Indian Journal of Trauma and Emergency Pediatrics
Volume 6 Number 3-4, July - December 2014

Colloidal Baby

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Abstract

Colloidion 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 condition is usually a manifestation of congenital ichthyosiform erythroderma (CIE) or lamellar ichthyosis (LI) and is responsible for about 2/3 of colloidion babies. Like a harlequin fetus, a colloidion baby appears to one phenotype for several genotypes. These two lesions may be the first signs of an ichthyosis transmitted as an autosomal dominant trait in about 10% of the cases. Colloidion baby is inherited in an autosomal recessive manner. Sex linked ichthyosis never begins with a colloidion baby syndrome (1–5). The term collodion baby is used for newborns in which all the body surface is covered by thick skin sheets, so called "collodion membrane". The collodion membrane is the result of an epidermal developmental dysfunction.

Key words: Collodion Baby; Ichthyosis.

Introduction

The term collodion baby refers to a clinic entity used for newborns who are encome passed by a translucent, tight and parchment paper like skin sheets so called collodion membrane, on the entire body surface [1, 2, 3]. Collodion baby as a term was first used by *Hallopeau* in 1884 [3, 4, 5]. Since then approximately 270 cases were reported [2, 4]. Although, some other diseases and situations may lead to collodion membrane formation in almost all the cases the cause is an autosomal recessive ichthyosiform disease.

In order of frequency, congenital ichthyosiform erythroderma (especially, nonbullous form), lamellar ichthyosis and harlequin ichthyosis (which are usually accepted to fea turing autosomal recessive inheritance) are responsible. Rarely there may be an asso- ciation with bullous congenital ichthyosiform erythroderma, *Gaucher*'s disease and *Sjögren-Larsson* syndrome. Furthermore, a new form of the disease with an autosomal recessive inheritance called "self healing collodion baby" has been notified where the newborn completely recovers in a couple of weeks. Nevertheless there some other collodion baby causes that have been notified in individual publications [3, 4, 6] (Table 1).

Case History

Our case is 1st product of 3rd degree consanguineous marriage, 39 weeks normal vaginal delivery; delivered at civil hospital, Ahmednagar admitted with complaints of excessive skin peeling and membranous covering all over body.

Table 1: Diseases Associated with Collodion Baby

• Autosomal recessive congenital ichthyoses [lamellar ichthyosis, congenital ichthyosiform erythroderma (nonbullous form), harlequin ichthyosis]
• Epidermolytic hyperkeratose (bullous congenital ichthyosiform erythroderma)
• *Gaucher*'s disease
• *Sjögren-Larsson* syndrome
• Self- healing collodion baby
• Neutral lipid storage disease
• Trichothiodystrophy
• Annular epidermolytic erythema
• Loricrin keratoderma
• X-linked to hypohydrotic ectodermal dysplasia
• Other diseases

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Pathogenesis

The collodion membrane occurs due to an epidermal cornification disorder just like all the ichthyosiform diseases. Although, the pathogenesis of molecular mechanisms apparently lead to an epidermal cornification disorder, keratinocyte protein and lipid me- tabolism defects resulting from autosomal recessive genetic mutations have also been notified as important cofactors [3].

The cause of both autosomal recessive la- mellar ichthyosis and congenital ichthyosi- form erythroderma (nonbullous) have been reported to be transglutaminase 1 gene mu- tation localized on the 14q11 [4, 7]. Moreover, both varied molecular pathogenesis mechanisms and 5 different gene localizations and more than 50 gene mutations in these genes have been detected [8, 9].

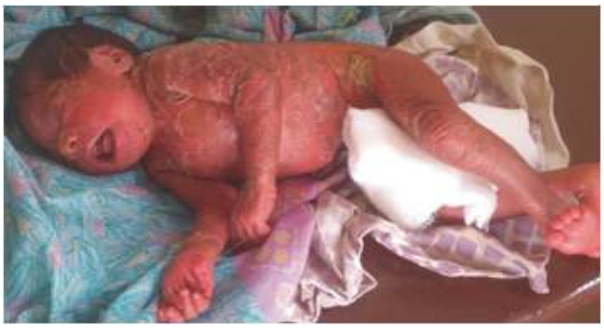
Histopathology

With light microscopic examination of the skin specimens of a newborn baby with a collodion membrane in the early periods an eosinophilic, PAS positive stratum corneum accompanied by hyperkeratosis can be observed. However, the epidermis is weakened due to the thinning of the granular layer. But electron microscopic examination reveals dense intracytoplasmic granules and convoluted corneocytes that can be seen in the upper portion of stratum corneum. Lamellar bodies are numerous but intercellular space and the desmosomes are well preserved. The thinned granular layer is structurally normal [3].

