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CASE REPORT

Collodion baby: A report of 4 cases

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Abstract *Introduction:* The term collodion baby refers to a clinical entity noted in newborns who are enmeshed by a translucent, cornified substance like sheets of uniform texture so called (collodion membrane) which gives the whole body surface a varnished appearance. Although, some other diseases and conditions may lead to collodion membrane formation, in almost all the cases the cause is an autosomal recessive ichthyosiform disease.
Case Presentation: The first three cases are all from a consanguineously married couple of Fulani decent. The abnormal appearances of the babies' skin were all noticed at birth. Parents are first cousins. The last case is a female born at GA of 40wks through emergency caesarian section due to poor progress of labor. She was admitted into the Newborn with abnormal skin. These series are crucial so as to enable the pediatrician have a high index of suspicion of its existence and to be equipped with the skills to tackle the numerous complications that follow the disease.
They contribute to mortality and morbidity among children in Nigeria and the exact etiology is unknown. However genetic and environmental factors among others are commonly implicated.
This report is thus aimed at presenting these abnormalities which are rather rare and to highlight that early intervention improves the outcome in patients with these conditions.
Conclusion: Early recognition of this clinical entity and early institution of appropriate therapy can definitely reduce morbidity and mortality in neonates.

Key Words: Collodion Baby, Neonates, Genetic Disorder

Introduction

Collodion baby is a characteristic clinical entity which may precede the development of one of a variety of ichthyoses or occur as an isolated and self-limiting condition. The first clinical description of collodion membrane by Pérez in 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] 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.[2-4] The tension exerted by the collodion membrane, distorts the features of the face and fingers. Rarely, the shedding of the membrane results in a normal integumen because shedding of the membrane results in erythema of varying intensity.[5] The clinical types of ichthyosis depend on the mode of inheritance as well as clinical and anatomic/ pathological data[4,6]. Ichthyosis can be classified into three groups: true ichthyosis, ichthyosiform states and epidemolytic hyperkeratosis.

There are several subtypes of each group. Among the true ichthyoses are three subgroups 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 (laminar ichthyosis, non bullous congenital ichthyosiform erythroderma).[7] 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.

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. 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.[2,4,9] We present these cases to highlight its rarity and need for presentation early to avert complications that follow it.

Case Presentation

Case 1

B.H is a day old male baby, the first child of a consanguineously married couple of Fulani decent. The abnormal appearance of the baby's skin was noticed at birth. Baby was delivered at home but mother presented immediately to the hospital. On examination the baby weighed 2.9kilogram.There was ectropion and the skin was semi transparent and peeling. Systemic examination was essentially unremarkable. Based on these findings, she was managed as collodion baby. The baby was managed in an incubator. The temperature, hydration and electrolyte status were monitored. The baby was managed with prophylactic antibiotics. Adequate nutrition was provided by the use of expressed breast milk.
She received IV fluids and antibiotics. Baby did well and was discharged.

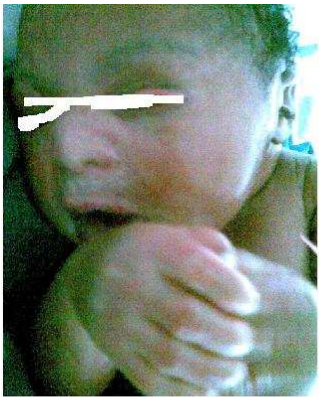

Fig 1: Before Treatment

Fig 1: After Treatment Note the disappearance of all skin features

Case 2

A.H is one day old female baby, the second consecutive child of a consanguineously married couple of Fulani decent. The abnormal appearance of the baby's skin was noticed at birth. Baby was delivered at home before presenting in the hospital. The birth weight at presentation was 2.8kg.
On examination, there was ectropion and absence of eyelashes and eye brows with an O-shaped mouth (eclabium). The skin was semi transparent, and had a parchment like feel with varying degrees of fissures at groin, axilla and joint regions.
The baby however died immediately of sepsis in the hospital.

Fig 2: Pictures of Case 2: Note ectropion on the forehead and collodion membrane on the body

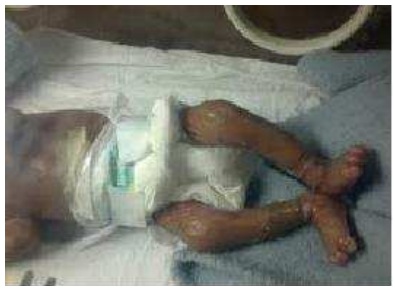

Fig 2: Note similar ectropion on the limbs

Case 3

Baby M.H, is a two day old male neonate, the fifth child in a polygamous family setting of Fulani descent. The parents are first cousins. The mother did not receive adequate antenatal care and the patient was delivered at home by his paternal grandmother. The abnormal appearance of the baby was noticed at birth. The patient is the third consecutive child of the mother with similar presentation at birth.
On examination the baby weighed 2.5kilograms, with a length of 48 cm and an occipito-frontal circumference of 34 cm. There was ectropion and absence of eyelashes and eye brows with an O-shaped mouth (eclabium).
The skin was semi transparent, and had a parchment like feel with varying degrees of fissures at groin, axilla and joint regions. Systemic examination was essentially unremarkable.

