

Sensory Exotropia Associated with Keratoconus and Review of Literature: Strabismus and Keratoconus

Suleyman Ciftci^{1,*}, Leyla Ciftci², Eyup Dogan³, Ali Simsek⁴

¹ Department of Ophthalmology, Diyarbakır Training and Research Hospital, Diyarbakır, Turkey

² Department of Cardiology, Faculty of Medicine, Dicle University, Diyarbakır, Turkey

³ Department of Ophthalmology, Siverek State Hospital, Sanliurfa, Turkey

⁴ Department of Ophthalmology, Faculty of Medicine, Adlyaman University, Adlyaman, Turkey

Abstract

Purpose: To present a case of sensory strabismus due to keratoconus with an ipsilateral nodular lesion of the bulbar conjunctiva.

Case presentation: The patient is a 17 year-old-boy. The fixing eye was the right eye and keratoconus in this eye was late onset. Vision in the left eye was poor and keratoconus was advanced in this eye. Due to longstanding of keratoconus and occurrence in the sensitive period, sensory exotropia had developed in the left eye. A nodular lesion of the bulbar conjunctiva was also present in the ipsilateral eye.

Conclusion: Sensory exotropia is not unusual but to be due to longstanding keratoconus is not a usual strabismus. If keratoconus develops before 7 years of age, sensory strabismus can develop. We reported and reviewed literature that keratoconus seldom begins in the sensitive period where abnormal visual experience affects visual development and may induce sensory exotropia. (*Eye Science 2013; 28:88–91*)

Keywords: keratoconus; sensory exotropia

Introduction

Keratoconus is the most common primary ectasia. It usually occurs in the second decade of life and affects both genders and all ethnicities. The estimated prevalence in the general population is 54 per 100,000. The diagnosis of keratoconus can be made by the presence of keratometry over 47.20 D¹. Sensory strabismus occurs due to temporary or permanent loss of vision in one or both eyes. The prevalence of sensory strabismus is 5–9%. When it occurs in adults,

the tendency is for it to be exotrophy².

Case presentation

We present a case of bilateral keratoconus, exotrophy, and nodular lesion of the bulbar conjunctiva in the left eye. The patient is a 17-year-old boy. The fixing eye is the right eye and keratoconus in this eye was late onset. Vision in the left eye is poor and keratoconus is advanced in this eye. The patient has a corrected acuity of 6/9 in his better and fixing eye and an acuity of 1/60 in the affected eye. Keratometric measures were 42.75 D at an axis of 28° and 49.0 D at an axis of 118° for the right eye, and 52.0 D at an axis of 154° and 58.75 D at an axis of 64° for the left eye. SimK1 was 45.24 D at an axis of 113 and SimK2 was 42.75 D at an axis of 25 for the right eye. SimK of the left eye could not be obtained because of the high, steep keratometry value. The refraction of the right eye was +1.5, –7.5 × 25 and the refraction of left eye could not be obtained (Figure 1).

The patient or the parents do not know when keratoconus started in his left eye and the parents indicated a childhood onset strabismus history. Keratoconus probably advanced in the left eye in childhood and sensory exotropia had developed due to prolonged reduced visual acuity in his poorer eye. A prism cover test revealed a 45 prism diopter exodeviation (Figure 2).

A nodular lesion of the bulbar conjunctiva is also present in the ipsilateral eye. This lesion has invaded the cornea and is surrounded by a vascular injection (Figure 3). It appears to be a pseudoepitheliomatous hyperplasia, which is an inflammatory lesion that

