

## CLINICAL INVESTIGATIONS

# Acute Angle-Closure Glaucoma Resulting From Spontaneous Hemorrhagic Retinal Detachment in Age-related Macular Degeneration: Case Reports and Literature Review

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**Purpose:** Acute angle-closure glaucoma resulting from massive subretinal hemorrhage is a rare and catastrophic complication in age-related macular degeneration. Anticoagulant usage had been strongly correlated with this complication in previously reported cases.

**Methods:** Four patients (4 eyes), 3 men and 1 woman, developed angle-closure glaucoma with diffuse subretinal hemorrhage and total hemorrhagic retinal detachment.

**Results:** Serial funduscopic examinations and echographic studies in 2 eyes showed that the blood gradually accumulated in the subretinal space. It took more than 10 days for the bleeding to build up to bullous hemorrhagic retinal detachment and secondary glaucoma. Antiglaucomatous agents were given and sclerotomy was performed in 3 of the 4 patients. Phthisical changes were observed subsequently in these 3 eyes. The eye that received early drainage of blood was an exception, and a small degree of residual acuity was retained. Three of the 4 patients had diabetes mellitus, and hypertension and vascular diseases were also present in the same 3 patients.

**Conclusions:** Diabetes mellitus might be a predisposing factor for the impaired hemostasis. Anti-glaucomatous agents were of no effect in the management of intraocular pressure. Sclerotomy and drainage of blood help control intraocular pressure and relieve ocular pain. Poor final visual acuity is inevitable. However, phthisical changes might be prevented with early sclerotomy and drainage of blood. **Jpn J Ophthalmol 2001;45:270–275** © Japanese Ophthalmological Society

**Key Words:** Acute angle-closure glaucoma, age-related macular degeneration, diabetes mellitus, retinal detachment, sclerotomy.

## Introduction

Spontaneous massive subretinal hemorrhage precipitating angle-closure glaucoma rarely occurs. To date, only 18 cases have been reported.<sup>1–11</sup> Bullous hemorrhagic retinal detachment or choroidal detachment pushes the lens-iris diaphragm forward, resulting in angle-closure glaucoma. Subretinal neovascularization is considered to be the bleeding source in most cases. Here we report another 4 patients with this disease entity. Clinical course, predisposing factors, and therapeutic strategies are discussed.

## **Case Reports**

## Case 1

A 57-year-old male patient came to our clinic on May 2, 1997, complaining of sudden onset of blurred vision in the right eye about 1 week previously. He had suffered from hypertension and diabetes mellitus for several years and had had a cerebrovascular episode 2 years earlier. Visual acuity was 6/100 in the right eye and 8/20 in the left eye. The intraocular

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pressure was within normal range bilaterally. The angles were 360° open by gonioscopic examination in both eyes. Funduscopic check disclosed submacular hemorrhage and choroidal neovascularization in the right eye, and some hard drusen at the posterior pole in the left eye. Fluorescein angiography performed 4 days later showed choroidal neovascularization at the parafoveal area in the right eye; the subretinal hemorrhage was progressing at this time as compared to his previous picture (Figure 1). Vision in his right eye continued to deteriorate while subretinal hemorrhage accumulated during following visits. Massive subretinal hemorrhage with total hemorrhagic retinal detachment and vitreous hemorrhage were noted by ultrasound on May 30 (Figure 2). Visual acuity in his right eye at this time was hand motion. To rule out the possibility of excessive bleeding, bleeding time and coagulation profiles were checked, and all were within normal ranges. Biochemical tests revealed an elevated triglyceride level (801 mg/dL). On June 8, the patient came to our emergency room due to severe pain in the right eye. Visual acuity in the right eye was negative for light sense. A collapsed anterior chamber, microcystic corneal edema and elevated intraocular pressure of 50 mm Hg were noted. Echographic study in the right eye at that time revealed bullous hemorrhagic retinal detachment, which completely occupied the vitreous space (Figure 3). Although under he was under full anti-glaucomatous medication, intraocular pressure could not be lowered. He was then lost to follow-up. He came back to our clinic 2 months later. A blood-stained cornea, collapsed anterior chamber, and undetectable low intraocular pressure were noted in the right eye. Echographic study revealed



**Figure 2.** B-scan ultrasound display of right eye showing massive subretinal hematoma on lower part with total hemorrhagic retinal detachment (case 1) on May 30, 1997.

that the subretinal hemorrhage had resolved. No intraocular malignancy was detected with either ultrasound or orbital computed tomography (CT). His right eye became phthisical soon thereafter.

