

A Rare Case Report of Lamellar Ichthyosis “Collodion Baby” with Severe Cicatricial Ectropion in Both Eyes



Medical Science

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ABSTRACT

The purpose is to report the rare occurrence of collodion baby among population of Central India. A 5 months old male child, first issue of a second degree consanguineous married couple, FTND in hospital, NICU admission for first 10 days after birth, c/o eversion of both upper & lower eyelids of both eye since birth. O/E exfoliated lesions present over scalp, palms, soles, fish mouth like oral cavity 'eclabium', eyebrows not developed, upper lids showed total cicatricial severe ectropion, lower lids showed mild ectropion, tarsal conjunctiva was exposed & bulbar conjunctiva exposed & keratinised, follows torch light, rest anterior segment WNL, fundus both eyes was within normal limits. Frequent lubricating eyedrops & antibiotic eyedrops were prescribed and baby was called for follow up. COLLOIDION BABY has rare incidence in Central India. One such case was reported in our institute.

INTRODUCTION

The word ichthyosis; is derived from a Greek word 'ikthus' meaning fish. Ichthyosiform dermatosis, are a group of hereditary disorders characterised by dryness and roughness of the skin with excessive accumulation of epidermal scales. Lamellar ichthyosis, is one of the rare congenital ichthyosiform dermatoses. The characteristic feature of the disease is a thin, dry, shining, brownish-yellow parchment-like membrane which completely envelopes the newborn. This gives a collodion or “baked apple” look to the newborn and such children are called “collodion babies”. This condition was first described by Seelingman in 1841. Ballantyne was the first author to make an extensive report on 33 cases. Till 1968 a total of 103 cases had been reported in world literature. We are reporting a case of lamellar ichthyosis, with bilateral ectropion of the eye lids in addition to the generalised involvement of the body surface by a membrane.

CASE REPORT HISTORY

A 5 months old male child resident of Nagpur, Muslim by religion, born of consanguineous marriage referred from private practitioner came to eye OPD with chief complaints of exfoliation of skin since birth, bleeding from exfoliated sites since birth and eversion of both upper and lower lids with watering from both eyes.

Mother was ANC registered case, regular ANC visitor. She received Iron & Folic acid tablets and 2 doses of TT injection. There is no h/o pregnancy induced hypertension, gestational diabetes mellitus or rash during pregnancy. No history of any ANC complications.

The child was full term normal vaginally delivered, cried immediately after birth with birth weight of 2 kgs. There is history of neonatal intensive care unit admission after birth for 10 days details of which are not available. Child is not immunized as per schedule and had received treatment from private practitioner for skin lesions, details of which are not available.

There is no history of any similar complaints in family, baby is born of consanguineous marriage and is first child. His bowel and bladder habits were normal and accepts feeds well.

On examination, general condition was fair, baby was hypothermic with heart rate 130 per minute, respiratory rate 28 per minute with no signs of distress.

Baby’s anthropometry parameters were as given in the table below :
ANTHROPOMETRY

	Observed	Expected	Percentile
Weight (kg)	2	6.9	<3 %
Height (cm)	46	64	<3 %
Head Circumference (cm)	32	41.5	<3 % rd
Chest Circumference (cm)	30cms		

On general examination, no pallor, icterus, cyanosis, clubbing or lymphadenopathy was present. Exfoliative lesions were present all over the body including scalp, palms & soles. Left limbs were in flexed posture. Toes & fingers were not well separated. The oral cavity was fish mouth shaped.

On systemic examination, cardiovascular system, respiratory system and central nervous system were found to be normal. Abdomen was soft, non- irritable with liver & spleen not palpable.

EYE :	OD	OS
LIDS	<ul style="list-style-type: none"> • Eyebrows not developed. • Eyelashes matted with discharge & mucus. • Upperlids : Total cicatricial severe ectropion • Lowerlids: Partial mild ectropion • Tarsal conjunctiva exposed 	<ul style="list-style-type: none"> • Eyebrows not developed. • Eyelashes matted with discharge & mucus. • Upperlids : Total cicatricial severe ectropion • Lowerlids: Partial mild ectropion • Tarsal conjunctiva exposed
V/A :DISTANT VISION	Follows torch light upto distance of 1ft	Follows torch light upto distance of 1ft
CONJUCTIVA: BULBAR	Palpebral: Exposed due to eversion & keratinisation of conjunctiva. Bulbar : Normal clear. no e/o congestion	Palpebral: Exposed due to eversion & keratinisation of conjunctiva. Bulbar : Normal clear. no e/o congestion
CORNEA :	BRIGHT	BRIGHT
SCLERA:	CLEAR	CLEAR
AC:	NORMAL DEPTH	NORMAL DEPTH
IRIS :	COLOUR PATTERN NORMAL	COLOUR PATTERN NORMAL
PUPIL : DIRECT & CONSENSUAL LIGHT REFLEX	NSRTL INTACT	NSRTL INTACT

EYE :	OD	OS
LENS :	ISC	ISC
IOT :DIGI-TALLY SCOTZ TON:OMETRY	DFN	DFN
OM	WITHIN NORMAL RANGE	WITHIN NORMAL RANGE

Fundus examination of both eyes was within normal limits.

CLINICAL PHOTOGRAPHS OF THE BABY



With these findings clinical impression of collodion baby with severe total upper lid ectropion of both eyes with mild ectropion of lower eyelid of both eyes was kept.

Baby's investigations were as follows :

CBC, WBC	:	1700/cumm
Electrolytes	:	Na : 143mEq/L
		K : 5.5 mEq/L
Total Bilirubin	:	8 mg/dl
Sr. Calcium	:	11.2 mg/dl
C-reactive protein	:	1.8 mg/L

TREATMENT -

Baby was given appropriate IV fluids with close monitoring of electrolytes in NICU by pediatrician, baby bath, thermoneutral environment with humidified incubator, skin emollients, prophylactic tobramycin 0.3% eye drops 6 hourly for 15 days, 0.5% Carboxymethylcellulose eyedrops 2 hourly to prevent dryness of eyes and tear substitute eye ointments at bed time.

DISCUSSION

In lamellar ichthyosis there is presence of disseminated echthysiform genodermatosis, dry skin, scaling, generalized erythroderma and hyperkeratosis. Eyes and mouth are open due to tightness of the skin. The collodion membrane desquamates within 2 weeks frequently leaving behind fissures and skin barrier dysfunction. Baby can develop complications like hypothermia, hypernatremic dehydration, skin infections and toxicity, fissures, conjunctivitis, lagophthalmos, sepsis, distal ischaemia and aspiration pneumonia. In long term > 75% of babies go on to develop a type of AR congenital ichthyosis and around 10% babies shed collodion membrane and has normal skin for its life.

CONCLUSION

Collodion baby has a rare incidence all over world. One such case was reported in tertiary eye care centre of central India.

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