The clinical types of ichthyosis depend on the mode of inheritance as well as clinical and anatomo-pathological data.^{2,4} Ichthyosis can be classified into three groups: 1) true ichthyosis, 2) ichthyosiform states and 3) epidermolytic hyperkeratosis. There are several subtypes of each group.

Among the true ichthyosis are three groups as follows: autosomal dominant ichthyosis (ichthyosis vulgaris, ichthyosis simple, fish skin disease), X-linked recessive ichthyosis (ichthyosis nigricans, ichthyosis of the male, saurodermia) and autosomal recessive ichthyosis (lamellar ichthyosis, non-bullous congenital ichthyosiform erythroderma).⁷ In 1986, Larregue (cited by Van Gysel et al., 2002)⁴ studied four children with ichthyosis and noted 11% mortality and neonatal complication rate of 45%. The majority of these children, who were born as collodion baby, eventually developed ichthyosis on a causal basis. An important outcome of this series is that the frequently reported clinical subtypes: congenital erythrodermic ichthyosis, 48%, lamellar ichthyosis, 12% and ichthyosis vulgaris dominant in 10% of patients. In 10% of cases, the skin eventually developed normally.

The same author reported years later a similar proportion: autosomal recessive lamellar ichthyosis erythroderma in 43% of cases, lamellar ichthyosis, autosomal recessive ichthyosis non-erythroderma in 19%, other forms 12% and normal skin in 25% of cases.⁴

Ichthyosiform states are recognized in the following syndromes: Sjogren-Larsson, Conradi-Hunermann, Rudd, Comel, Tay, Refsum, Netherton, Kid and Bid as well as erythrokeratoderma variabilis of Mendes da Costa and progressive symmetric erythrokeratoderma. Other acquired states are lymphoma, leprosy, malnutrition and medications.⁷

Neonatal ichthyosis, in its most severe form, is known as harlequin ichthyosis, harlequin fetus or maligna keratoma. Harlequin ichthyosis is also a keratinization disorder with extremely rare autosomal recessive hereditary traits. The skin of the affected baby is markedly thick and hard (resembles cardboard) with deep grooves running both transversely and vertically.

Hands and feet are hard and ischemic and there is poor development of the distal digital area. Most babies are premature at between 32 and 36 weeks of gestation. Complications include sepsis, distal gangrene and difficulty feeding and breathing. Aspiration pneumonia of squamous cells in amniotic fluid is a potential complication.⁶ ABCA 12 gene (adenosine triphosphate binding cassette A 12), located at chromosome 2q33-q35, is recognized as the cause of lamellar ichthyosis and mutation of this gene as being responsible for harlequin ichthyosis.⁸

The frequency of collodion baby is very low. It is estimated that there are 1:300,000 cases of newborns in the worldwide.^{3,5,9} In Mexico there are some cases of ichthyosis, although not reported as collodion baby. In a study conducted in the previously known Mexican Institute of Child Care (IMAN), currently Instituto Nacional de Pediatría (INP), a ratio of one case per 3250 patients was estimated.¹⁰ Corona-Rivera, in turn, reported a case of congenital ichthyosis vulgaris in 1995, but the case is more closely related to the presence of trisomy 18 and little was said of the ichthyosis. Characteristics of collodion baby were not reported.¹¹ The incidence of collodion baby as a subtype of ichthyosis has a frequency even lower than this. The only Mexican cases found in the literature are those by Rodríguez et al., who reported on four patients in the south of Veracruz but were presented alluding to autosomal recessive form.³ The case presented here corresponds to the first report of collodion baby in the Tlalnepantla General Hospital "Valle Ceylan" in 35 years of existence and is perhaps the first published case in Mexico. After reviewing 57 years of the *Boletín Hospital Infantil*

de Mexico (1947-2007) and 27 years of *Acta Pediatrica de Mexico* (1980-2007), in addition to searching the databases of Medigraphic, LILACS and ARTEMISA, no reports were found

Clinical case

We present the case of a female infant who was the sole product of 19-year-old parents. The mother had prenatal care from the first trimester of pregnancy with a total of eight visits. During the third month she had a threatened abortion, developed cervicovaginitis at the sixth month and presented premature rupture of membranes 18 h prior to birth. She was then at 38 weeks gestation according to the date of her last menstrual period. She was admitted in labor, ending in vaginal eutocia. The neonate was a product of a gravida 2, para 2 and she cried and breathed at birth. She had an Apgar score of 7–8, birth weight of 2725 g and had Silverman Anderson score of 0. Upon initial physical examination the patient presented with a hard whitish covering that involved the whole body, generalized edema, and eyelids with ectropion, as well as abundant hyaline type ocular secretion. The ears had a dysmorphic appearance due to the keratotic coat, which retracted them (Figure 1) and elabium lips (Figure 2). Skin of the neck and trunk had the appearance of fissures (Figures 3 and 4). The anterior fontanel was without any compromise. Pulmonary fields showed adequate air movement without evidence of wheezing or rales. Rhythmic heart sounds had good intensity without aggregated phenomena. The abdomen was distended due to hepatomegaly of 5 cm below the costal margin. Genitalia were consistent with age and sex, and gestation was estimated at 34 weeks by Capurro. Extremities were complete and symmetrical, although hardened and with poor distal and digital development (Figure 5).