#### Case 2

A 78-year-old male patient had progressively blurred vision in the left eye for 2 months. He had visited another hospital where age-related macular degeneration was diagnosed. Severe ocular pain in his left eye and elevated intraocular pressure had developed 1 month earlier. Although a local doctor had given him full anti-glaucomatous agents, his pain could not be relieved. He then came to our clinic for help. He was suffering from coronary arterial disease, and a coronary artery bypass graft had been



**Figure 1.** Fundus photograph shows choroidal neovascularization with subretinal hemorrhage in right eye of case 1.



**Figure 3.** B-scan ultraound display of right eye showing bullous hemorrhagic retinal detachment (case 1) on June 8, 1997.

performed 2 years previously. He had stopped taking medication 1 year after the bypass surgery. He denied having other systemic diseases, including hypertension or diabetes mellitus. On ophthalmologic examination, his visual acuity was 20/40 in the right eye but only light-sense negative in the left eye. Intraocular pressure was 12 mm Hg in the right eye and 50 mm Hg in the left eye. A collapsed anterior chamber, blood-stained cornea, and microcystic corneal edema were noted in the left eye. Gonioscopic examination of the right eye showed that the angle was open at all four quadrants. Funduscopic check revealed a mottled macula in the right eye. Examination was not possible in the left eye due to media opacity. Echographic study in the left eye showed diffuse subretinal or choroidal hemorrhage. Ultrasound and orbital CT did not reveal any indication of intraocular malignancy. Bleeding time, coagulation profiles, and results of biochemistry studies were all within normal ranges. Blood was drained through sclerotomy, and the anterior chamber was reformed in the left eye. A massive amount of semisolid, dark blood was drained from the sclerotomy site. Intraocular pressure was successfully lowered with this procedure, and ocular pain was greatly relieved. Echographic studies showed that the subretinal hemorrhage had been much reduced. The left eye subsequently became phthisical.

## Case 3

A 55-year-old male patient was referred to our clinic on May 15, 1998, with a tentative diagnosis of age-related macular degeneration with subretinal hemorrhage in the right eye. His visual acuity was counting fingers on referral. When he arrived at our clinic 2 days later, his visual acuity in the right eye had dropped to hand motion. A funduscopic check showed a massive subretinal hematoma at the submacular area. Results of a fundus check in the left eye were normal. Gonioscopic check showed widely open angles for both eyes. He had complained of a rapid loss of vision and visual field during the past week. Echographic study in the right eye showed hemorrhagic retinal detachment at the temporo-inferior aspect of the disc (Figure 4). According to the patient, he suffered from hypertension, which had been under regular medical control for more than 10 years. He denied other systemic disease. However, in a routine biochemical check, hyperglycemia and elevated serum triglycerides were noted. Results of checks for bleeding time and coagulation profiles were both within normal ranges. The hemorrhagic



**Figure 4.** B-scan ultrasound display of right eye showing diffuse subretinal hemorrhage (case 2).

retinal detachment in the right eye became total on May 18 (Figure 5) and was noted to be retrolenticular (Figure 6) on May 24. A progressively shallow anterior chamber and elevated intraocular pressure in the right eye developed thereafter. Although the patient was under full anti-glaucomatous medication, intraocular pressure could not be well controlled (it was around 42 mm Hg). Visual acuity in his right eye was negative for light sense at this time. We then performed sclerotomy. A massive amount of dark, thick blood was drained. The subretinal hematoma was much reduced and the anterior chamber was deepened after this procedure. Intraocular pressure returned to normal with no medication. The subretinal blood gradually resolved. However, a shallow inferior retinal detachment persisted (Figure



**Figure 5.** B-scan ultrasound display of right eye on May 22, 1998 (case 3) showing that hemorrhagic detachment is progressing.