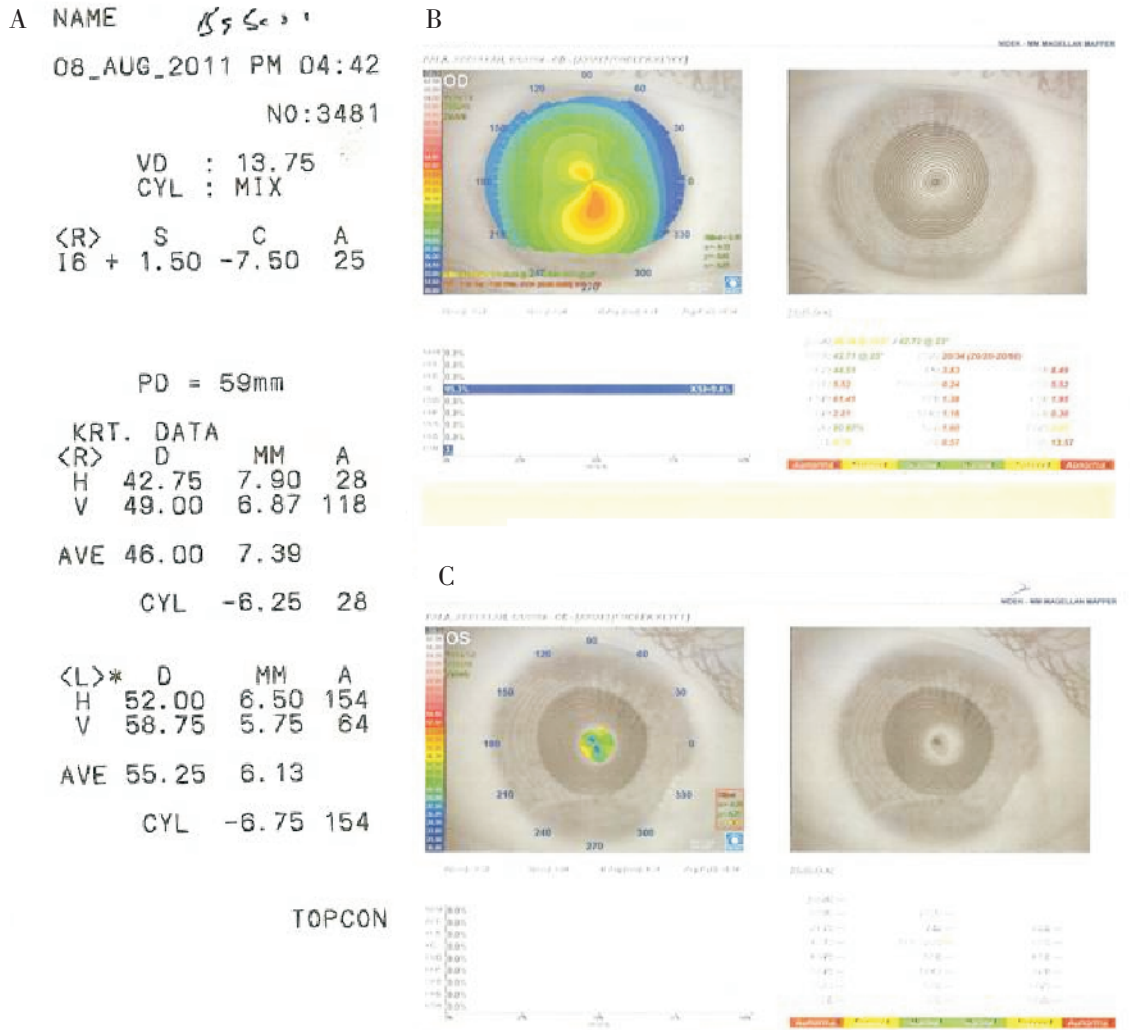


Figure 1 A: the refraction and keratometer value of the patient; B: corneal topography of the right eye; C: corneal topography of the left eye.

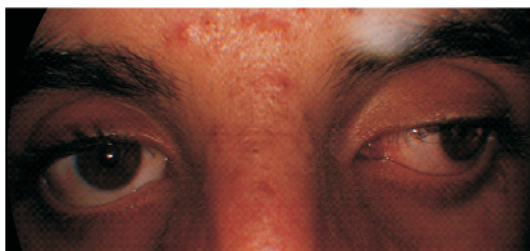


Figure 2 Clinical photography of the patient shows exotropia in the left eye.

usually occurs on the eyelid but rarely occurs in the bulbar conjunctiva. No active inflammation of the conjunctiva was present but the patient has a history of vernal keratoconjunctivitis.

Discussion

Sensory strabismus is commonly encountered, but

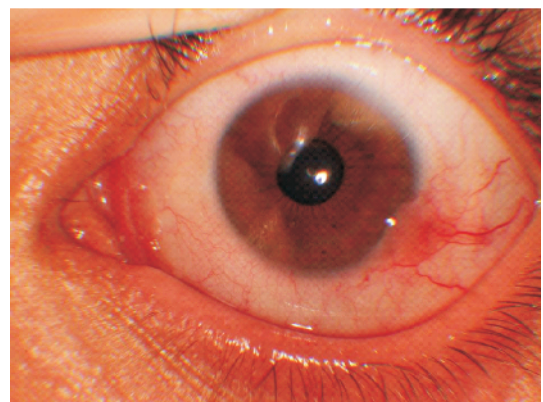


Figure 3 Clinical photography of the nodular lesion of the bulbar conjunctiva.

the cause of poor sight is varied. Chorioretinal atrophy, congenital cataract, optic atrophy, disease of

retina, complicated cataract, leucoma, coloboma, high myopia, congenital glaucoma, penetrating trauma, contusional eye trauma, and traumatic cataract are the common causes³. The prevalence of keratoconus in the general population appears to be relatively high, but the occurrence of sensory strabismus due to keratoconus is not a common entity. Because keratoconus generally develops bilaterally, the visual loss in uncorrected keratoconus is gradual and myopic. Loss of visual acuity usually is prevented by glasses or contact lenses and typically first occurs around the age of puberty. Therefore, the sensitive period for the development of suppression finishes when keratoconus settles in.

