Case report

Retinal pigment epithelium changes in pediatric patients with glaucoma drainage devices

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ABSTRACT

Purpose: Retinal changes secondary to hypotony are usually described as wrinkling or folding of the inner portion of the choroid, the retinal pigment epithelium (RPE), and the outer retinal layers in the macular area due to scleral wall collapse. We describe a new retinal finding in children with suspected hypotony after implantation of Baerveldt Glaucoma Implant (BGI).

Observations: Four patients in our series developed significant RPE defects after BGI implant. The RPE defects appeared as elongated white lines observed solely in the posterior pole, in no particular pattern, and seemed to be worse in infants with anterior segment dysgenesis and with collagen disorders.

Conclusion and importance: Children have thinner and more elastic scleral walls than adults. This characteristic may cause the inward scleral wall to collapse when the eye is hypotonic. The resulting redundancy of the retina leads to wrinkling and RPE defects characterized by hypopigmented lines predominantly in the macular area. Such findings, to our knowledge, have not been previously reported in pediatric patients.

1. Introduction

Glaucoma drainage devices such as the Baerveldt Glaucoma Implant (BGI) (Abbott Medical Optics, Santa Ana, CA) are used in pediatric glaucoma patients when control of the intraocular pressure (IOP) is not achieved with medical therapy or with angle surgeries. Success rates after BGI implantation have been reported to be 80–94% in several studies.\textsuperscript{1–7} However, postoperative complication rates associated with shunt surgeries remain high, with most early complications occurring secondary to post-operative hypotony, including choroidal effusions and/or hemorrhages, shallow or flat anterior chambers (AC) with or without induced aqueous misdirection, or retinal changes.\textsuperscript{7} Post-operative hypotony is a significant occurrence following pediatric BGI implantation, with greater than one-third of patients experiencing hypotony within the first 6 months following surgery.\textsuperscript{9}

There is a paucity of information regarding the specific, chronic retinal changes that can occur in pediatric patients following this complication, as well as their pathologic mechanism. Our purpose is to describe a new retinal finding in children after implantation of Baerveldt Glaucoma Implant. We suspect these changes occurred during a period of hypotony after implantation of the BGI.

2. Findings

2.1. Case 1

A seven-month old male with infantile glaucoma underwent trabeculectomies in both eyes within the first month of life. Due to poorly controlled intraocular pressure of 28 mm Hg, a BGI was implanted in the left eye. Five months later, an exam under anesthesia revealed an IOP of 11 mm Hg (Fig. 1) and hypopigmented RPE changes involving the temporal half of the posterior pole of the left eye (Fig. 2).

2.2. Case 2

A four-month old infant with SHOX gene mutation, and Peters anomaly in both eyes presented with congenital glaucoma and uncontrolled IOP of 40 mm Hg in the right eye and 30 mm Hg in the left eye. The patient underwent corneal transplants in both eyes and BGI placement on the right eye, followed by the left eye two weeks later.

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Thirteen months after surgery, an exam under anesthesia revealed an IOP of 9 mm Hg in the left eye (Fig. 3), as well as RPE changes in the posterior pole extending from the optic disc to the foveal area (Fig. 4).

2.3. Case 3

A one-year old female with an undefined collagen vascular disorder, hip dysplasia and club feet was referred for lens opacity in the left eye and bilateral corneal opacity and congenital glaucoma associated with Peter's Anomaly and Axenfeld Rieger's Syndrome. Due to uncontrolled IOP in both eyes of approximately 40 mm Hg, a pars plana vitrectomy and lensectomy with BGI placement was performed in the left eye, followed by a pars plana vitrectomy, penetrating keratoplasty and BGI placement on the right eye 4 weeks later. An exam under anesthesia 3 months later showed IOP on the left eye of 6 mm Hg (Fig. 5) and fundus RPE changes compromising the macular area (Fig. 6).

2.4. Case 4

A four-month old female presented with endogenous candida endophthalmitis on the right eye at 34 weeks gestational age. She underwent pars plana vitrectomy and lensectomy. Subsequently, she developed aphakic closed angle glaucoma with IOP 35 mm Hg and underwent BGI placement. On follow-up exams, IOP fluctuated between 21 and 32 mm Hg. Nine months later, an exam under anesthesia revealed IOP of 29 mm Hg (Fig. 7) and RPE changes in the macular area.
were noticed on the fundus examination (Fig. 8). Fluorescein angiography showed RPE changes.

3. Discussion

The differential diagnosis for patterns of alternate dark and light colored retinal streaks includes several diseases affecting Bruch’s membrane and the RPE. A choroidal rupture may be considered in cases following a closed globe injury from blunt trauma. Lacquer cracks are a possible diagnosis when the breaks in Bruch’s membrane are bilateral and occurring in highly myopic eyes. Breaks in a weakened Bruch membrane may also result in angiod streaks, which are viewed as bilateral, irregular subretinal lines radiating from the optic disc. However, in our series of cases the RPE changes were unilateral and first noticed after the placement of a BGI in the affected eye, suggesting ocular hypotony as the most probable cause.

