Advanced keratoconus in a child with juvenile scleroderma

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An 18-year-old male presented with complaints of gradually decreasing vision in both eyes for 6 years. He was a diagnosed with juvenile scleroderma at the age of 11 years by a pediatric rheumatologist. Clinical slit lamp examination showed features of ectasia, thinning, Vogt's striae, and apical scarring in both eyes. Bulbar and tarsal conjunctiva was quiet and normal. Corneal tomography revealed mean keratometry 65.8 and 65.4 diopters, thinnest pachymetry of 351 and 224 microns in the right and left eye, respectively. There was no history of itching and eye rubbing in the past. Patients of juvenile scleroderma may have associated keratoconus. The management of advanced keratoconus presents challenges related to handling and insertion of contact lenses in this condition. Keratoplasty is an option in those patients when contact lenses are not acceptable.

Key words: Connective tissue disorder, juvenile scleroderma, keratoconus

Keratoconus is a bilateral and usually asymmetrical disease in which the ectatic cornea becomes conical in shape.[1] It usually presents in first to second decade of life and tends to progress till the third decade. The exact cause is not fully understood but eye rubbing has a strong association either in isolation or when associated with allergic eye disease.[2] Few cases are also believed to be hereditary in nature. It affects both genders and all ethnicities.

Scleroderma is an autoimmune multisystem rheumatic disorder. The disease is characterized by stiffening of the skin that is caused by extra deposition of collagen in the skin and subcutaneous tissue followed by scarring. Although, the condition is usually seen in women in the age group 35–55 years, the disease can occur in children.[3] The condition occurs in two main forms–localized or generalized.

The various ocular manifestations in scleroderma are - eyelid changes (tightness of skin of eyelids, shortening of interpalpebral fissure, telangiectasia of eyelid skin), disturbed lacrimal glandular function, conjunctival changes (vascular congestion, telangiectasias), episcleritis, lenticular opacities, anterior uveitis, vitreous changes, and retinal abnormalities.[4,5]

Corneal involvement is rarely reported. There are isolated reports of corneal ectasia in two adults with systemic scleroderma, one with keratoconus in a 50-year-old and second with pellucid marginal degeneration in a 55-year-old.[6,7] Herein, we report a case of bilateral advanced keratoconus in an 18-year-old male with juvenile scleroderma.

Case Report

An 18-year-old male presented to our refractive clinic services with complaints of progressive decrease in vision in both eyes for 6 years. He was using glasses for the past 8 years and his current glasses were a year old. His birth history was normal with age appropriate mile stone development. There was a history of first-degree parental consanguinity. At the age of 11 years, he was diagnosed with juvenile systemic scleroderma by a pediatric rheumatologist and since then he was on varied oral immunosuppressants (methotrexate, mycophenolate mofetil, azathioprine) and corticosteroids intermittently. General examination revealed puckered skin around the mouth, and his teeth were widely spaced. The skin of the dorsum of the hands showed tightness, with little bit of flexion contracture of fingers and the tip of the digits showed healing ulcers [Fig. 1a-c]. On ocular examination, his uncorrected visual acuity was 20/320 in both eyes which improved to 20/160 and 20/200 in the right and the left eye, respectively. Palpebral skin felt firm on palpation, with adequate eyelid closure although the palpebral aperture was slightly reduced. Slit lamp examination showed a healthy ocular surface with normal upper tarsal conjunctiva on eye lid eversion, corneal thinning, ectasia, Vogt's striae, and apical scarring in both the eyes [Fig. 2a and b]. The bulbar and tarsal conjunctiva was normal and limbus had normal appearance. The anterior and posterior segment was unremarkable. Corneal tomography using Oculyzer (Alcon Inc., Fort Worth, Texas) showed the right eye having a steep cornea with a central cone with a mean keratometry of 65.8 D and a K max of 77.2 D. In the left eye, the mean keratometry was 65.4 D and the K max was 94.4 D [Fig. 2c and d]. The thinnest pachymetry was 351 and 224 microns and located inferotemporal and inferonasal to the pupil center in the right and left eye, respectively. Both the eyes showed a deranged corneal thickness spatial profile (CTSP) and percentage thickness increase (PTI) as evident on Belin-Ambrosio enhanced ectasia display on Oculyzer [Fig. 2e and f]. Based on the ocular and systemic features, an ocular diagnosis of both eyes keratoconus and juvenile scleroderma was made. The patient did not have any history of redness, itching, and eye rubbing in the past. A contact lens trial was performed to assess the visual potential. A scleral prosthetic replacement

