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Case report

Pars plana vitrectomy in the management of ghost cell glaucoma

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Abstract

In this report we describe, herewith, 5 cases of ghost cell glaucoma that followed spontaneous vitreous hemorrhage complicating branch retinal vein occlusion in a phakic eye (one case), and traumatic vitreous hemorrhage (4 cases). Because intraocular pressure was uncontrolled, pars plana vitrectomy was performed to remove the reservoir of ghost cells. Vitrectomy resulted in successful control of intraocular pressure without medications and visual improvement.

Introduction

Ghost cell glaucoma is a recently described form of secondary open angle glaucoma that occurs as a consequence of vitreous hemorrhage. Following vitreous hemorrhage, the red blood cells (RBCs) degenerate into ghost blood cells (GBCs) and pass slowly into the anterior chamber through a disruption in the anterior hyaloid face [1]. The pressure rise is due to primary obstruction of the intertrabecular spaces by the nonpliable GBCs [2, 3].

When medical therapy fails in cases of dense and old vitreous hemorrhage, vitrectomy is effective in controlling the intraocular pressure by removing the source of ghost cells and also in improving vision by clearing the intraocular opacities [4]. In this report we present our experience in 5 cases with this infrequent indication for pars plana vitrectomy.

Case 1

A 45-year-old hypertensive man was referred with the chief complaint of decreased vision, severe pain, and redness in the right eye of one week duration. The patient underwent laser photocoagulation for branch retinal vein occlusion in the right eye elsewhere 5 months prior to presentation that was followed by vitreous hemorrhage. Examination revealed a visual acuity of light perception, with good light projection in all quadrants in the right eye and 20/20 in the left eye. The intraocular pressure was 48 mmHg in the right eye and 14 mmHg in the left eye. Examination of the left eye was normal. The right eye had moderate ciliary injection, and slit lamp examination showed mild bullous keratopathy and corneal stromal edema. The anterior chamber showed 4+ khaki-colored GBCs. The GBCs layered in the angle inferiorly, creating a 20% khaki-colored 'pseudohypopyon'. There were no evidence of rubeosis iridis. The lens showed mild posterior subcapsular opacities. There was no lens sublaxation, and no clinical evidence of anterior hyaloid face disruption. On gonioscopic examination the anterior chamber angle was open without evidence of neovascularization. The anterior vitreous gel showed dense yellow-colored blood. The retina could not be visualized because of the dense vitreous hemorrhage. B-scan ultrasonography showed a dense vitreous hemorrhage without evidence of retinal detachment. A clinical diagnosis of ghost cell glaucoma was made. Despite maximum antiglaucoma therapy intraocular pressure was 30 mmHg. A pars plana vitrectomy was performed and as much of the opaque vitreous gel as possible was excised. The retina was seen to be attached and the macula appeared normal. The blood vessels along the upper temporal arcades were noted to be sheathed with

patches of neovascularization. Argon laser endophotocoagulation was applied in a scatter pattern along the distribution of the obstructed upper temporal vein. One year after surgery the intraocular pressure was 12 mmHg without medications, and visual acuity was 20/30.

Case 2

An 8-year-old boy was struck in the left eye by a piece of stone 5 days prior to referral. Examination revealed a visual acuity of 20/20 in the right eye and light perception, with good light projection in all quadrants in the left eye. The intraocular pressure was 16 mmHg in the right eye, and 35 mmHg in the left eye. Examination of the right eye was unremarkable. The left eye showed total hyphema. B-scan ultrasonography revealed dense vitreous hemorrhage and flat retina. Sickle cell disease was ruled out. The patient was treated with oral acetazolamide, topical timptol 0.5%, and propine 0.1%, and the antifibrinolytic agent tranexamic acid. Twelve days later, the hyphema cleared, and the intraocular pressure was controlled. The antiglaucoma medications and tranexamic acid were discontinued. Gonioscopy revealed an open angle for 360° without recession. Two weeks later, the patient presented with a painful, red left eye. Visual acuity was counting fingers at 2 feet. The intraocular pressure was 40 mmHg. Slit lamp examination revealed moderate ciliary injection, and mild, diffuse bullous keratopathy. The anterior chamber showed 4+ GBCs. The lens was clear without evidence of sublaxation. There were no clinically detectable discontinuities of the anterior hyaloid face. Dense hemorrhage was present in the anterior vitreous gel. The retina could not be visualized because of the dense vitreous hemorrhage. B-scan ultrasonography revealed a dense vitreous hemorrhage with a shallow retinal detachment noted inferiorly. A diagnosis of ghost cell glaucoma was made and the patient underwent a pars plana vitrectomy. A retinal dialysis was noted inferiorly, that was treated with cryotherapy and supported on a scleral buckle. An intraocular gas bubble was used to tamponade the retinal break during the initial postoperative time. Postoperatively the eye healed without complications. The retina became reattached, and the retinal break was sealed. When the patient was last seen one year later, the intraocular pressure was 14 mmHg without medications, and visual acuity was 20/80.

