

## Spontaneous subconjunctival abscess in congenital lamellar ichthyosis

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Congenital lamellar ichthyosis is an autosomal recessive, heterogeneous disorder presenting at birth with generalized skin involvement. The most common ophthalmic manifestation noted is bilateral ectropion of the lower eyelids. A 1-month-old female neonate, the second born of a nonconsanguineous marriage, presented with 4 days' history of redness, discharge, and swelling in the right eye. There was severe right upper eyelid ectropion, conjunctival injection, chemosis, a subconjunctival mass on the temporal bulbar conjunctiva spontaneously draining pus and corneal haze. The anterior chamber, iris, lens and fundus appeared normal. Congenital lamellar ichthyosis was suspected because of scaling and excessive dryness of the entire body. The occurrence of a spontaneous subconjunctival abscess is not known in lamellar ichthyosis. We thus report the management of a rare case of unilateral upper eyelid ectropion, subconjunctival abscess with orbital cellulitis in congenital lamellar ichthyosis.

**Key words:** Lamellar ichthyosis, ocular manifestations of lamellar ichthyosis, subconjunctival abscess

Ichthyoses are a group of heterogeneous, hereditary disorders characterized clinically by excessive scaling and thickening of the skin.<sup>[1]</sup> Congenital lamellar ichthyosis, an autosomal recessive condition, is the rarest with a prevalence of 1 in 300,000.<sup>[2]</sup> The main ocular manifestations as noted by Arnold in 1834 were bilateral lower eyelid ectropion, exposure keratopathy, and corneal scarring.<sup>[3]</sup> The frequency of occurrence of ectropion is estimated at 45%–80%.<sup>[4]</sup> We report a rare case of severe right upper eyelid ectropion, subconjunctival abscess with orbital cellulitis in lamellar ichthyosis.

### Case Report

A 1-month-old female neonate presented to the pediatric department with redness and discharge associated with a swelling in the right eye which had progressively worsened over the preceding 4 days. She was the second born of a

nonconsanguineous marriage at 36 weeks of gestation through a normal vaginal delivery with no perinatal trauma. The mother had no adverse obstetric history. At birth, her entire body was covered with a transparent, dry, parchment-like shiny membrane with mild erythema. Thus, a clinical diagnosis of congenital lamellar ichthyosis was made. The neonate was put on topical hypromellose 2% gel 2 times/day due to the risk of dry eye. On examination, multiple scales were present over the chest, abdomen, and all the four limbs with wrinkling [Fig. 1]. The right eye showed Grade 3 upper eyelid ectropion with thickening of the anterior lamella, the skin over the eyelid being erythematous with small scales with severe lagophthalmos, leading to an increased risk of dry eye. The lower eyelid showed Grade 1 ectropion laterally. On gently separating the lids' conjunctival chemosis, congestion was seen with a subconjunctival mass measuring 10 mm × 5 mm on the temporal bulbar conjunctiva spontaneously draining pus through a central opening, the posterior extent of which could not be made out [Fig. 2]. The cornea appeared hazy, measuring 10 mm in diameter horizontally. The anterior chamber was well formed, iris was normal in color and pattern, pupil was 2 mm round, sluggishly reacting to both direct and indirect light reflexes, and lens appeared clear. Restriction of extra-ocular movements was seen in all gazes with no evidence of proptosis.



**Figure 1:** Multiple scales over bilateral upper limb, lower limb, chest, and abdomen with wrinkling

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**Figure 2:** Right eye examination – A subconjunctival mass measuring 10 mm × 5 mm on the temporal bulbar conjunctiva spontaneously draining pus with Grade 3 ectropion of the upper eyelid

The left eye examination showed Grade 1 ectropion of the lateral third of the lower eyelid with no involvement of the upper eyelid or the underlying conjunctiva. On B-scan ultrasonography, an ill-defined soft-tissue mass measuring 3 cm × 1.9 cm involving the temporal aspect of the right globe with increased vascularity and areas of necrosis was seen suggestive of an abscess. The discharge on culture sensitivity showed methicillin-resistant staphylococcus growth sensitive to moxifloxacin. Based on the above clinical features, a diagnosis of lamellar ichthyosis with right eye spontaneous subconjunctival abscess with orbital cellulitis with upper eyelid ectropion was made. The neonate was started on intravenous ceftriaxone, amikacin with frequent topical instillation of carboxymethylcellulose, and moxifloxacin 0.5% eye drops 6 times/day. Given that the abscess was already draining, we elected to withhold immediate surgical treatment. Good patient compliance made close outpatient follow-up possible. The child was treated for the skin lesions with lubricating and keratolytic agents. After 2 weeks of starting treatment, there was complete resolution of the subconjunctival abscess [Fig. 3]. On the 1<sup>st</sup> monthly follow-up Grade 2 cicatricial ectropion of the right upper eyelid, Grade 1 ectropion of the lateral third of the lower eyelid with a macular grade corneal opacity was noted [Fig. 3].

## Discussion

Ichthyoses are a group of inherited heterogeneous disorders. Based on the hereditary pattern and clinical and histological findings, the congenital ichthyotic disorders are divided into the following main types: ichthyosis vulgaris, X-linked ichthyosis, and autosomal recessive primary ichthyosis.<sup>[5]</sup>

Autosomal recessive primary ichthyosis is further divided into lamellar ichthyosis and congenital ichthyosiform erythroderma. Neonates with lamellar ichthyosis are affected from birth, born as collodion babies. Clinically, it is the rarest and most severe form of the autosomal recessive ichthyoses, the estimated incidence is 1:300,000 live births.<sup>[6]</sup>

Affected neonates are embedded in an inelastic membrane at birth which later on drying is associated with an increased



**Figure 3:** Two weeks' follow-up: Complete resolution of the subconjunctival abscess with Grade 2 cicatricial ectropion of the right upper eyelid and Grade 1 ectropion of the lateral third of the lower eyelid with a macular grade corneal opacity

risk of dehydration and infection. As the membrane gradually begins to exfoliate a few weeks after birth, it is replaced by a generalized scaling rash involving the skin flexures, palms, and soles. It is not associated with erythema or bullae formation.<sup>[7]</sup>

The most common ocular manifestations previously described with congenital lamellar ichthyosis include eyelid abnormalities, bilateral ectropion of the lower eyelids being the most common.<sup>[8]</sup> The other findings include exposure keratitis secondary to ectropion,<sup>[9]</sup> megalocornea, absence of the meibomian gland, lacrimal puncta, trichiasis, and madarosis. Deep stromal corneal opacities are more commonly seen with the X-linked recessive form.<sup>[3]</sup> Mucosal surface involvement including that of the conjunctiva is not a recognized association of lamellar ichthyosis.<sup>[3,6]</sup>

Our case is unique, since there is a unilateral involvement with ectropion of the upper lid with marked thickening of the anterior lamella and the conjunctiva shows a spontaneously draining subconjunctival abscess with no known risk factors.

There are a few reports describing the occurrence of a subconjunctival abscess. It is a rare condition, mostly associated with a history of trauma, infectious scleritis, or surgery.<sup>[10]</sup> Following aggressive treatment with intravenous antibiotics, the abscess and orbital cellulitis completely resolved negating the need for any surgical intervention.

## Conclusion

Patients with bacterial conjunctivitis associated with lamellar ichthyosis should be closely examined to rule out a subconjunctival abscess which may require aggressive medical or surgical management to prevent vision-threatening complications and sequelae.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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#### Conflicts of interest

There are no conflicts of interest.

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