Clinical Features

The collodion babies may be born with a collodion membrane covering the entire skin surface just like an armor (Fig. 1). This situation limits both the baby's respiration and sucking function.

Fig. 1: Collodion baby



The collodion membrane peels off in two or more weeks frequently leaving behind fissures and skin barrier dysfunctions. As a result serious complications like risk of infection, fluid loss, hypernatraemic dehydration, electrolyte imbalance and thermal instability may be encountered. The collodion babies are usually premature at birth. Therefore, these babies should be monitored very carefully [2, 6]. The eyelids and the lips may be everted and tethered (ectropion and eclabion). In such cases loss of proper management can result with keratitis due to xerophthalmia and eventually blindness. While the skin peels off, the residual skin becomes dry and tough. Particularly, tight membrane on limbs may lead to construction and loss of function [3, 6]. While the child grows up, also the symptoms and findings of the premier diseases which have caused the collodion baby appearance begin to arise (Table 1). However, some cases of collodion babies that have healed spontaneously in a few weeks and no associated disease have ever been determined are also been reported [1, 2, 3].

The Clinical Forms of Diseases Often Associated with Collodion Baby

In a long time follow up study of 17 collodion babies it has been reported that, after the membranes peel in a 1–4 weeks period, 7 cases (41%) were determined as congenital ichthyosiform erythroderma, 3 cases (18%) as lamellar ichthyosis, one case as *Sjögren-Larsson* syndrome, one case as epidermolytic hyperkeratosis (bullous congenital ichthyosiform erythroderma) and one case as *Gaucher*'s disease. The rest of the patients (4 cases, 24%) showed any other skin disease [2].

Lamellar Ichthyosis

Lamellar ichthyosis may cause collodion baby. In these cases after the collodion membrane peels the skin is almost com pletely erythematous and later on an almost generalized desquamation is observed. Gradually the size and thickness of the scales increase. Soon after, the body surface becomes covered by thick scales. Especially, the face and the lower legs are involved. Such a thickening of the stratum corneum can bring about a lot of secondary problems. Because of the dysfunction of the sweat glands evaporation can be insufficient and as a result hyperthermia occurs. If the scalp is greatly involved, alopecia areata can be seen and even a hazardous

cicatricial alopecia can occur. Besides these rather more commonly seen unwanted affects deep fissures and extremity contractures can also be observed. Ectropion may lead to development of eye dryness (xerophthalmia) and keratitis and eventu- ally may evolve to blindness [4].

Congenital Ichthyosiform Erythroderma

This clinic entity is usually dealt as bullous and nonbullous congenital ichthyosiform erythroderma in the literature. However, bullous congenital ichthyosiform erythroderma is also named as epidermolytic hyperkeratose by some authors. One of the most important distinct features between these two clinic forms depends on the clinical course. Bullous lesions are characteristic for bullous congenital ichthyosiform erythroderma. In addition to this bullous form shows an autosomal dominant inheritance. On the other hand, nonbullous congenital ichthyosiform erythroderma is the most frequent cause for the collodion baby [2, 3, 4, 5, 6, 10]. The collodion baby membrane exfoliates in time leaving diffuse fine scales covering all the body. But, thick skin sheets cannot be compared with the thick and hard scales of lamellar ichthyosis. However, in the temporal region and the lower parts of legs areas of thick sheets like lamellar ichthyosis can be seen. Palmoplantar skin involvement is also possible and deep fissures may ensue at this part of the skin. But in such cases generalized erythema is much more frequently encountered. Skin barrier dysfunction and secondary calorie loss may cause developmental retardation and short structure. Moreover, hypohidrosis may occur, on account of sweat gland dysfunction [4].