Case 3

A 38-year-old male was referred 5 days after repair of a corneo-scleral laceration that followed blunt trauma to the right eye. Examination revealed a visual acuity of counting fingers at one foot in the right eye, and 20/30 in the left eye. The intraocular pressure was 34 mmHg in the right eye, and 15 mmHg in the left eye. Examination of the left eye was unremarkable. Slit lamp examination of the right eye showed moderate ciliary injection, and the corneo-scleral laceration seemed well healed. The anterior chamber showed 60% hyphema. There were posterior synechiae and the lens was clear. B-scan ultrasonography revealed a dense vitreous hemorrhage. No evidence of retinal detachment or an intraocular foreign body was seen. Sickle cell disease was ruled out. The patient was treated with antiglaucoma medications, and topical atropine and steroid compounds. Twenty days later, the intraocular pressure of the right eye was 30 mmHg despite maximum medical therapy. The anterior chamber showed 4+ GBCs. The hyphema cleared, but a dense yellow fibrinous clot was left on the anterior lens capsule in the pupillary area. Gonioscopy revealed an open angle for 360° without recession. A clinical diagnosis of ghost cell glaucoma was made. Tissue plasminogen activator 25 μg in 0.1 ml was injected into the anterior chamber, that resulted in complete dissolution of the fibrinous clot. The lens was clear, and there was no sublaxation. There was no clinical evidence of anterior hyaloid disruption. A dense yellowish brown blood was seen in the anterior vitreous gel. Two days later, a pars plana vitrectomy was performed on the right eye. The vitreous hemorrhage was removed and no complications occurred. Postoperatively, vision improved to 20/60, and intraocular pressure was 20 mmHg without antiglaucoma medications during a follow-up of 7 months.

Case 4

A 20-year-old male was referred with the chief complaint of decreased vision, pain, and redness in the right eye. The patient sustained a blunt trauma to the right eye by a wooden stick 25 days prior to presentation. Examination revealed a visual acuity of hand motions, with good light projection in all quadrants in the right eye and 20/25 in the left eye. The intraocular pressure was 47 mmHg in the right eye and 12 mmHg in the left eye. Examination of the left eye

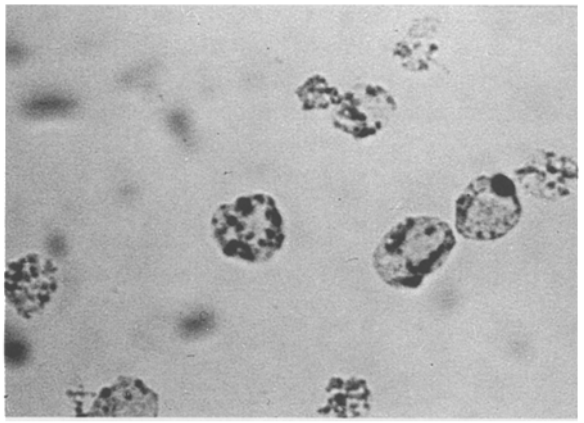

Fig. 1. Light micrograph of vitreous sample showing ghost red blood cells with Heinz bodies (Giemsa stain × 100).

was normal. The right eye had moderate ciliary injection, and slit lamp examination showed epithelial and stromal corneal edema. The anterior chamber showed 4+ GBCs. The GBCs layered in the angle inferiorly, creating a 10% khaki-colored 'pseudohypopyon'. Gonioscopy showed a small area of peripheral anterior synechiae inferonasally and about 20% angle recession. The lens was clear and there was no sublaxation. There were no clinically detectable discontinuities of the anterior hyaloid face. A dense yellow-colored blood was present in the anterior vitreous gel. The retina could not be visualized because of the dense vitreous hemorrhage. B-scan ultrasonography revealed dense vitreous hemorrhage and flat retina. Screening for sickle cell disease was negative. A clinical diagnosis of ghost cell glaucoma was made. The intraocular pressure was inadequately controlled on maximum antiglaucoma therapy. A pars plana vitrectomy was performed on the right eye. The optic disc was healthy, the retina was seen to be attached, and the macula appeared normal. Giemsa staining of the vitreous specimen revealed the presence of typical ghost cells with Heinz bodies (Fig. 1). Six months after surgery, the intraocular pressure was 17 mmHg without medications, and visual acuity was 20/20.

