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Case Report

Triple Trouble–Microcornea, Iris Coloboma and Hard Cataract: Outcome of Phacoemulsification in This Perilous Journey

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Abstract

Ocular colobomas are malformations of eye and are associated with early cataractous changes. Cataract surgery in eyes with coloboma require extra care and planning at each stage as these eyes are at greater risk for complications during cataract surgery. Presenting here, the outcome of cataract surgery in a 45 years female who presented with micro cornea, iris coloboma and advanced cataractous changes with nystagmus. The preoperative planning and management, the complications dealt during phacoemulsification and the outcome are discussed.

Keywords: Microcornea, coloboma, cataract, nystagmus, phacoemulsification.

INTRODUCTION

Coloboma comes from the Greek word that means "curtailed." The eye develops quickly during a fetus' first three months of growth. A gap, known as the choroidal fissure, appears at the bottom of the stalks that eventually forms the eye. This fissure generally closes by the seventh week of pregnancy. If it does not close, a coloboma or space forms. It can involve lids, iris, lens, optic disc, retina. Degree of vision loss depends on the location of the coloboma. Coloboma involving macula and optic nerve are associated with reduced vision while those involving the retina are associated with defect in visual field.

Eyes with congenital coloboma develop cataract at relatively young ages and are at greater risk for complications during cataract surgery [1, 2]. Zonular deficiency commonly seen in such cases increases the risk for vitreous loss [3, 4], complicates intraocular lens (IOL) implantation and centration of lens in the capsular bag [5]. Microcornea alongwith nystagmus present may make things further difficult. Cataract surgery not done well can result in complications like glaucoma and retinal detachment [1, 2].

CASE REPORT

A 45 year old female presented to the Ophthalmology Outpatient Department at tertiary health centre with complaints of gradual painless loss of vision since 6 months in left eye with no history of trauma, watering, redness of eye. There was no history of wearing glasses. Patient was not a known case of diabetes mellitus, hypertension and bronchial asthma.

On Slit lamp examination of Right eye, there was microcornea (vertical diameter: 7 mm and horizontal diameter 7.5mm), central nebulomacular corneal opacity, total typical iris coloboma in lower nasal quadrant, irregular depth of anterior chamber, with sluggish reacting pupil and advanced cataract and faint fundal glow with nystagmus.

On Slit lamp examination of Left eye, there was microcornea (vertical diameter: 8mm and horizontal diameter 8.5mm), iris coloboma in lower nasal quadrant with briskly reacting pupil and immature senile cataract (grade: NS3) with faint fundal glow, with nystagmus (Figure 1a & 1b). The cornea was clear, and the endothelium appeared to be healthy. The anterior chamber, however, was on shallow side.

Fig-1a: LE showing microcornea, irregular AC depth and dense cataract

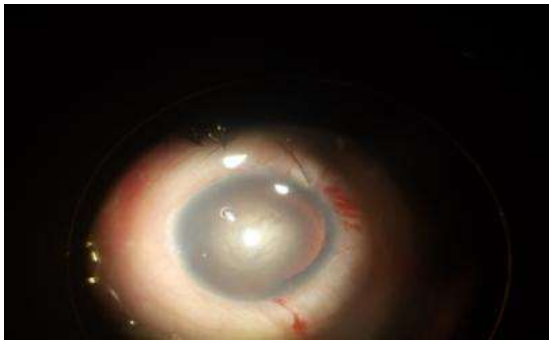

Fig-1b: Pupil dilated showing nuclear cataract with inferior iris coloboma

Best corrected visual acuity showed perception of light positive in right eye and Finger count at 2 m in left eye. Dilated fundus examination of both eyes with indirect ophthalmoscopy was attempted. Through hazy media one could appreciate retinal choroidal coloboma in lower half in left eye with no involvement of optic disc. Ultrasonography of both eyes showed choroidal coloboma and also that retina was attached (Figure 2a, 2b). The intraocular pressure in right eye was 12 mmHg and it was 14 mm Hg in left eye. Patient was planned for cataract surgery in left eye under guarded visual prognosis. As corneal diameter in right eye was very less with corneal opacity since birth, along with diminuition of vision since childhood, it was decided to be left alone as of now.