The baby was managed in an incubator. The temperature, hydration and electrolyte status were monitored. Bland lubricants were applied to the skin to facilitate desquamation. The baby was managed with antibiotics. Adequate nutrition was provided by the use of expressed breast milk.
The Collodion membrane peeled off within 19 days revealing normal raw skin underneath. Patient was subsequently discharged home in a fair condition. The first of the preceding sibling of the index patient died of severe sepsis due to late presentation while the second

was managed in a teaching hospital and improved remarkably.

Case 4

NR is a female born at 40wks gestation through emergency caesarian section due to prolonged labor. APGAR score was 8/9 and birth weight 3.3kg.
On examination we found a baby with parchment like, taut membrane covering the whole body. There was bilateral ectropion, a flattened nose, an O shaped mouth and dysmorphic fingernails. Other systems were grossly normal.
A diagnosis of collodion baby was made. Full blood count, Serum electrolyte urea and creatinine (SEUCr) were normal. She was nursed in an incubator at high humidity and was given IV fluids, antibiotics, multivitamins and genticine eye drops. Dermatologic and Ophthalmologic teams were invited to review, following which she received keratolytics, moisturizer and sufratyl gauze eye dressing. Blood culture results are negative.

By the 10th day there was appearance of normal skin.
The patient was discharged on 11th day to Ophthalmology and Dermatology Clinic. Unfortunately, she was lost to follow up. It is important to note that none of these patients presented with sepsis, electrolyte imbalance or dehydration (safe the second who died of sepsis) because of the early and vigorous management which was given to them. These involve antibiotics, fluid management and joint management with other specialists.

Fig 1: Pictures of case 4 before treatment

Discussion

The term collodion baby applies to newborns who appear to have an extra layer of skin (known as a *collodion membrane*) that has a collodion-like quality. It is a descriptive term, not a specific diagnosis or disorder (as such, it is a syndrome).[10] This is exactly noted in our patients who had extra layers of transparent skin.
When the Colloidion membrane was shed, the underlying skin appeared normal however a skin biopsy would be performed by the dermatologists to determine the specific type. 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 (25%).[4] However, surveillance and genetic counseling will continue during follow-up. Unfortunately, we lost our patient to follow up. This is because their parents are herdsmen and are always migrating.

The natural course of collodion membrane is intriguing. For instance, approximately 75% of collodion baby will go on to develop a type of autosomal recessive congenital ichthyosis, either lamellar ichthyosis or congenital ichthyosiform erythrodema).[11]
Another 10% of cases the baby sheds this layer of skin and has normal skin for the rest of its life.[12] This is known as *self-healing collodion baby*. This is akin to our series. The remaining 15% of cases could stem from variety of diseases involving keratinization disorders.[11]
Known causes of collodion baby include ichthyosis vulgaris and trichothiodystrophy.[13] Less well documented causes include Sjögren-Larsson syndrome, Netherton syndrome, Gaucher disease type 2, congenital hypothyroidism, Conradi syndrome, Dorfman-Chanarin syndrome, ketoadipiaciduria, koraxitrachitic syndrome, ichthyosis variegata and palmoplantar keratoderma with anogenital leukokeratosis.[13]

There is a striking relationship between ichthyosis and collodion membrane formation. Lamellar ichthyosis may cause collodion baby. In these cases after the collodion membrane peels, the skin is almost completely erythematous and later on an almost generalized desquamation is observed. Soon after, the entire body surface becomes covered by thick scales. Such a thickening of the stratum corneum is called ichthyosis.[14]
Three of our series were associated with consanguinity, the parents being first cousin. It has been noted with interest that since many of these conditions have an autosomal recessive inheritance pattern, they can be associated with consanguinity.[13] Frenk and his cohort, in their study, noted that five spontaneously healing collodion babies recorded in large Swiss kindred all had consanguineous parents. Their distribution in the family indicates autosomal recessive inheritance. At birth they had the typical features of collodion babies. [15]