Case 5

A 10-year-old boy sustained a trauma to the left eye by a piece of stone. The visual acuity was 20/30 in the right eye and light perception in the left eye. The intraocular pressure was 12 mmHg in the right eye, and 38 mmHg in the left eye. Examination of the right eye was unremarkable. The left eye showed 70% hyphema. B-scan ultrasonography revealed dense vitreous hemorrhage. No evidence of retinal detachment was seen. Two weeks later, evacuation of the anterior chamber clot was performed. Fifty days after the procedure, the patient was referred to us for consultation. Visual acuity in the left eye was hand motions, with good light projection in all quadrants. The intraocular pressure was 30 mmHg. Slit lamp examination showed a central localized rounded area of corneal blood staining 1.5 mm in diameter. The anterior chamber showed 4+ GBCs. The lens showed moderate posterior subcapsular opacities and a localized anterior capsular opacity, and there was no sublaxation. A dense yellow-colored blood was present in the anterior vitreous gel. The retina could not be visualized because of the dense vitreous hemorrhage. Gonioscopy revealed the presence of 30% angle recession. B-scan ultrasonography showed dense vitreous hemorrhage without evidence of retinal detachment. A diagnosis of ghost cell glaucoma was made. The patient underwent a pars plana lensectomy, and vitrectomy. During surgery, a chorioretinal scar was noted in the macular area. One month later, the intraocular pressure was 16 mmHg without antiglaucoma medications, and visual acuity improved to 20/100.

Comment

Ghost cell glaucoma is an unusual and often overlooked cause of persistent elevation of intraocular pressure following vitreous hemorrhage. Following vitreous hemorrhage, fresh biconcave pliable red blood cells are converted within one to two weeks into more rounded less pliable khaki-colored ghost cells. Intracellular hemoglobin leaves the cells and precipitates extracellularly as large clumps that become enmeshed within, and adherent to vitreous strands. The remaining intracellular hemoglobin aggregates into clumps, known as Heinz bodies, around the inner surface of the plasma membrane of the ghost cells [1, 2].

Ghost cells pass into the anterior chamber whenever the anterior hyaloid face is disrupted, as seen following vitrectomy, cataract extraction, and penetrating or blunt trauma [2, 4–6]. Clinically, these cells are frequently mistaken for white blood cells, leading to the erroneous diagnosis of uveitis. The intraocular pressure rise is due to direct mechanical obstruction of the

trabecular meshwork by the nonpliable GBCs. GBCs were found to be far more effective in causing trabecular meshwork obstruction in vivo than fresh RBCs [1, 3].

The intact hyaloid face constitutes an effective barrier to the forward migration of GBCs [6]. Spontaneous vitreous hemorrhage in phakic eyes is a rare cause for ghost cell glaucoma. Ghost cell glaucoma developed in Case 1 following vitreous hemorrhage complicating branch retinal vein occlusion. The patient was phakic with no prior ocular trauma or surgery. Spontaneous disruption of the anterior hyaloid face is postulated which allowed the passage of GBCs into the anterior chamber. Five similar cases of ghost cell glaucoma in phakic eyes after spontaneous vitreous hemorrhage were reported [7–9].

In cases 2, 3, 4, and 5 ghost cell glaucoma followed traumatic vitreous hemorrhages. The patients 2, 3, and 5 had vitreous hemorrhages in addition to anterior chamber hemorrhages. The initial hyphema was associated with a high initial pressure. This was followed by the appearance of GBCs in the anterior chamber that occurred at a time when the hyphema was cleared or almost cleared. The ghost cell stage caused a persistent elevation of intraocular pressure or a second elevation after returning of pressure toward normal. Campbell [6] noted that ghost cell glaucoma was not seen in a pure, anterior chamber hemorrhage. In the anterior chamber, red blood cells retain their red color for long periods of time, often many months, whereas red cells quickly change hue in the vitreous cavity. Higher levels of oxygen and nutrients, coupled with a more rapid circulation of fluid in the anterior chamber may constitute reasons for this difference. Also, quantitatively, the vitreous cavity can hold much more blood than the anterior chamber and can therefore provide many more GBCs following ocular hemorrhage. These factors may explain the observation that vitreous hemorrhage seems to be necessary for the development of ghost cell glaucoma.

Control of ghost cell glaucoma requires surgical treatment in most cases because the intraocular pressure remains elevated despite maximum medical therapy [6]. Removal of the GBCs from the anterior chamber by irrigation is successful although temporary measure to lower the intraocular pressure. This procedure can be repeated several times [1, 6]. Frazer et al. [8] controlled the glaucoma in two cases by trabeculectomy. Pars plana vitrectomy is effective in controlling the intraocular pressure by removing the reservoir of GBCs. In addition, the procedure improves vision by clearing the intraocular opacities [4, 10]. It is necessary to excise as much vitreous hemorrhage as possible in order to remove the reservoir of GBCs. Otherwise the elevated intraocular pressure can recur postoperatively [4].

In conclusion, we are reporting 5 cases of ghost cell glaucoma that were treated by pars plana vitrectomy. Vitrectomy resulted in control of the intraocular pressure and visual improvement in all the cases.

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