Case Report

A Patient with Keratoconus, Axial Hyperopia and Pigmentary Retinopathy

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Abstract

Introduction: Keratoconus has been known to be associated with various ocular conditions. **Objective:** To describe a case of high hyperopia, keratoconus and pigmentary retinopathy. **Case:** An eight year old boy, whose initial refractive error was high hyperopia with short axial length of 20mm, presented as bilateral advanced keratoconus with acute hydrops in one eye. Subsequently his other eye also developed hydrops. He had nystagmus and pigmentary retinopathy from his infancy. He was managed conservatively and the hydrops resolved in both the eyes leaving paracentral scar in the cornea. **Conclusion:** The triad of keratoconus, high hyperopia and pigmentary retinopathy has been quite rare. There could be syndromic association between the three simultaneous ocular findings.

Introduction

Keratoconus is a corneal disorder in which the central portion of the cornea becomes thinner and bulges forward in cone-shaped fashion resulting in myopia, irregular astigmatism and visual impairment. The onset of the disease occurs in puberty, affects both genders and is prevalent in all races (Gordon-Shaag A et al 2015, Feder RS & Kshettry P 2005). The disease is bilateral, although asymmetrical. As the disease progresses, the descemet's membrane can develop breaks leading to stromal edema and pain. Corneal topography helps in diagnosis and early detection of keratoconus. In the initial stage of the disease, vision can be improved by spectacles but as keratoconus progresses rigid gas permeable contact lenses will be needed. However about 20% of patients will eventually need keratoplasty. Recently, a technique called collagen cross-linking had been developed which arrests the progression of keratoconus (Gordon-ShaagA et al, 2015). Both environmental and genetic factors contribute to the pathogenesis of keratoconus. Keratoconus is known to be associated with various systemic and ocular conditions, the most common ones being atopy, Down's syndrome and vernal keratoconjunctivitis. Other systemic findings known to exist are connective tissue disorders like osteogenesis imperfecta, Ehlers-Danlos syndrome and mitral valve prolapse. Ocular associations that have been reported are Lebers congenital amaurosis, pigmentary retinopathy, progressive cone dystrophy, aniridia and corneal dystrophies (Feder RS & Kshettry P, 2005). We present a case of keratoconus, axial hyperopia and pigmentary retinopathy occurring simultaneously.

Case

A seven month old male child thought to have poor vision was brought by his parents

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for ocular examination. His perinatal history was normal. He has no siblings. There was no family history of any ocular disorder. There was no history of consanguineous marriage in the family. On examination the patient had nystagmus but could follow light. Cyclorefraction revealed a hyperopia of +7.00 DS in both eyes. Patient was prescribed glasses and was advised to follow up in 6 months. But there had been no improvement in his vision even with regular wearing of glasses. At the age of 3 years, visual acuity was counting finger at half a metre distance. Cyclorefraction revealed net hyperopia of +9 DS/-1 DC at 180^0 in the right eye and +10 DS/ - 1 DC at 180^0 in the left eye. His fundus examination showed salt and pepper appearance of pigmentary retinopathy and bony spicules in the mid and peripheral retina. Fundus photography could not be taken due to nystagmus. Parents were explained guarded prognosis for vision due to posterior segment pathology. They were advised to continue the glasses and were advised to follow up every year, during which the patient seemed to have stable visual acuity, manifest refractions and anterior segment examination. However the parents noticed that their child's ability to see was worse in the night.

At 8 years of age, A- scan axial length was 20.11mm in right eye and 20.20 mm in the left eye. Rest of the examination findings remained the same. After this visit the patient did not turn up for follow up for two years. At the age of ten he was brought to the emergency department with history of whitish lesion in the left eye noticed suddenly. (Figure 1) It was associated with foreign body sensation and watering. There was no history of redness, itching, rubbing of eyes or trauma. There was bulging of central cornea due to edema and epithelial bullae. Peripheral cornea was clear (Figure 2). In the right eye, cornea was clear but there was thinning at the central cornea assuming cone shape and Vogt's striae could also be seen. Munson sign was positive. Patient was diagnosed as having axial hyperopia,