The major cause of collodion baby syndrome is not well known. However it has been known to be inherited in an autosomal recessive fashion.[16] Placental insufficiency and post maturity have also been implicated in some forms of collodion membrane formation .This could be due to the effects of DNA repair and transcription gene abnormalities in human pre-natal life. Trichothiodystrophy (TTD), a rare (affected frequency of 10^{-6}) recessive disorder caused by mutations in genes involved in nucleotide excision repair (NER) pathway has also been implicated.[16] However our series were all born at term with no placental abnormalities
The Collodion baby is at the risk of high losses of transcutaneous fluid, risk of dehydration, hyponatremia and skin infections (gram-positive and *Candida* spp.). There is also the risk of pneumonia secondary to aspiration of desquamated material in the amniotic fluid.[3, 4, 17]

our first and third cases were managed in a humidified incubator and were given antibiotics but the second series died of sepsis due to late presentation. Taïbeb reported that one of the most important decisions is placing the babies in incubators with humidifiers that vary from 90-100%.[18] Fluid and broad spectrum antibiotics were also used. We did not document any electrolyte imbalance.

In the study by other workers [4, 19] affected babies were treated with emollients as prophylaxis. While one report documented systemic infections, the other study did not. In our two series, we used bland emollients for lubrication and no infection was documented.
Specific practical guides on management in a resource poor country should include This includes half-hourly applications of emollients, frequent oiling of skin, nursing in humidified incubator with careful temperature monitoring, aseptic handling, investigation and treatment of signs of sepsis and, meticulous fluid and electrolyte balance. Constriction bands if present should be divided.
In the third case, shedding of the Collodion membrane occurred by the second week of hospital stay and no keratolytic agents were used.

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.

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References

1. Cortina WJ, Cruz MJ, Villalobos OA, Espinoza MA. Ictiosis congénita (feto arlequín). *Bol Med Hosp Infant Mex 1975; 32:699-702.*
2. O'Connell JP. A collodion baby. *Proc R Soc Med 1977;70:212-13.*
3. Rodríguez GR, Belman PR, Dander AH, Cruz MW. Ictiosis autosómica recesiva: informe de 4 casos del sur de Veracruz. *Bol Med Hosp Infant Mex 2002;59:372-78.*
4. Van Gysel D, Lijnen RLP, Moekti SS, de Laat PCJ, Oranjet AP. Collodion baby: a follow-up study of 17 cases. *J Eur Acad Derm Venereol 2003; 16:472-75.*
5. Monteagudo GL, Aleman PN, Navarro RM. Ictiosis lamelar. Presentación de un paciente. *Medicentro 2005; 9:2-4.*
6. Conlon JD, Drolet BA. Skin lesion in the neonate. *Pediatr Clin North Am 2004;51:863-88*
7. Arenas R. Dermatología. Atlas, Diagnóstico y Tratamiento. México: *McGraw-Hill-Interamericana; 1996. p. 225-30.*
8. Akiyama M. Pathomechanisms of harlequin ichthyosis and ABCA transporters in human diseases. *Arch Dermatol 2006; 142:914-18.*
9. Zapalowicz K, Wygledowska G, Roszkowski T, Bednarowska A. Harlequin ichthyosis□difficulties in prenatal diagnosis. *J Appl Genet 2006; 47:195-97.*
10. Larrègue M, Ottavy N, Bressieux JM, Lorette J. "[Collodion baby: 32 new case reports]" (in French). *Ann Dermatol Venereol 1986; 113: 773–85*
11. Dermatology at the Millennium.By Delwyn Dyall-Smith, Robin Marks, Page 586, Published by Informa Health Care, 1999, ISBN 1-85070-005-2
12. 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*
13. Taïeb A, Labrèze C. "Collodion baby: what's new". *J Eur Acad Dermatol Venereol 2002; 16: 436–7*
14. Shwayder T, Akland T. Neonatal skin barrier: structure, function and disorders. *Dermatol Therapy 2005; 18: 87-103*
15. Frenk E, de Techtermann F. Sehealing collodion baby: evidence for autosomal recessive inheritance. *Pediatr Dermatol. 1992; 9:95-7.*
16. Moslehi R, Signore C, Tamura D, Mill JL , DiGiovanna JJ, Tucker MA et al. Adverse effects of trichothiodystrophy DNA repair and transcription gene disorder on human fetal development. *Clin Genet 2010; 77: 365–373.*
17. Akcakus M, Gunes T, Kurtoglu S, Ozturk A. Collodion baby associated with asymmetric crying facies: a case report. *Pediatr Dermatol 2003;20:134-36.*
18. Taïeb A, Labrèze C. Collodion baby: what's new. *J Eur Acad Derm Venereol 2002; 16:436-37.*
19. Arturo G. Estrada-López, G. García -Miranda A, Dulce M. Meza-Zamora. Collodion baby. *Bol Med Hosp Infant Mex 2010;67: 348-54*