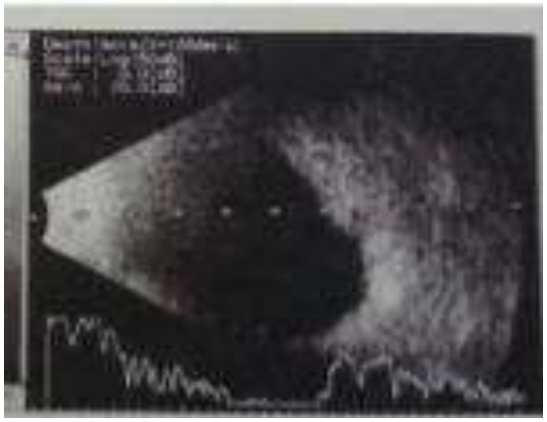

Fig-2a: B scan of both eyes, Right and Left showing choroidal coloboma

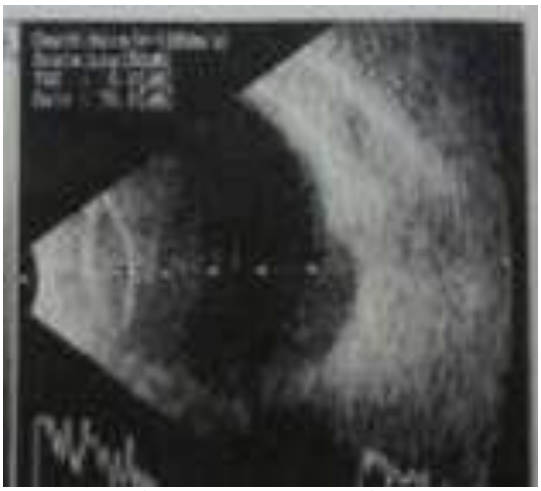

Fig-2b: B Scan of both eyes, Right and Left showing choroidal coloboma

When deciding our surgical approach, we considered several factors. One was the small corneal diameter in combination with the shallow chamber and full-sized hard cataract. Added to our concern was the inferior coloboma with a given degree of absent zonules, although there was no sign of phacodonesis.

Finally, we needed to determine the best way to anchor an IOL should capsular damage occur during surgery. We could not rely on an ACIOL as a backup plan, given the patient's anterior segment anatomy. Iris and scleral suturing would prolong surgery significantly, again risking an intraoperative suprachoroidal hemorrhage.

Biometry relieved our concern regarding the IOL by showing a very steep cornea. The average keratometry reading was approximately 48.50 D. A-scan ultrasonography showed an elongated axial length (24.21 mm), and B-scan ultrasonography ruled out a staphyloma. As a result, the patient required only a 5.00 D PCIOL for emmetropia. Given that finding, we theorized that in case of an eventuality of not being able to place the IOL, we could leave the eye aphakic, and she could easily manage the necessary spectacles.

Phacoemulsification was planned and performed on this patient. After completing capsulorrhexis phaco was started (Figure-3). The parameters kept for phaco 1 were : bottle height 100 cm, power 60%, vacuum of 50 mm Hg and aspiration flow rate 35/cc (Figure-4). However , to our great dismay while doing trenching in phaco 1, the weak zonules gave way and nucleus drop occured. Surgery was stopped, without much manipulation a suture was placed to close the section and it was planned to refer the case to vitreoretinal surgeon for further management.