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Figure 1: (a) Dorsum of hands showing skin tightness; (b) distal end of digits showing ulceration; and (c) perioral skin stretching causing dental abnormalities

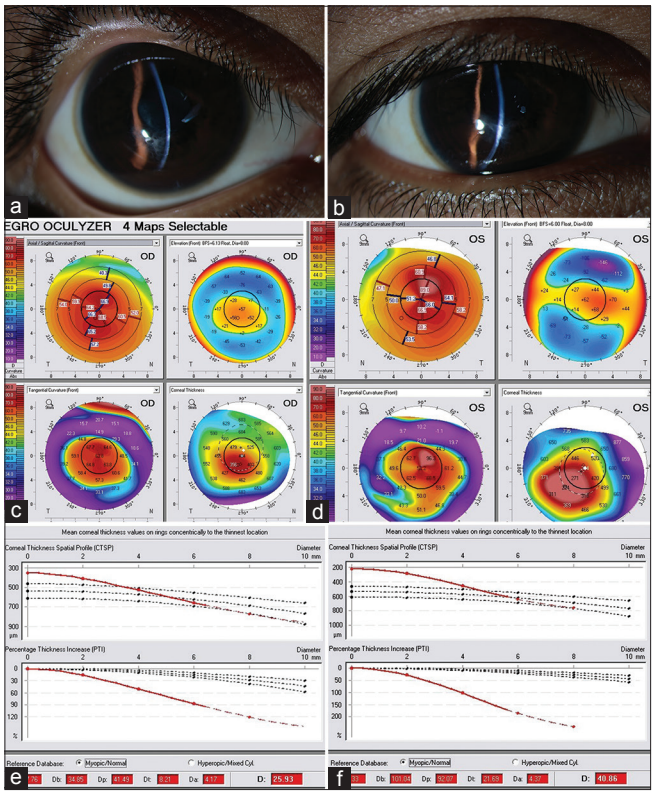

Figure 2: (a-f): Slit lamp photographs of the right (a) and the left eye (b) showing thinning, ectasia and apical scarring. Oculyzer maps of the right (c) and the left eye (d) showing advanced ectasia and corneal thinning; and deranged CTSP and PTI in right eye (e) and left eye (f)

of the ocular surface ecosystem contact lens trial helped in improvement of vision to 20/50 and 20/30p in the right and left eye, respectively. In view of recurrent ulceration at the distal end of digits, the patient was not keen on using contact lenses. An option of keratoplasty was also discussed with the patient and his parents. The pros and cons of keratoplasty, postoperative care, and need of medication instillation were discussed. The patient decided that he will undergo keratoplasty at a later point in time.

Discussion

The prevalence of keratoconus ranges from 0.0003% to 2.3% in various studies.[8] This is the first case report of keratoconus in association with juvenile scleroderma. Our patient did not have any history of eye rubbing or allergic eye disease in the past. His eyelid showed tightness, although lid closure was complete and adequate. The human cornea and skin are predominantly composed of collagen type 1.[9] Considering the collagen abnormality in both these disorders and progressive gradual tightness of the eyelid skin, it is likely that the primary systemic condition may have been an attributable risk factor for keratoconus progression.

The case reported by Anayol *et al*.[6] described a middle-aged female patient of scleroderma with keratoconus who was managed conservatively with lubricants and spectacles. There were few dilemmas regarding the management in our patient who is 18 year adolescent. Ulceration of distal end of digits made him less receptive to using and handling contact lenses. The specific issues related to keratoplasty that needed consideration in this patient were feasibility of the periodic steroid medication instillation in the postoperative period and how the systemic condition affects the success and graft survival after keratoplasty. As the patient had advanced keratoconus and needed immunosuppressants for the systemic condition, keratoplasty was performed.

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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