Pars Plana Vitrectomy for Retinal Detachments Associated with Chorioretinal Coloboma

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Abstract

***Purpose*:** To evaluate the anatomical and functional outcomes of surgical management of retinal detachments associated with choroidal colobomas

***Methods*:** In this retrospective study, 31 eyes of 31 patients with coloboma-related retinal detachments enrolled. All the eyes underwent standard three-port pars plana vitrectomy with internal tamponade with nonheavy silicon oil in 24 (77.4%) or 20% sulfur hexafluoride (SF6) in 7 (22.6%) eyes. Endolaser photocoagulation at the borders of the coloboma was performed in all cases to isolate the colobomatous area. Encircling band was placed in 12 (38.7%) eyes based on surgeon's intraoperative judgment. Intraoperative lensectomy was performed in 16 (51.6%) eyes.

***Results*:** Mean (±SD) preoperative visual acuity (VA) was 2.41 (±0.73), which showed a statistically significant improvement to 1.78 (±0.93), postoperatively (P<0.001). Placement of encircling band significantly increased the chance of postoperative intraocular pressure (IOP) elevation (P=0.027). Retinal reattachment was ultimately achieved in 29 patients (93.5%) after an average of 1.32 operations per patient. Intractable glaucoma, retinal redetachment, band keratopathy, corneal decompensation and macular hole formation were the complications observed.

***Conclusion*:** Using vitrectomy techniques with intraoperative silicon oil or SF6 tamponade and endolaser photocoagulation at the borders of the coloboma is highly successful in anatomical retinal reattachment accompanied by significant visual improvement.

***Keywords*:** Choroidal Coloboma, Retinal Detachment, Vitrectomy

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Introduction

Choroidal colobomas result from faulty closure of the embryonic fissure. The reported incidence is 0.14% among general population, and 40% of affected individuals will develop retinal detachment during their lifetime.[1] The colobomatous area consists of a thin layer of hypoplastic retinal tissue. At this area, the choroid and retinal pigment epithelium are not developed and the sclera is usually thin and ectatic, producing a staphyloma.

Retinal detachments associated with choroidal coloboma are classified into two broad subtypes.[2] Type I detachments are caused by breaks outside the coloboma. These detachments are managed by conventional scleral buckling techniques. Type II detachments are caused by breaks inside the coloboma. These detachments are difficult to manage; retinopexy is not effective because of the absence of choroid and retinal pigment epithelium. In addition, the posterior location of the breaks makes them extremely difficult to buckle and preoperative identification of the breaks is difficult due to the lack of contrast in the colobomatous area and the nystagmus which is sometimes seen in these patients.

In this article, we present surgical outcomes of 31 eyes of 31 patients in whom pars plana vitrectomy with internal tamponade was performed to treat coloboma-related retinal detachments.

Methods

In a retrospective study of 31 patients (14 women and 17 men) admitted to a tertiary referral center between January 2000 and December 2005 with coloboma-related retinal detachments, we reviewed their medical records. Recorded variables including demographics, preoperative data, details of the procedure and postoperative data including follow-up visits were gathered. Retinal detachments caused by breaks outside the coloboma were excluded from the study. Visual acuities (VA) were recorded by the logarithm of the minimal angle of resolution (logMAR). A detailed fundus examination had been done pre and postoperatively, using a binocular indirect ophthalmoscope and a slit lamp biomicroscope with a +90 diopter (D) lens. A detailed retinal chart was drawn and details of the retinal detachment were noted. Data from the last follow-up examination was analyzed and compared with preoperative data using the Chi-square, paired samples correlations, and one-way ANOVA tests with SPSS software for Windows (Ver.11.0).