Ocular hypotony is defined as low ocular pressure leading to functional and structural changes.\(^{10}\) Studies have reported that approximately 3.5%–27% of patients present with hypotony following glaucoma drainage device surgery.\(^{11-17}\) Patients with hypotony are at risk for vision loss through several mechanisms, including keratopathy, induced irregular astigmatism, cataract formation and progression, choroidal effusion, choroidal hemorrhage, optic nerve edema, and retinal changes.\(^{18}\)

The most commonly described retinal change is hypotony maculopathy. Dellaporta first reported this pathology in 1954, through a series of four cases of patients with findings of hypotony associated with papilledema ex vacuo, vascular tortuosity, and chorioretinal folds in the macular area, usually occurring after antiglaucomatous surgery or perforating eye injuries.\(^{19}\) In this condition, the chorioretinal folds present in a radial distribution around the fovea, due to the thicker perifoveal retina surrounding the very thin foveal center. These folds then result in distortion of the layers and components of the retina, with possible compression of pigment epithelial cells in the trough of the fold and thinning of this cell layer over the crest. These changes are thought to produce the alternate dark and light colored streaks observed in these cases.\(^{20}\) As IOP normalizes, these folds may completely disappear.\(^{20}\) Sometimes, however, changes in the posterior pole may

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**Fig. 4.** A. Fundus picture before Baerveldt Glaucoma Implant placement. B. Retinal pigment epithelium changes noticed after Baerveldt Glaucoma Implant placement.

**Fig. 5.** Case 3 IOP measurements at time of BGI placement and of retinal findings.

**Fig. 6.** Retinal pigment epithelium changes noticed after Baerveldt Glaucoma Implant placement.
persist due to anatomical alterations in the retinal pigment epithelium and fibrosis within the retina, choroid or sclera.

In our series, the RPE changes were noticed mainly around the macular area, as in hypotony maculopathy. However, noticeable differences can be observed in the thickness, distribution, and pattern of these findings. In all cases, RPE changes consisted of thick hypopigmented lines without a specific pattern, mainly localized to the macular area but with extension to the temporal retina and superior to the optic nerve.

To our knowledge, these findings have not been previously described in the literature. Hence, pathologic studies are necessary to determine the histological changes taking place. Based on the clinical findings, we postulate that, similar to the mechanism of hypotony maculopathy, IOP changes after glaucoma device implantation cause the inward scleral wall to collapse, resulting in redundancy and wrinkling of the retina, with subsequent RPE defects. In infants, the sclera is significantly thinner than in adults, measuring approximately 0.45 mm in neonates and increasing to 1 mm in adults. It is also up to four times more pliable than in adults, with approximately a half of the tensile strength. These differences explain the structural changes seen in congenital glaucoma such as increased axial length and buphthalmos. Due to these structural differences of the scleral wall in infants, the vitreous cavity may have a greater tendency to collapse following the rapid decrease in intraocular pressure after BGI implantation. This would lead to increased, irregular wrinkling and RPE defects at higher pressures than in adults. As the wrinkling or folding becomes more prominent, there may be increased displacement of RPE cells over the crest of the fold. As the IOP normalizes, the folds disappear but the RPE changes might remain as hypopigmented irregular lines in the posterior pole. The fluorescein angiography findings in one of these patients include hyperfluorescent streaks secondary to thinned RPE, supporting this hypothesis. Optic coherence tomography studies would have provided useful information. However, the challenging media (opaque cornea) resulted in poor quality study results.

Although we did not record hypotony values for one of our patients (case 4), we assume that the mechanism is a similar one as for the other three cases, due to the resemblance in clinical findings. Given that reliable IOP values were obtained mainly during EUAs, it is possible that the hypotony period might have been missed in between monthly and tri-monthly exams. Another possibility might be that the highly variable IOP changes, with rapid decreases and increases in IOP, could have caused inward scleral collapse in a similar manner as in hypotony.

4. Conclusions

We postulate that the retinal findings in this small case series may be a result of retinal pigment epithelium defects after variation in intraocular pressure. The increased thickness and irregular distribution of these hypopigmented changes may be secondary to increased scleral elasticity in infants. Such findings, to our knowledge, have not yet been reported in pediatric patients. The effect of these changes in the vision and visual development of these young patients is yet to be determined.

Patient consent

The patients provided written consent for publication of case information including medical record details and photographs. This study was approved by the Institutional Review Board of University of Miami: IRB ID 20150996.

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Conflicts of interest

The following authors have no financial disclosures: CJO, SG, MPF, CVV, SA, TCH, EH, SRD, AB.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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