

Reporting a rare case of ectropion associated with Collodion baby.



Ophthalmology

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ABSTRACT

A rare case of collodion baby is being reported with severe form of ectropion bilaterally.

Introduction

Collodion baby is an uncommon clinical presentation of several genetic conditions, primarily disorders of cornification. Ichthyosis is an infrequent clinical entity worldwide (1:300,000 births). The word Ichthyosis; is derived from a Greek work '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. The characteristic feature of the disease is a thin, dry, shining, brownish-yellow parchment-like membrane which completely envelopes the new born. This gives a collodion or "backed apple" look to the new born and such children are called "collodion babies". Almost always associated with ectropion and eclabium. This condition was first described by Seelingman in 1841.

Collodion baby is a phenotype resulting from different congenital disorders of keratinization. Most affected individuals later have development of classical lamellar Ichthyosis or non bullous congenital ichthyosiform erythroderma. Although other syndromes such as sjogren larssen syndrome, infantile gaucher's disease etc. can be causative.

CASE REPORT

A 24-year-old lady delivered a full term 2.7 Kg female baby by LSCS. New born was covered all over with a tight, shiny yellowish membrane. Due to tautness of the membrane the baby's eyes were remaining open with marked ectropion of the lids of both eyes and the mouth was fixed in an open position [Figure - 1]. The membrane at neck region cracked open by few hours of birth. On examination of the eyes severe bilateral ectropion were seen at time of birth with congestion of the conjunctiva but no corneal or conjunctival defect. Treatment was initiated with frequent light massage of the entire body surface with white soft paraffin. Tobramycin eye drops 2 hourly, Lubrex eye drops 2 hourly given alternately and plastic protective eye shield was given to the new born to prevent any form of damage to the eye (exposure keratitis etc. A wet saline gauze was lightly placed over the face covering the eyes totally. After two weeks of birth, the condition improved and the child could open his eyes partially [Figure 2]. The recovery was asymmetrical; the left eye responded better than the right eye. However, due to tautness of the membrane, the baby's eyes remained open. The mother was advised to continue the same treatment. Frequent follow-ups were done to detect any complication. Further improvement was seen after 1 month in both eyes (Figure 3). Tobramycin eyedrops was replaced by Apdrops eyedrops for 2 hourly which was stopped after 1 month along with Lubrex eyedrops 5 times and Eyemist eye gel twice a day.

Discussion

Lamellar Ichthyosis is known to be an inherited autosomal recessive disorder. Though not a feature in our case, 8% of such cases give a history of first degree consanguinity. Similarly, our case was a female child though the disease is reported to be two times more common in

males. 25% of such children born premature and 51% have similarly affected siblings. Our case was born at full term and doesn't have a similarly affected sibling. Although 80% patients show a generalised involvement, others have the disease limited either to the trunk or one or two extremities. Flexor aspects of the body are most severely affected. The other reported associations of the disease are bilateral ectropion (33%), diminished or absent sweating (10%), nail dystrophies (less than 5%) and seasonal recurrence of the dermatosis in summer (15%). These children are extra susceptible to systemic infection. Alternate formation and shedding of scales on the skin from time to time has been described. The histopathology has been discussed and marked hyperkeratosis, normal to thickened granular layer on occasion prominent rete ridges was found. These children are also susceptible to hypothermia and hypernatraemic dehydration.

Follicular orifices are filled with keratin and hair follicles and sebaceous glands are smaller than normal. There is universal agreement regarding management 3 of such cases with plain ointments, keratolytic preparations and in severe cases administration of systemic and topical corticosteroids.

The present case illustrates the severity of ectropion of eyelids and high-lights the importance of proper treatment to prevent exposure keratopathy

Conclusion

The diagnosis of the collodion baby is clinical and management is conservative. Detailed history of the patient should be obtained for better management. Collodion baby is susceptible to infections so care must be taken and minimal handling should be done. Naso Gastric feed was given in our case to prevent aspiration. Along with Naso Gastric feed, IV fluids, proper antibiotics, were given. One should not hesitate in upgrading the antibiotics depending on septic screening. Management of collodion baby requires a team comprising of paediatrician, ophthalmologist and dermatologist.



Figure 1 : Bilateral upper lid ectropion with collodion membrane



Figure 2: Partial resolution of ectropion after 2 weeks



Figure 3: Further improvement seen after 1 month

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