Surgical procedure

The operations were performed by twelve vitreoretinal surgeons in the retina service of Farabi Eye Hospital between January 2000 and September 2005. A standard three-port vitrectomy procedure was performed with a 4 mm infusion cannula and 20-gauge endosurgical instruments. At the beginning of the procedure, pars plana lensectomy was performed in 16 (51.6%) eyes due to either the presence of lens opacities or anterior proliferative vitreoretinopathies necessitating thorough vitreous base shaving. Encircling band was placed 9 to 12 millimeters from the limbus in 12 (38.7%) eyes in order to overcome tractional forces at the vitreous base. The placement of band was decided according to individual surgeon's judgment. Endolaser photocoagulation was done in all patients around the coloboma area in three to four rows of confluent grade 3 laser burns avoiding areas near the disc or papillomacular bundle. The presence of schlieren (subtle wavy lines that develop when clear fluids of different viscosities mix) during gas-fluid exchange was used as an important clue to identify the location of the break. Relaxing retinotomies were not required in order to flatten the retina. After gas-fluid exchange, intraocular tamponades were used in all patients consisted of a 20% mixture of sulfur hexafluoride (SF6) with air in 7 (22.6%) and nonheavy silicon oil in the remaining 24 (77.4%) eyes. Retinal reattachment was achieved in all (100%) patients at the conclusion of the procedure.

Results

The study population consisted of 14 women and 17 men indicating no gender predilection. The mean follow-up was 13 months (minimum 4 and maximum 32 months), twenty eight patients being followed for at least six months after surgery. Patients' mean age was 29.9

years (from 14 to 65 years). Preoperative logMAR VA ranged from light perception to 1 with a mean of 2.41. Preoperative intraocular pressure (IOP) ranged from 8 to 40 mmHg with a mean of 16.9 mmHg. Iris coloboma was present in 20 (64.5%), microcornea in 3 (9.6%), and nystagmus in 6 (19.3%) patients. In 6 patients (19.3%), the preoperative anterior segment examination was normal. In 7 patients (22.5%) the other eye was phthisic due to complications of choroidal coloboma. In 4 (12.9%) other patients, the fellow eye had choroidal colobomas and in the remaining 20 (64.5%), the other eye was normal. Proliferative vitreoretinopathy of grade C or higher was present in 5 (16.1%) eyes. 24 patients (77.4%) did not have previous surgeries but 7 patients (22.5%) had previously undergone surgical procedures to repair retinal detachments including 3 (9.75%) scleral buckling procedures and 4 (12.9%) pars plana vitrectomies.

Mean (±SD) preoperative VA was 2.41 (±0.73) which improved to 1.78 (±0.93); P<0.001. IOP increased from a preoperative mean (±SD) of 17.0 (±8.9) to 20.07 (±7.2), but this change was not statistically significant (P=0.075).

Whether the procedure was the first or a reoperation, did not correlate with patients' final VA (P=0.221), the level of visual improvement (P=0.143), and the chance of developing postoperative IOP elevation (P=0.325).

Performing lensectomy during the procedure did not affect the final visual outcome (P=0.199) or the mean postoperative IOP (P=0.193).

Placement of encircling band did not change the visual prognosis of the procedure (P=0.622), but significantly increased the likelihood of IOP elevation in the latest follow-up visit (P=0.027).

The type of intraocular tamponade used during the procedure did not have any impact on the visual prognosis (P=0.588) or the IOP (P=0.875).

Only one case of re-detachment was observed before silicon removal which was managed successfully with a reoperation consisting of membrane peeling, endolaser photocoagulation and heavy silicon oil injection. Two eyes were lost due to uncontrollable glaucoma which did not respond to any treatment. In five other eyes, silicon oil was removed after a mean postoperative period of 12.25 months, leading to two cases of redetachments (40%) which were further managed by reoperations. Other complications observed were macular hole formation in two cases, band keratopathy in two cases, and corneal decompensation in one case. Retinal reattachment was ultimately achieved in 29 patients (93.5%) after an average of 1.32 operations per patient. LogMAR VA at the latest follow-up examination ranged from no light perception to 0.5 with a mean of 1.8.

Discussion

Coloboma of choroid was first described by Von Ammon (cited by Duke-Elder) in 1831.[3] Retinal detachments caused by coloboma of the fundus are relatively rare.[4] The treatment of these patients had been difficult with external scleral buckling techniques as evidenced by the reported poor results.[1,2] This is because of the difficulty in identifying and closing the causative retinal breaks.