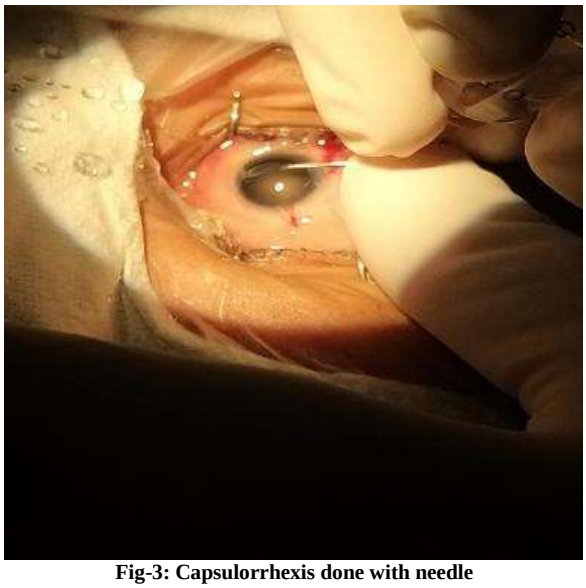

Fig-3: Capsulorrhexis done with needle

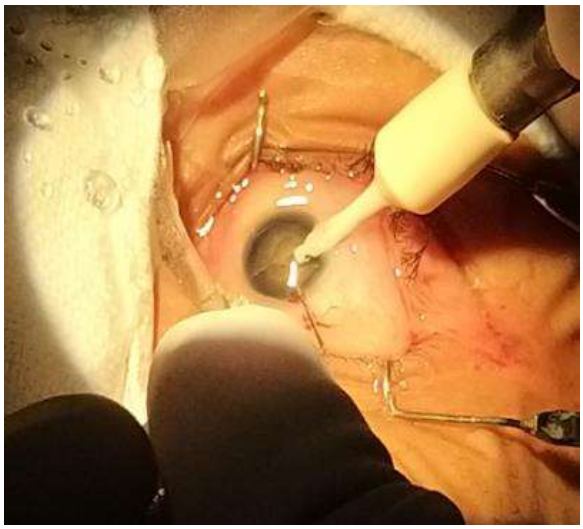

Fig-4: Trenching done in Phaco 1

Postoperatively patient was put on systemic steroids and topical antibiotics, steroid and antiglaucoma drops. Repeat fundus examination was done by vitreoretinal surgeon which showed nucleus lying over retina (Figure-5). Vitreoretinal surgery was planned after a week (Figure-6). Nucleus removed, pars plana vitrectomy was done. IOL was deferred and the eye was left aphakic .But however, because the calculated IOL power was low due to long axial length, in spite of aphakia she maintained her visual acuity.