The patient was admitted to the neonatal intensive care unit in isolation, where various laboratory studies were performed in order to rule out congenital infectious process due to the history

of hepatomegaly. TORCH test was requested and was reported as negative. Cultures of blood, stool, urine, and ocular and umbilical secretions and spinal fluid were requested and did not show any bacterial growth. Cytological study of the spinal fluid was also normal. Blood work-up was also performed and reported 25,200 leukocytes,



Figure 1. Large areas of scaly scalp and flattened ears are shown with increase in volume.



Figure 2. Collodion baby at birth. Ectropion, elabium and collodion membrane are noted.

19,900 neutrophils, 2300 lymphocytes, and 1009 bands. Based on these results it was decided to begin an antibiotic regimen using dicloxacillin and amikacin. Skin fissures were covered with mupirocin cream.

The infant subsequently presented purulent discharge from both eyes, prompting another culture of the eye discharge, which isolated coagulase-negative *Staphylococcus aureus*. Results of susceptibility testing justified the addition of chloramphenicol ophthalmic antibiotic therapy.

Due to the high suspicion of ichthyosis, consultation with the Dermatology Department was done and confirmed the diagnosis. Treatment with oleocalcareo liniment was added, as well as increase of fluid intake because of fluid losses and was managed under sterile conditions.

In addition, three punch biopsies were taken and sent to the Pathology Department and were reported as normal because the sample was taken during the shedding stage (Figure 6). To complete the study, families were referred for genetic counseling and karyotype determination; however,



Figure 3. Patient at 48 h. Severe ectropion due to the decrease in humidity of the skin is noted.



Figure 4. Collodion baby at 96 h of birth. Note the presence of cracks and crevices in the skin and flaking in large layers.



Figure 5. Prominent fissures in areas of flexion in limbs, poor development of distal digital area and thickening of the fingers.

the parents did not attend and this study was not available. After 18 days of birth the patient was discharged because of improvement in general condition and was to be followed-up as an out-patient (Figure 7).

Discussion

Collodion baby is similar to an extremely premature infant in terms of the skin barrier due to the high losses of transcutaneous fluid, risk of dehydration and hyponatremia, and skin infections (gram-positive and *Candida* spp.). The seriousness increases due to compressive mechanisms especially over all of the distal extremities and pneumonia secondary to aspiration of desquamated material in the amniotic fluid.^{3,4,12} There are other genetic and physiological deficiencies and patient management will reflect the prognosis.

In the study by Van Gysel et al., babies received treatment on the basis of emollients (petrolatum,

lanolin and cetomacrogolis) as topical prophylaxis. Despite this, systemic infections were reported.⁴ The patient we are reporting on received skin lubrication. Fissures were treated with mupirocine as prophylaxis and no infectious complications were reported. However, the patient presented an eye infection due to coagulase-negative *S. aureus*. Because of this clinical data associated with the presence of bandemia, the patient was treated with a double scheme of systemic antibiotics as well as topical eye treatment without needing to expand treatment.

Taïbeb reported that one of the most important decisions is placing the babies in incubators with humidifiers that vary from 90-100%.¹³ Similar steps were undertaken in this case to avoid risks of skin colonization, maintaining the patient in contact isolation. The incubator was always maintained with humidity >50%. No complications were reported during the hospital stay.

In the case presented here, shedding of the collodion membrane took place approximately on the fifth day of birth. There was no need to use kera-



Figure 6. Scaling in large layers.



Figure 7. Collodion baby at 15 days after admission to the NICU and treatment.

tolytic agents such as retinoids that are used when shedding of the collodion membrane is delayed. Sedation with opioids may be considered in cases where the patient is in pain; however, risks of teratogenicity and toxicity are high.⁶ Other less aggressive topical treatments have been introduced such as those noted by Ümit et al. with N-acetylcysteine. Treatment is controversial because the evidence is inconclusive for its recommendation.¹⁴

When the collodion membrane was shed, a skin biopsy was performed and was reported as normal. As noted previously, the evolution reported in some studies included various types of ichthyosis: congenital ichthyosis erythroderma (43%), lamellar ichthyosis (19%), dominant ichthyosis vulgaris (12%) and normal skin, as in our case (25%). However, surveillance continues in order to observe changes in clinical status.

Children who survive ichthyotic genodermatoses require different treatments and palliative care. It is our obligation to refer patients for typing and timely identification as well as to provide strict surveillance and to refer for genetic support. The progress presented by this patient was satisfactory and similar to that discussed by different authors. It is suggested that these types of cases be followed-up for future publications.

As part of multidisciplinary treatment, children need psychological support as they grow so that they accept their disease. It is also needed to reeducate those persons closest to the patient so that these children can pursue normal activities.²

In conclusion, because this is a rare disease it is indispensable to have very clear and precise information on the steps to follow and the complications that may arise.

Despite its longevity, Tlalnepantla General Hospital has no record of any patient with features similar to the case presented here. Literature review does not offer much information, making it important that these experiences be shared. This patient continues being followed-up under multidisciplinary and inter-institutional surveillance and management.

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Correspondence to: Dr. Arturo G. Estrada López
Servicio de Neonatología, Hospital General de Tlalnepantla
Edo. de México, Mexico
E-mail: centinelaurus@yahoo.com.mx

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