Schepens advocated drainage of subretinal fluid, followed by the production of chorioretinal adhesion around the periphery of the coloboma using cryopexy, photocoagulation, or both[3] Patnaik and Kalsi achieved reattachment of the retina in one case by applying a radial buckle which extended to the optic disc[5] when both sides of the coloboma are involved in the detachment, Wang and Hilton recommended that two radial buckles be applied along the two edges of the coloboma. They reviewed the literature and found that of 42 eyes with coloboma-related retinal detachment, 18 (43%) had successful reattachment. In their own series of 20 eyes, success was achieved in 7 eyes (35%). They concluded that vitrectomy with intraocular gas tamponade and creation of chorioretinal adhesion along the edge of the coloboma is the preferred treatment.[6] Silicon oil also has been used as a temporary tamponade in the management of these cases. In 1983, Gonvers reported the successful use of the silicon oil tamponade to treat one eye with a retina detachment associated with a choroidal coloboma.[7] Direct closure of the break with cyanoacrylate glue is advocated by Henneken et al[8] whereas others advocated

photocoagulation of the retina bordering the coloboma.[9] Michaels et al have advocated postoperative krypton laser treatment to the peripapillary portion of the coloboma to avoid damage to the nerve fiber layer at this critical location.[10]

To develop a rational approach to the treatment of these patients, one has to understand the pathogenesis clearly. Hermann Schubert, describing the histopathology of the colobomatous border in eight eyes, has shown that the retina splits into two layers near the margin of the coloboma.[11] This split in the layers of the retina has been identified at the level of inner nuclear layer or outer plexiform layer or both. The junction where this reversal occurs has been termed "locus minoris resistentiae". The intercalary membrane progressively becomes thinner as it is traced centrally. Breaks can occur at two locations in this altered anatomy of the fundus:[12]

1. at the locus minoris resistentiae:
Clinical identification of the breaks at this location would be impossible.

2. in the intercalary membrane:
Breaks in the intercalary membrane can be identified preoperatively or intraoperatively or both, although with some difficulty. Breaks at the intercalary membrane, whether or not associated with breaks at the locus minoris resistentiae, can cause retinal detachments that are able to extend beyond the boundaries of the coloboma. The above-mentioned breaks were addressed in our study. Retinal detachments caused by breaks in the peripheral retina were excluded because they are considered to be unrelated to the coloboma and therefore were managed by conventional scleral buckling techniques. Anatomical success as described by the presence of complete retinal reattachment at the latest follow-up visit was achieved in 29 patients (93.5%) which is comparable to most recent reports that incorporated similar surgical techniques.[11-19] Considering the fact that all patients with choroidal colobomas have some degrees of visual handicap attributable to the retinal pathology, mean (±SD) postoperative VA of 1.78 (±0.93) in our patients seems to be a satisfactory result specially when compared with the mean (±SD) preoperative VA of 2.41±0.73 ($P<0.001$). Redetachment of retina after silicon oil removal is reported to occur in 30-50% of patients with coloboma-related retinal detachments.[13] This happened in 40% of our cases. Because the number of cases who underwent silicon removal in our series was too small (5 patients), no definitive conclusions can be reached by the above-mentioned figures. Although not statistically significant, IOP elevations were commonly seen in the postoperative period of our patients, indicating the importance of careful IOP monitoring and timely treatment in such patients. In our series, placement of encircling band significantly increased the likelihood of IOP elevation in the latest follow-up visit ($P=0.027$), having no significant effect on final visual outcome. Therefore, it is reasonable not to advise against placement of encircling band in such patients. To our knowledge, the effect of encircling bands in IOP elevations among patients with choroidal coloboma has never been addressed in previous studies. Lensectomy did not alter the visual or anatomical outcome in our patients, so it should be reserved for situations in which presence of lens opacities interferes with intraoperative visualization or presence of anterior proliferative vitreoretinopathy that necessitates meticulous vitreous base shaving. The number of patients in whom gas tamponade was used was so small that no statistical comparison can be made between the gas and silicon tamponade groups.

Conclusion

Retinal detachments in colobomatous eyes are difficult to manage due to altered anatomy and physiology of the involved eye. Using vitrectomy techniques with intraoperative tamponade and endolaser photocoagulation at the borders of the coloboma is highly successful in anatomical retinal reattachment. Visual prognosis is also promising. Increased IOP is one of the main postoperative problems encountered in these patients.

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