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Case Report

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Management of Congenital Cataract Associated with Axenfeld Rieger Anomaly

Rao Muhammad Arif Khan*, Ashal Kaiser Pal and Abdul Qayyum

Pediatric Ophthalmologist, King Edward Medical University, Makkah, Saudi Arabia

***Corresponding author**

Rao Muhammad Arif Khan, MCPS, FCPS, FPO, FACS, Pediatric Ophthalmologist, King Edward Medical University, Al-Awali Street, Taif Road, Makkah, Saudi Arabia, Tel: 00966560479694; E-mail: raoarefkhan@gmail.com

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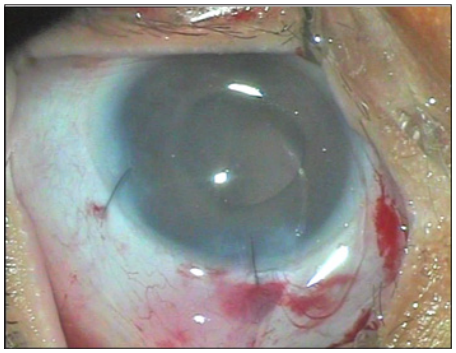
Anterior Segment Dysgenesis (ASD) represents a spectrum of developmental disease involving neural crest mesenchyme [1]. Axenfeld anomaly refers to presence of posterior embryotoxon (anteriorly displaced schwalbe's line) with attached iris strands [1]. When these peripheral abnormalities are associated with iris changes such as corectopia or atrophy it is called Rieger's anomaly [1]. In association with systemic anomalies such as abnormal teeth and facial abnormalities, particularly hypertelorism, it is referred to as Rieger syndrome [1]. Families with Axenfeld Rieger anomaly show an autosomal dominant pattern of inheritance [1]. Gene loci have been mapped to 4q25 (PITX2 gene), 6p25 (FKHL7) and 13q14 (RIEG2) [2]. Axenfeld Rieger anomaly has no gender predilection and there is associated glaucoma [2]. Cataract is not an association with it. We report a case of anterior segment dysgenesis with associated congenital cataract but no congenital glaucoma, managed by lens aspiration, primary posterior capsulotomy and deep anterior vitrectomy. To our knowledge no case of congenital cataract with associated Axenfeld Rieger anomaly is reported, so this is probably the first case report.



Case Report

A one month old baby girl presented to us with white looking right eye and small left eye. She was the youngest amongst the siblings. The elder sister was healthy and without any ocular complaints. Her parents were first cousins and ophthalmologically normal. She was full term and was delivered by spontaneous vaginal delivery. Her past medical history was unremarkable and family history noncontributory.

The mother noticed white right eye a week before. In clinics corneal opacity in right eye and congenital cataract in left eye was recorded. Both eyes appeared smaller than normal. There was low frequency low amplitude fine rotary nystagmus in both eyes. B scan ultrasonography revealed normal posterior segment topographies for both eyes. Both eyes were labeled to be micro ophthalmic.

An examination under general anesthesia showed a prominent, anteriorly displaced schwalbe's line (often referred to as posterior embryotoxon), a clear cornea and a geographically hypoplastic iris without corectopia and no iridoschisis. Gonioscopic examination revealed iridocorneal adhesions in seven clock hours separated by zones of normal iris insertions, bridging anterior chamber angle from peripheral iris to the prominent schwalbe's ridge; beyond the tissue strands the angle was open. Maximum pupillary dilation was 4mm but it remained round. There was central nuclear cataract 3mm in diameter with clear lenticular periphery as much as visible. Fundus view was very blurred and no details could be delineated. There was gross morphological asymmetry in both eyes with all above mentioned features present in left eye. In right eye cornea was totally opaque with very hazy anterior segment details. However there was no bulging of cornea.

Corneal diameters were measured and were 9.5mm for right eye and 8.5mm for left eye in both meridians. Intraocular pressure with perkins tonometer was found to be 10mm of Hg in right eye and 06mm of Hg in left eye with adjusted IOP reading of 16 and 12 mm of Hg respectively.