Bullous Congenital Ichthyosiform Erythroderma (Epidermolytic Hyperkeratose)

The mutations that are localized on the keratin 1 and/or 10 genes play an important role in the occurrence of this clinical picture [5, 11]. In severe cases, large bullous lesions, diffuse erythema and a scaly appearance may be seen at birth. The bullous lesions easily rupture and the skin usually shows erosive, erythematous patches. Bullous lesions are frequently painful and hence, they may be troublesome for the infant in the early periods of life. Especially, diffuse erosive lesions may cause risk of high infection. In differential diagnosis, staphylococcal scalded skin syndrome and epidermolysis bullosa should be considered. In the course of the disease, sites of diminishing bullous lesions leave their place to a hyperkeratotic skin layer seemingly a cobblestone configuration. This apperance in times may cover the skin surface. The cobblestone appearance is pre- dominantly because of the deep fissures. The most important feature of this stage of the disease is the predisposition to infection and plenty of different micro-organisms easily inoculated and proliferated in the deep fissures. This gives the baby a peculiar bad odor. Particularly, at adulthood with the activation of the apocrine glands this malodorous condition caused by the micro-organisms can be really annoying for the patients [4].

In this clinic form varied degrees of palmoplantar involvement can also be observed. *Digovanna* and *Bale* according to a study have classified bullous congenital ichthyosiform erythroderma in 6 groups. Three groups showed palmoplantar involvement, while the other 3 groups did not [10].

Harlequin Ichthyosis

Harlequin ichthyosis is the most severe and striking form of ichthyosiform diseases. These collodion babies are born covered with a thick skin sheets resembling an armor. In addition, there are deep fissures on the skin. The ears and the nose are flattened, because of the loss of skin elasticity. The everting of the eyelids and mouth may cause a remarkable and almost a specific appearance for this clinical entity. Harlequin ichthyosis is a rare and probably because of an autosomal recessive inheritance. In a study consisting of 10 harlequin babies, the pathogenesis of the disease has been reported to be the result of both struc tural and functional default of keratin, filaggrin and the lamellar body [3, 12]. Keratin, filaggrin and lamellar body are the main elements of stratum corneum. Also, in an- other study, the occurrence of serintreonin protein phosphatase enzyme deficiency re lated protein phosphatase gene mutations localized on 11^{th} chromosome were notified as another possible cause of this hazardous disease [3, 13].

Treatment

In collodion babies fluid and electrolyte balance and body temperature must be carefully monitored. In addition to this the membrane must be lubricated and to achieve elasticity and desquamation an

adequate hydration of the skin are the major components of management. Suitable eye care and pain control should be carried out for the collodion babies with ectropion. Humidified incubators and water dressings followed by emollient agents are the essentials of the management. If there is a respiratory failure ventilative support for the collodion babies may surely be needed. In cases of epidermolytic hyperkeratose (bulous congenital ichthyosiform erythroderma) which show generalized erythema, bullae and erosions an antibacterial will be needed among the standard therapies. The collodion babies with large areas of skin erosions are always under the risk of heavy infections an even sepsis therefore suitable local and systemic antibacterial agents must be cautiously determined and preferred [6]. The drugs such as salicylic acid, lactic acid and propylene glycol may be applied in order to remove the hyperkeratotic sheets from the skin. But in such cases with generalize lesions, particularly in newborns it must not be forgotten that the application of salicylic acid locally in extreme doses may cause salicylic acid toxemia. Therefore, local remedy in these cases should be cautiously monitorized and carried out in this way [4].

However, in the collodion babies with localized lesions local retinoic acid and calsipotriol treatments have been reported to be successful [14]. Systemic retinoids which are currently a preferred treatment method giving impressive results in cases with generalized lesions have been notified as a perfect alternative. When systemic retinoids came into the practice the mortality rates decreased expressively. In a study, acitretin has been used at the dose of 0.5–0.75 mg/kg/day and the mortality rate of collodion babies has decreased to 11% (1986) compared to the numbers of 1960 (50%). In cases of lamellar ichthyosis, systemic retinoids have been begun at doses of 0.5 mg/kg/day and later on the doses have eventually been increased to 2 mg/kg/day. It is notified by the authors that the greatly thick scales have improved expressively. Systemic retinoids have also shown to have been effective for cases with congenital ichthyosiform erythroderma.

Although, harlequin fetus is rare among all the ichthyosiform diseases, without any dispute it is the severest form. Hence, high mortality rates have been observed. Nonetheless, in recent years, these rates have also been declined because of the entrance of systemic retinoids into the clinical practice and the advanced care methods which are used in intensive care units [4].

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