Consulting PubMed, we could find only two reports about sensory strabismus developing after keratoconus. In these studies, the binocular function of the patients with longstanding asymmetric keratoconus was investigated. The first study by Sherafat et al. investigated a database of approximately 350 patients with keratoconus. Only twenty patients were found with abnormal binocular function⁴. The abnormalities described in this study were central suppression of the deviating eye, reduction or loss of stereopsis with a microtropia when wearing their scleral lens, and a manifest exotropia with suppression when the poorer eye was uncorrected. The earliest age at which keratoconus was diagnosed in their series of patients was 12 years. However, we think that these authors may have made an error when they accepted the onset age of keratoconus as the age at which the diagnosis made and they claim that the adult visual system is susceptible to prolonged visual deprivation. However, the general assumption is that the sensitive period ends at about 7 years of age and the adult visual system is not affected by visual deprivation in humans⁵⁻⁹.

Khan and Al-Shamsi investigated a database of 103 patients with keratoconus and reported only seven patients with abnormal binocular function due to longstanding keratoconus¹⁰. In their series, six patients had constant exotropia. After intervention to improve visual acuity, the six patients complained of constant binocular diplopia, which was resolved after successful surgical alignment. Khan and Al-Shamsi explained that resolution of diplopia by realigning

the eyes allowed the patients to use their suppression scotoma and be diplopia-free. The major limitation of this study was a failure to report the onset age of keratoconus in their series and they could not explain the etiology of development of suppression in keratoconus.

Conclusion

Sensory exotropia is not unusual but to be caused by longstanding keratoconus is unusual. Because keratoconus generally develops bilaterally, the visual loss in uncorrected keratoconus is gradual and myopic, and visual acuity loss can usually be prevented by glasses or contact lenses. This condition typically first occurs around the age of puberty, but if keratoconus develops before 7 years of age, sensory strabismus can develop. The presence of a nodular lesion of the bulbar conjunctiva with keratoconus and history of vernal keratoconjunctivitis suggest that keratoconus began in our patient before 7 years of age, and in our opinion, in the series that we mentioned above, keratoconus also likely began before 7 years of age.

We reported a patient who showed approximately 45 prism diopters of constant sensory exotropia. The etiology of strabismus due to keratoconus has not been well defined in literature. We reported and reviewed literature that shows that keratoconus seldom begins in the sensitive period, where abnormal visual experience affects visual development and may induce sensory exotropia.

References

- 1 Sugar J. Stromal corneal dystrophies and ectasias. In: Yanoff M, Duker JS, eds. *Ophthalmology*. Second edition. St. Louis, MO: Mosby 2003; 443.
- 2 Merino P, Mateos C, Gómez De Liaño P, et al. Horizontal sensory strabismus: characteristics and treatment results. *Arch Soc Esp Ophthalmol*, 2011, 86(11): 358-362.
- 3 Oliveira BF, Bigolin S, Souza MB, et al. Sensorial strabismus: a study of 191 cases. *Arq Bras Oftalmol*. 2006; 69(1): 71-74.
- 4 Sherafat H, White JE, Pullum KW, et al. Anomalies of binocular function in patients with longstanding asymmetric keratoconus. *Br J Ophthalmol*, 2001, 85(9): 1057-1060.
- 5 Boothe RG, Dobson V, Teller DY. Postnatal development of vision in human and non-human primates. *Ann Rev*

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- Neurosci, 1985, 8:485–545.
- 6 Von Noorden GK. Amblyopia: a multidisciplinary approach. *Invest Ophthalmol Vis Sci*, 1985, 26:1704–1716.
- 7 Wiesel TN. Postnatal development of the visual cortex and the influence of environment. *Nature*, 1982, 299:583–591.
- 8 Sloper JJ. Competition and co-operation in visual development. *Eye*, 1993, 7:319–331.
- 9 Vaegan, Taylor D. Critical period for deprivation amblyopia in children. *Trans Ophthal Soc UK*, 1979, 99:432–439.
- 10 Khan AO, Al-Shamsi H. Binocular diplopia following vision restoration in patients with keratoconus and sensory strabismus. *Strabismus*, 2008, 16(1):19–22.