Cataract extraction was planned for left eye and biometry was done. Average K-reading for left eye with automated hand held keratometer was 6.42mm. Axial length with Quantel a scan was 15.03 mm. IOL power calculation with SRK-T formula gave IOL power +50.74D. With residual ametropia of +8D adjusted IOL power was calculated to be +41D.

Pediatric medical review revealed no systemic associations. Multipiece acrylic IOL was arranged to manage cataract and it was planned to keep a vigilant follow up for IOP.

Thorough debate was done to decide whether to leave the patient aphakic or facilitate the visual system with IOL implantation. Keeping in view the practically only eyed status of the toddler it

was decided to give his visual system maximum support with a very efficient postoperative follow up. So we decided to get ready for cataract extraction with primary IOL implantation and take the final decision after assessing response of ocular tissues and behavior of lens-iris diaphragm per operatively.

Pupil was dilated with 1% tropicamide eye drops, 1% phenylephrine eye drops and 0.25% cyclopentolate eye drops. All three were instilled thrice an hour prior to planned surgery, each group dosage 10 minutes apart. Maximum papillary dilatation achieved was 4 mm. Iris dilator hooks were considered to enhance dilation to maximum. To avoid over handling in an only eyed infant minimum touch approach was preferred and dilator hooks' idea was dropped.

Due to narrow palpebral aperture and small eyeball the eye was underexposed despite application of traction speculum. To enhance exposure of eyeball superior rectus bridal suture with 4/0 black silk was applied. Three step superior limbal incision with 3.2 mm keratome was given. Vision blue was injected intracamerally and removed after thirty seconds. Anterior capsule was found to be poorly stained with poor visibility. Dye reinjected for another thirty seconds. Anterior capsule was stained mildly with somewhat improved visibility. No corneal staining was noticed grossly.

Anterior continuous curvilinear capsulorhexis approximately 4mm in diameter was done with capsular forceps just encroaching the papillary plane. Anterior capsule was found to be extremely elastic and more retractile than in other congenital cataracts. Very slow, controlled and firm rotation of capsule was required to keep the rhexis regular. Vitrectorhexis and diathermic rhexis were avoided to prevent excessive instrumentation, manipulation and risk of early capsular phimosis and scarring.

Hydrodissection was done to isolate cataractous nuclear zone from cortical outskirts. Irrigation- aspiration of cataract and cortex was smooth and uneventful.

Per-operatively generalized corneal haze appeared which hampered visualization moderately but not to an extent to abandon procedure.

Anterior chamber was continuously kept maintained with viscodispersive viscoelastic material. Posterior capsule was extremely thin especially in center unlike other congenital cataracts. It was found bulging forward centrally. Primary posterior capsulotomy of 3 mm diameter was done with ocutome. No vitreous prolapsed. 5mm deep anterior vitrectomy was done.

Fundus examination with indirect ophthalmoscopy was normal. Pupil size throughout the procedure remained uniform.

Considering corneal haze development peroperatively and anterior segment crowding of microphthalmos, primary intraocular lens implantation was withheld.

The wound was closed with 10/0 nylon suture. Retinoscopy was done at 2/3m distance and reflex was neutralized at+33DS. To cater one and a half meter circle of vision of baby the same i.e. 33DS was prescribed. Subconjuntival steroid-antibiotic combination was given in lower fornix.

Postoperative topical steroids and antibiotics along with mydriatics were instituted four hours from surgery. On first postoperative day eye was quiet. B scan repeated and was normal. Parents were recounselled to get ready for long follow-ups. For right eye they were referred for pediatric corneal grafting.

Discussion

This case of Axenfeld Rieger anomaly was unusual in presentation because of its association with congenital cataract. We are not aware of any other reported case with this composite challenge. Also this case demonstrates that to manage congenital cataract in a mixed scenario needs open ended approach. We have to keep thought process on continuously and weigh situation at each step carefully. Initial induction of aphakia with optical correction followed by secondary implant at a later date is the theme. Using this approach, success was achieved without sacrificing vision or critical ocular structures.

References

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