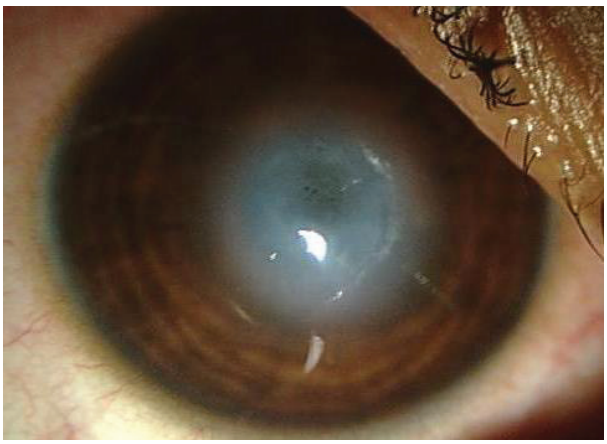

Figure 1: Acute hydrops

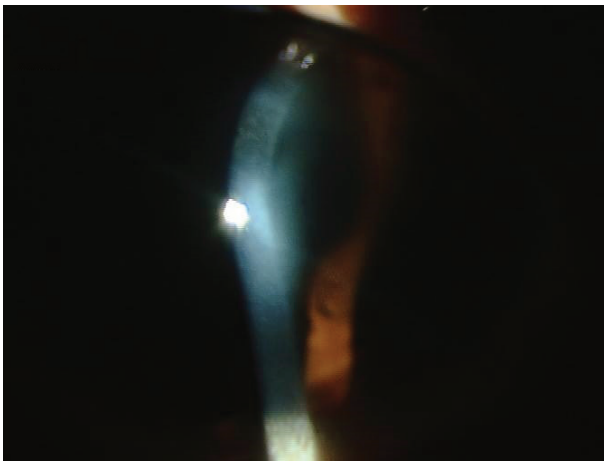

Figure 2: Acute hydrops showing central corneal bulging.

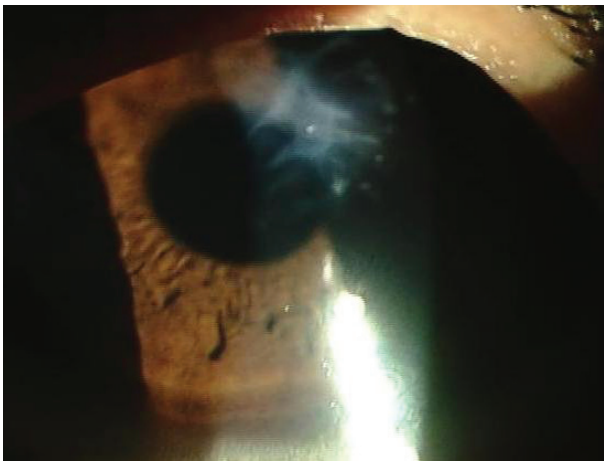

Figure 3: Corneal scar after resolution of hydrops.

pigmentary retinopathy and advanced keratoconus in both eyes with acute hydrops in the left eye. Corneal topography could not be done due to nystagmus. The patient was given hypertonic normal saline. NaCl 5% eye drops four times a day, eye drops. Timolol 0.5 % twice a day and Ciprofloxacin ointment at bed time in the left eye. After three weeks the hydrops of the left cornea resolved with small paracental scar (Figure 3). The patient was referred to low vision services where he was advised stand magnification for near vision. Parents were counseled about the nature of the condition and the reason for keratoplasty surgery, not being beneficial for the condition. They were hence advised to protect their child from any trauma. Within 3 months the fellow eye also developed acute hydrops and was managed in the same way. This eye also developed paracentral scar after resolution of the hydrops.

Discussions

Most of the keratoconus patients present with myopic refractive error and myopic astigmatism. Ernst BJ & Hsu HY (2011) as well as Touzeau O et al (2004) had studied relation of keratoconus and axial length. Both studies quoted similar conclusion that the axial length, anterior chamber depth, and posterior segment length were all significantly longer in the keratoconic group than in the emmetropic control group. Ernst BJ & Hsu HY (2011) reported that the ratio of mean keratoconic axial length versus mean emmetropic axial length was 24.40 vs. 23.24 mm (p=0.001). The same ratio in the study by Touzeau O et al (2004) was calculated as 23.97 mm versus 23.21 (p=0.001). Touzeau O et al (2004) also mentioned posterior segment length was significantly greater in the keratoconus group than in the emmetropic group (16.54 mm versus 15.99 mm, p<0.001). Hence both studies quoted that keratoconic eyes have on average longer axial lengths that are primarily because of longer posterior segment lengths than emmetropic eyes. But in our case the axial length was around 20mm which indicated the presence of high hyperopia which was a unique finding in this case. Martin R (2009) had reported one case of high hyperopia with keratoconus but there was no retinopathy like in our case. The patient in our case had a high hyperopia of +9 DS and the refractive error did not show astigmatism during the course of follow up. The cornea also looked grossly normal. Hence keratoconus was not thought of. Retrospectively there could have been subclinical keratoconus which might have progressed rapidly when patient missed follow up for two years. It was diagnosed only after having acute hydrops.

Systemic and ocular association of keratoconus has been studied frequently. Increased incidence of atopy, Down's syndrome and vernal keratoconjunctivitis has been well known.

(Feder RS & Kshettry P, 2005). In our study, there has not been any systemic association with keratoconus but had ocular association of axial hyperopia and pigmentary retinopathy. Peduzzi M et al (1991) mentioned similar case of keratoconus and pigmentary retinopathy in a child of five years but without hyperopia. So far to our knowledge there has been only one reported case having axial hyperopia, pigmentary retinopathy and keratoconus in the literature which was described by Sammouh SK et al (2016). In his case, the diagnosis was made at early adulthood and there was extra feature of lipodermoid with restriction of extraocular motility along with the mentioned triad.

Although rare, keratoconus may occur in cases of high hyperopia. Association of pigmentary retinopathy gives a notion of syndromic association between the three features. Visual rehabilitation in such cases is quite disappointing due to existence of retinopathy in otherwise excellent prognosis of keratoplasty surgery in isolated keratoconus.

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