Vernal Keratoconjunctivitis spectrum in two generations of a family

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Key words: Burnt-out Limbal Vernal Keratoconjunctivitis, limbal stem cell deficiency, Limbal Vernal Keratoconjunctivitis

Limbal Vernal Keratoconjunctivitis (VKC) is one of the known causes of Limbal Stem Cell Deficiency (LCSD)[1] and has recently gained importance because of the long-term implications of chronic limbal inflammation being documented.[2] We report 2 cases from the same family, presenting with features of either end of the spectrum. One was a 11-year-old male with history of recurrent episodes of both eyes (BE) itching and watering. His best-corrected visual acuity (BCVA) in BE was 20/20 with a significant cylinder of -2.5D, for which a baseline pentacam was done to rule out forme fruste keratoconus [Fig. 1]. His examination revealed BE upper tarsal papillae with limbal gelatinous nodules [Fig. 2], suggestive of active mixed VKC (predominantly limbal), with a normal fundus and intraocular pressure (IOP).

The second case was his grandfather, a 65-year-old male who gave a history of episodes of itching and redness in childhood being treated with topical medications, and whiteness in superior half of cornea since 30 years. He was a known case of glaucoma, under medication, with a currently normal IOP, bilateral cupping of 0.8 and a BCVA RE 20/60, LE 20/30. Examination of BE seemed suggestive of partial LSCD [Fig. 3] and old burnt-out VKC with secondary steroid-induced glaucoma.

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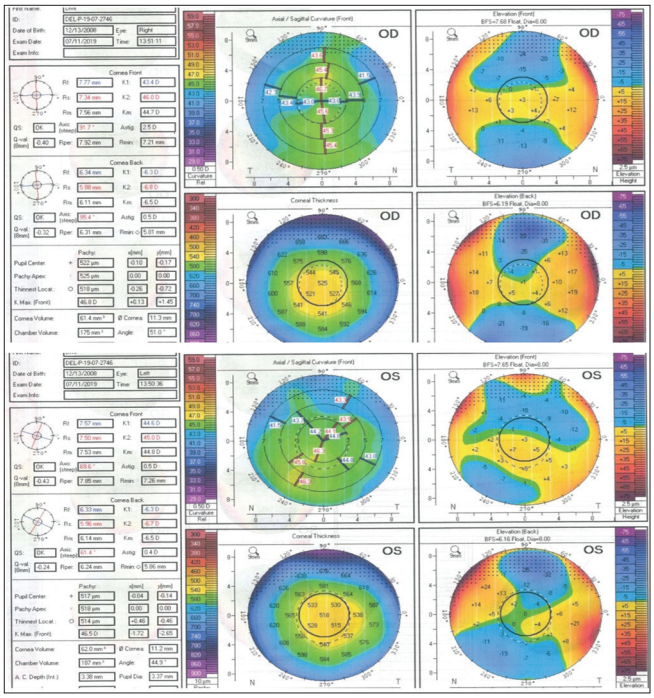

Figure 1: Pentacam of both eyes of the child showing a normal scan

Discussion

VKC is multifactorial disease for which a hereditary association has been suggested. No direct genetic associations nor a clear correlation with specific genetic loci has been found, but the role of cytokine gene cluster on chromosome 5q has been questioned.[3] Although VKC is seen more often in patients who have atopic family histories,[4] it is indeed rare to find 2 cases of different generations in the same family, with features of both active limbal VKC and old chronic limbal inflammation leading to LSCD.

Declaration of patient consent

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

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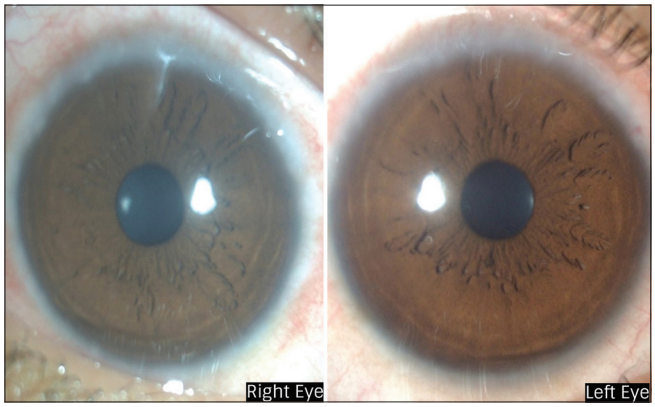

Figure 2: Bilateral limbal gelatinous nodules (active limbitis) seen in the child suggestive of active VKC

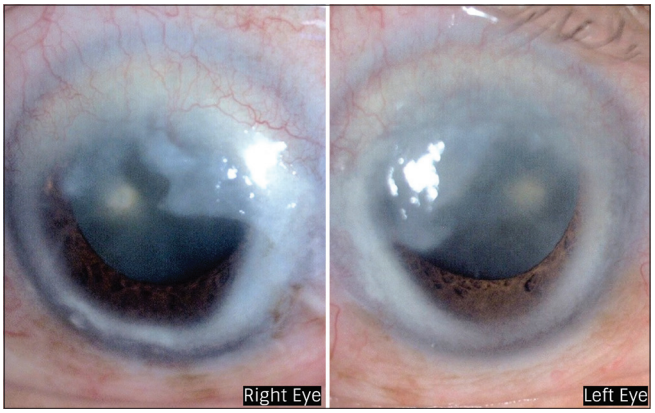

Figure 3: Bilateral features of partial LSCD seen in the grandfather with superior conjunctivalization, dense scarring, and superficial vascularization

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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