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Case Report

Bilateral ectropion in a 3 months old baby with lamellar ichthyosis: a rare case report

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ABSTRACT

Ichthyosiform dermatoses are a group of hereditary disorders characterized by dryness and roughness of the skin with excessive accumulation of epidermal scales. Four main types of Ichthyosis are Ichthyosis vulgaris, x-linked recessive, lamellar ichthyosis and epidermolytic hyperkeratosis. Lamellar ichthyosis is the rarest form with an incidence of 1 in 3, 00,000. It has autosomal recessive inheritance. Ocular manifestations include exposure keratitis secondary to ectropion, unilateral megalocornea, and enlarged corneal nerve, and blepharitis, absence of the meibomian gland, trichiasis, madarosis and absence of lacrimal puncta. Ectropion of both upper and lower lids have been documented. We are reporting a rare case of lamellar ichthyosis with bilateral upper eyelid ectropion in a child.

Keywords: Lamellar ichthyosis, Ectropion, Collodion membrane

INTRODUCTION

The word ichthyosis is derived from a Greek work 'Ikthus' meaning fish. Lamellar Ichthyosis (LI) is an autosomal recessive disorder that is apparent at birth and is present throughout life. The new-born is born encased in a collodion membrane that sheds within 10-14 days. The shedding of the membrane reveals generalized scaling with variable redness of the skin. Lamellar Ichthyosis, is one of the rare congenital ichthyosiform dermatoses.[1]

Congenital eversion of the upper eyelid was first described by Adams in 1896 and was termed 'double congenital ectropion'. The condition is usually bilateral but unilateral cases have been reported. We are reporting a rare case of lamellar ichthyosis with severe bilateral upper eyelid ectropion in a child.

CASE REPORT

A 3 month old boy came into the Dermatology OPD with chief complaints of dryness of skin, scaling, fissuring at places and itching. On close examination the skin of the boy was characterised by extensive scaling which resembled "fish scales". This scaling was present over the entire body including the head, face, groin, back and joints. The teeth, hair and nails were normal. The child's mother gave the history of this condition being present since birth. On being asked further she revealed that the child was born full term, weighed 2.5 kg and there was a history of consanguineous marriage. The mother was a primi gravida.

A preliminary diagnosis of congenital lamellar ichthyosis was made and it was confirmed after a skin biopsy from the right groin which showed orthokeratosis along with thickening of stratum corneum and stratum granulosum consistent with the condition.

The child was referred to ophthalmology for mild conjunctival discharge along with severe bilateral upper lid ectropion. On examination with a slit lamp which showed conjunctival and corneal xerosis with bilateral upper eyelid ectropion. No other ocular abnormality was detected. The child was managed conservatively with

topical keratolytics, 3% salicylic acid applied twice daily. Emollients were applied two to three times a day. Oral isotretinoin 10mg given once daily. Secondary infection was controlled with appropriate antibiotics. Hourly application of methyl cellulose eye drops 1%, moxifloxacin eye drops 0.5% four times a day and hydroxy methyl cellulose ointment 2% at night was given.

Figure 1(a): "Fish" like scales involving entire body.

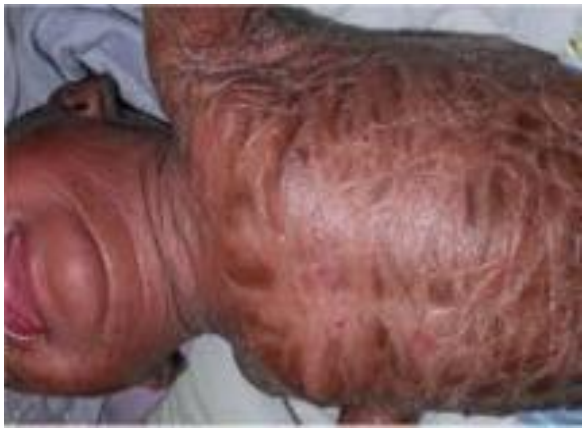

Figure 1(b): "Fish" like scales involving entire body.

A further systemic examination revealed no other abnormality other than a mild nutritional deficiency. The child's mother was further counselled about the child's condition and was referred to a higher centre for further management of the severe ectropion.

Figure 2(a): Severe upper lid ectropion.

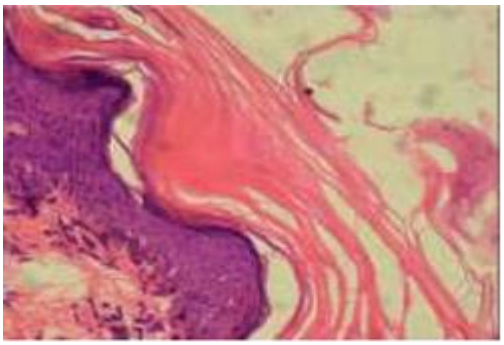

Figure 2(b): Histopathology showed hyperkeratosis, hypergranulosis and elongation of rete ridges.

DISSCUSION

Ichthyosis is a skin disorder characterized by excessive dryness of the skin and increased formation of epidermal scales. Four main types of Ichthyosis are Ichthyosis vulgaris, x-linked recessive, lamellar Ichthyosis, and epidermolytic hyperkeratosis. Lamellar Ichthyosis is the rarest form with an incidence of 1 in 300,000. It has autosomal recessive inheritance.[2]

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". 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.[3]

Patients with lamellar Ichthyosis are typically born encased in a translucent collodion membrane, which is replaced over the first month of life with generalized scale that is accentuated in flexural areas as well as on the forehead and lower extremities. Eclabium, ectropion and scarring alopecia at the periphery of the scalp are often observed as sequelae of excessively taut skin and heat intolerance frequently occurs due to obstruction of the sweat ducts by plates of scales.

About 8% of such cases give a history of consanguinity which is also a feature in our case. The disease is reported to be two times more common in males, also a feature of our case. 25% of such children are born premature and 51% have similarly affected siblings. Although 80% patients show a generalised involvement, as in our patient, 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 infections.[4]

Ocular manifestations include exposure keratitis secondary to ectropion, unilateral megalocornea, enlarged corneal nerves, blepharitis, absence of the meibomian gland, trichiasis, madarosis and absence of lacrimal puncta. Ectropion of both upper and lower lids have been documented.[5]

Ectropion in lamellar ichthyosis may respond to conservative management in the early period of life in about half of the cases.[2]

It is important to treat these children early in order prevent exposure keratopathy in order to maintain a good visual acuity for adult life.

Although the disorder is not life threatening, it is quite disfiguring and causes considerable psychological stress to affected patients. Hence, these conditions require a combined team of Dermatologist, Ophthalmologist, Paediatrician and a child psychiatrist in order to treat, counsel as well as rehabilitate the patient to the best of their efforts.

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