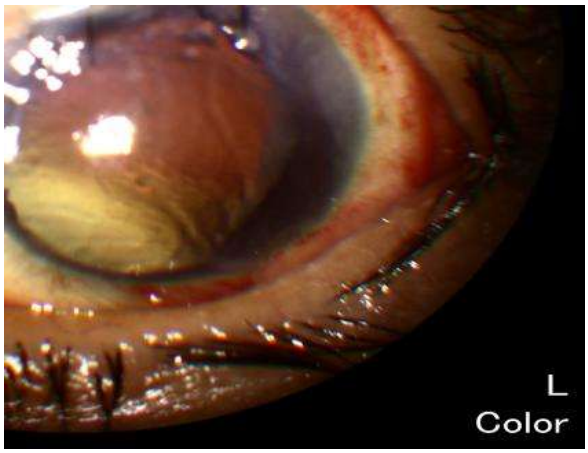

Fig-5: Fundus picture showing nucleus lying over retina

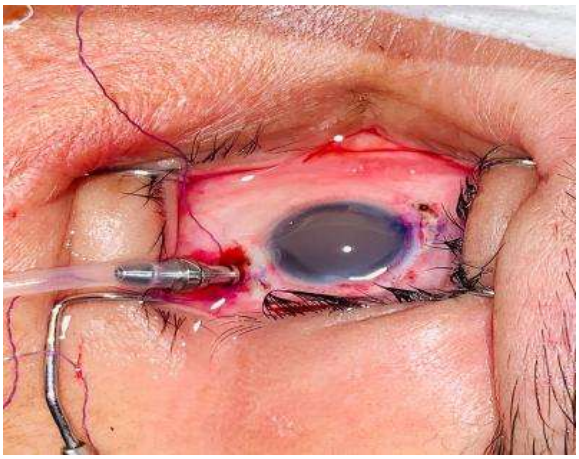

Fig-6: Nucleus extraction with vitrectomy

Postoperatively, the patient was continued on topical drops for six weeks and regular follow up was maintained (Figure-7). The patient fared remarkably well, and we more than achieved the targeted visual acuity. Three months after surgery, the patient's UCVA was 6/60 OS, with a BCVA of 2.00 -2.25 X70 = 6/24. She was able to read large newsprint with a 3.50 D add, and her IOP was 16 mm Hg. Postoperative fundus findings included an extensive inferior choroidal coloboma (Figure-8). Most importantly, however, the patient now functions as a sighted individual.

Fig-7: Immediate postoperative period

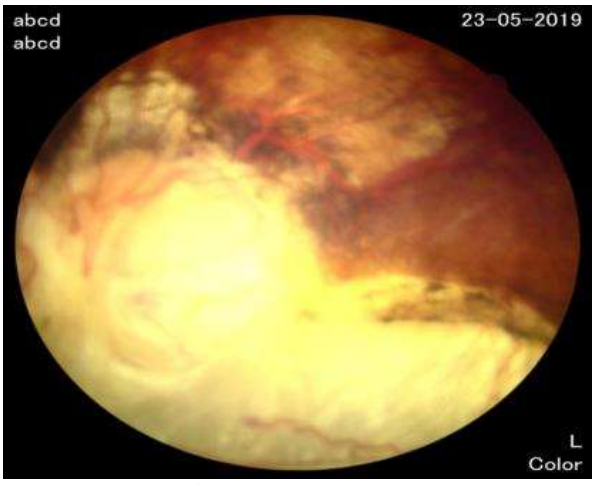

Fig-8: Postoperative fundus picture showing choroidal coloboma

DISCUSSION

Microcornea is the condition in which the horizontal diameter of cornea is less than 10 mm [6]. These patients usually have shallow anterior chamber. Microcornea can be associated with glaucoma, congenital cataract, optic disc hypoplasia. Coloboma refers to the congenital defect in the formation of eyeball. Depending on the structure involved vision impairment is seen. Cataract in eyes with microcornea and typical coloboma pose a great challenge for surgery due to small size of cornea and shallow anterior chamber which pose a challenge for corneal incisions and are at greater risk for complications during cataract surgery. Phacoemulsification though not a contraindication, but can be challenging. In these cases, zonular dialysis, phacodonesis, poor pupillary dilatation, capsular laxity can be observed and difficulty can be faced with in capsulorhexis due to these findings. Zonular deficiency can also lead to vitreous loss which pose difficulty in intraocular lens implantation and placement of lens in capsular bag.

Rafi *et al*., compared the outcome of phacoemulsification in eyes of 10 patients with coloboma in their study on "Management and surgical outcomes in phacoemulsification and intraocular lens placement in eyes with cataract and congenital iris coloboma [7]" and also found that cataract is usually formed at an early age in such patients. They concluded that cataract surgery done has good outcome with extra care and good planning.

Nanda L *et al*., presented a case report on "Phacoemulsification done in microphthalmic eye with micro cornea and coloboma [8]" the surgery was uneventful with good visual outcome.

Our case also presented with microcornea and typical coloboma which had an intraoperative complication of nucleus drop due to zonular dehisence while performing phacoemulsification. The complication was detected timely and was managed accurately.

CONCLUSION

Microcornea with iris coloboma with choroidal coloboma is not a contraindication for cataract surgery. Phacoemulsification in eyes with coloboma is more problematic compared to normal eyes. Our case highlights that in spite of microcornea, iris coloboma, hard cataract, choroidal coloboma, with shallow anterior chamber along with nystagmus, the phacoemulsification cataract surgery was attempted and despite of the complication encountered, it was managed efficiently with satisfactory end result.

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