Bilateral viral keratitis in lamellar ichthyosis: A rare ocular manifestation

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Key words: Bilateral HSV keratitis, lamellar ichthyosis, viral keratitis

Congenital ichthyosis is a rare group of genetic skin keratinization diseases characterized by generalized scaling and a variable degree of erythema and hyperkeratosis. Lamellar ichthyosis (LI) is the rarest form with an incidence of 1 in 500,000. Previous reports of ocular involvement in LI includes ectropion of both lids which leads to subsequent corneal exposure, dry eyes and even microbial keratitis. Herein, we report an unusual presentation of bilateral HSV keratitis in LI.

An eight-year-old boy with lamellar ichthyosis (LI), presented with complaints of photophobia in both eyes. He was on tablet acitretin 10 mg OD for the past five years which he stopped in the previous month, as per the advice of treating physician and was also using lubricants regularly. Presenting visual acuity was 6/6 in both eyes. On examination, he had bilateral severe upper lid ectropion and moderate lower lid ectropion with generalized scaling of skin. Since the child was photophobic, eyes were examined under sedation, which showed bilateral congestion with sparse and coalesced punctate epithelial lesions in the right eye and left eye, respectively [Fig. 1a]. He was treated with sodium hyaluronate eye drops, ointment and antibiotic drops. On one-week review, patient worsened symptomatically with increasing photophobia. Suspecting dry eye related inflammation, 0.5% loteprednol eye drops were added three times per day. Though there was symptomatic improvement in the following days, one week later, the mother noticed whitish lesion in both eyes. Repeat examination under sedation revealed bilateral geographic ulcer 5 × 4 mm with active margins. An empirical diagnosis of herpes simplex virus (HSV) keratitis was made and he was started on ganciclovir eye ointment 0.15% five times a day along with lubricants and antibiotic drops [Fig. 1b and c]. Since there was pre-existing compromised ocular surface due to bilateral ectropion, amniotic membrane grafting was done in both eyes for non-healing ulcers and tablet valacyclovir 500 mg OD was given for the next one month. Complete resolution of the epithelial defect was seen in three weeks, and at six-month follow-up, both eyes showed nebular corneal opacity with visual acuity of 6/12. One year after the primary episode, patient presented again with bilateral redness and watering.

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for two days [Fig. 2a]. Slit-lamp examination revealed stippled epithelial lesions (similar to the first episode) in both eyes. The tear film was subjected to polymerase chain reaction (PCR) for HSV-1 DNA, which tested positive. The patient was successfully treated with topical ganciclovir five times a day with complete resolution in two weeks. He was advised regular usage of preservative-free topical lubricants four times a day along with ointment at night.

**Discussion**

The initial presenting symptoms of our patient led us to a provisional diagnosis of dry eye secondary to ectropion. However, the worsening of symptoms with topical steroids alerted us and the characteristic geographic ulcer helped us initiate appropriate antiviral treatment. The second episode of viral keratitis was treated without delay and the PCR test aided in making an established diagnosis of HSV keratitis. Some of the key learnings are that classical dendritic lesions are typically absent in the initial stages probably due to the altered ocular surface, and prompt early treatment is especially important in these patients due to poor epithelial healing.

Hence a high degree of clinical suspicion for viral keratitis is necessary while treating patients with lamellar ichthyosis. PCR can be a good adjunctive in confirmatory diagnosis, especially in the absence of classical clinical picture.

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**Conflicts of interest**

There are no conflicts of interest.

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