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Case Report

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Bilateral Congenital Ectropion Uveae, Anterior Segment Dysgenesis and Aniridia with Microspherophakic Congenital Cataracts and RubeosisIridis

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Abstract

In recent times, multiple eye diseases have been seen associated with an increase in the rate of Demodex infestation as a possible cause, but in the particular case of dry eye syndrome in patients treated with platelet-rich plasma, this increase in mite may be relevant to guide a more adequate treatment focusing on the elimination of the mite in conjunction with the recovery of the ocular ecology. The demodex mite is a commensal parasite that lives in hair follicles, sebaceous glands and meibomian, which in a high rate of infestation can generate alterations in the ocular area. Performing an adequate diagnosis for the detection of the mite and treatment for its eradication can be effective for the recovery of the normal physiology of the tear film that constitutes a cause of dry eye.

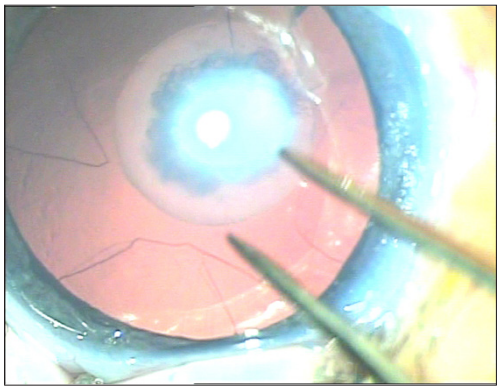
Introduction

Congenital ectropion uvea is a rare ocular manifestation of neural crest syndrome [1]. It is a non-progressive anomaly characterized by presence of iris pigment epithelium on anterior surface of iris from the pigment ruff [2]. Congenital glaucoma is its common association [3-8]. Classically it is unilateral with ipsilateral glaucoma [2]. There is one case report of bilateral congenital ectropion uvea with bilateral glaucoma [6].

Here we are documenting a case report with bilateral congenital ectropion uvea with bilateral congenital glaucoma, bilateral megalocornea,bilateral anterior segment dysgenesis, bilateral microspherophakic cataract, bilateral rubiosisiridis, bilateral non developed zonules and optically empty vitreous and bilateral nystagmus in a three months old full term baby.

Anterior segment dysgenesis spans an anomaly ranging from posterior embryotoxon with iris strands adherent to it to hypoplastic iris and dyscoria. Severe iris hypoplasia manifests as aniridia which is an autosomal dominant bilateral condition with variable appearance of iris from a rudimentary stump to mild iris atrophy. Central corneal pannus, cataract, foveal hypoplasia, decreased vision and nystagmus can be an association of aniridia.

Our case report is a conglomeration of all these independently reported ocular anomalies, never found coexistant and reported before. Throughout the literature review we did not find a single case report of such a stark bilateral anterior segment disfigurement and developmental disarray. It is a novel case report.

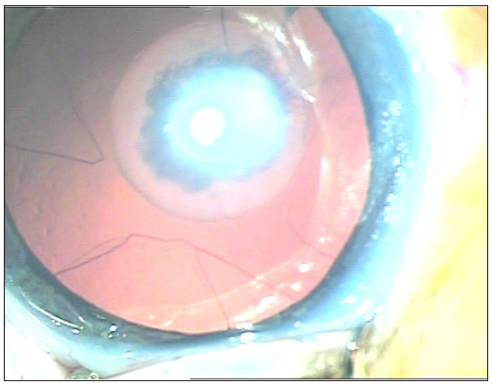


Case Report

A three month old baby girl of normal perinatal status was referred to us with a diagnosis of bilateral aniridia, congenital glaucoma and cataractousmicrospherophakia. Trabeculectomy with mitomycin-c had been done in right eye and opinion for goniotomy in left eye was asked for.

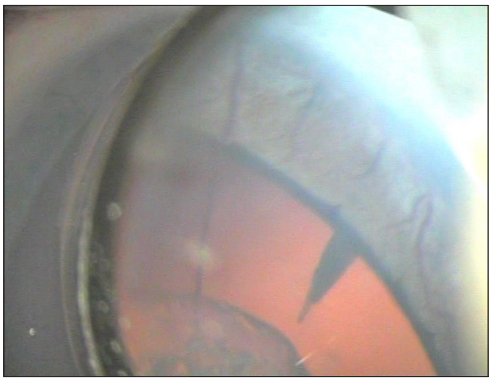
On examination under general anesthesia cornea of left eye was clear centrally while of right had central scarring due to ruptured Habb'sstriae in the zone of palpebral aperture. Corneal diameters in right eye were 14mm vertically and 15mm horizontally and in left eye were 14mm vertically and 14.5mm horizontally. Intraocular pressures were 17mm of Hg bilaterally with perkinsapplanation tonometry. Central corneal thickness was 750u in right eye and 580u in left eye.

There was very prominent posterior embryotoxon bilaterally. There was severe bilateral ectropionuveae bilaterally. There was bilateral partial aniridia with stump of iris ranging from 3-5mm in its dimensions. Even in its incompleteness it was hypoplastic in its stromal bed with hypoplasia more marked inferiorly bilaterally.



There was bilateral microspherophakia. The lens was 6mm in diameter and cataractous bilaterally. There were no zonules at all at any region in both eyes.

There was severe rubeosisiridis in all the four quadrants of both eyes. The vessels were thick and cord like running across the angle,travelling on the surface of iris, climbing down onto the non-developed zonular zone and running all across it, reaching the small crystalline len, reflecting back from margin of microspherophakia and reaching back the anterior chamber angle zone.



The angle recess was very narrow. There was no angle structure at all in any quadrant bilaterally. The posterior segment was found to be normal on indirect ophthalmoscopy bilaterally. Retinoscopy was attempted but no reflex attained centrally. The axial length was 20.19mm in right eye and 20.43mm in the left.

B scan of both eyes showed optically empty vitreous cavity with normal posterior segment topography.Posterior segment photography with RETCAM was attempted but remained poor quality due to cataract in left eye and cataract and corneal scarring in right eye. The proceeding with goniotomy was abandoned.

The family was entailed in clinics. It was a family of four siblings and the parents were first cousins around there forties. The gap between third child and this fourth baby was ten years.There was no history of maternal contraception. Pregnancy was uneventful.

There was very low frequency, low amplitude, bilateral, fine rotary nystagmus in baby with good fixation effort to light though it was not central, unsteady and unmaintained.There was intermittent bilteral lid retraction. Father's eye examination was normal. The mother and the three kids were having mild to moderate clinically insignificant anterior segment dysgenesis.Younger two kids were having visually insignificant sutural cataracts. Bilateral cataract extraction for the baby was planned and for intraocular pressure a close follow up was charted.

Discussion

Congenital ectropion uvea, axenfeldrieger anomaly, megalocornea, aniridia, rubiosisiridis, microspherophakia, congenital cataract, non-developed zonules, optically empty vitreous and disparate corneal thickness have never been mentioned together in a single eye before in literature. Our baby has all these features in tandem and that too bilaterally. The special thing about this child is that there is total disfigurement of anterior segment in both eyes and posterior segment is totally spared. Surface ectoderm, mesoderm and neural crest origins are involved with apparent sparing of neural ectoderm roots. Despite all this havoc in the anterior chamber baby still tries to take fixation.We expect some improved fixation after clearing the media but the exercise is going to be very exhaustive for sure.

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