**Figure 6.** B-scan ultrasound display of right eye on May 25, 1998 (case 3) demonstrating retrolental hemorrhagic retinal detachment.

7). Dense posterior subcapsular opacity developed in the right eye. His visual acuity was hand motion at the end of follow-up.

## Case 4

A 67-year-old female patient was referred to our clinic due to uncontrolled glaucoma in the left eye for 1 month. She had had progressive blurred vision in the left eye for 2 years. Age-related macular degeneration with disciform scar was noted at that time. Her systemic history was positive for diabetes mellitus, hypertension, and viral hepatitis C. She had had rapid deterioration of vision followed by severe eye pain in her left eye for 1 month before she visited our clinic. A collapsed anterior chamber and elevated intraocular pressure (up to 70 mm Hg) were



**Figure 7.** B-scan ultrasound display of right eye in January of 1999 (case 3) showing that hemorrhage had completely reabsorbed with residual shallow retinal detachment.

noted by a local doctor at that time. She was suspected of having malignant glaucoma, although no previous ocular surgery had been performed. After unsuccessful control with anti-glaucomatous medications, she had received a filtering procedure. While doing this procedure, the local doctor noted that dark blood was spurting from the wound, and he immediately closed the wound. The patient still suffered from elevated intraocular pressure and eye pain. One month later, she was referred to our clinic. At our clinic, a blood-stained cornea with a folded Descemet's membrane and collapsed anterior chamber were noted in the left eye. The intraocular pressure was 38 mm Hg under full anti-glaucomatous agents. Gonioscopic check of the right eye showed that the angle was grade-one narrow superiorly and widely open at the other quadrants. Ultrasound showed diffuse subretinal hemorrhage and total hemorrhagic retinal detachment. Drainage of blood through sclerotomy and reforming of the anterior chamber were performed. Her ocular pain was relieved, and the intraocular pressure was lowered with no medication after the sclerotomy. The eye subsequently became phthisical.

## Discussion

Spontaneous massive hemorrhagic retinal or choroidal detachment resulting in angle closure glaucoma seldom occurs. Only 18 cases have been reported previously.<sup>2-12</sup> Bullous hemorrhagic retinal or choroidal detachment pushes the lens-iris diaphragm forward resulting in angle-closure glaucoma. According to previous reports,<sup>1-5,7-13</sup> disciform scar with choroidal neovascularization in age-related macular degeneration is the bleeding source in most cases. El Baba et al<sup>1</sup> made a histopathologic study of 7 patients with massive subretinal hemorrhage. They concluded that the origin of the hemorrhage was from the disciform scars. Rupture of the large choroidal vessels that extend into the disciform scar accounts for the massive hemorrhage that erupts through all layers of the retina and into the vitreous. Gass and Wolter<sup>2</sup> described similar histologic findings and reached similar conclusions in two cases with massive subretinal hemorrage. The process presumably starts from choroidal neovascularization, which actively leaks transudate. The serous or serosanguineous exudate causes retinal pigment epithelial detachment. As the process goes under the disciform scar, it tears a choridal vessel that feeds the disciform scars and hemorrhaging begins. In our series, all four cases were diagnosed as, or suspected of having, age-related macular degeneration either at the time of presentation or previous to this event. All patients experienced rapidly progressive visual decline and visual field defect within 2 to 4 weeks followed by severe eye pain and elevated intraocular pressure.

We made serial funduscopic and echographic recordings after the initial stage of bleeding in cases 1 and 3. We found that the bleeding did not follow a rapid course as occurs in expulsive suprachoroid hemorrhage in intraocular surgery. The bleeding also did not occur in a hypotensive state, which is the prerequisite in most cases for postoperative suprachoroidal hemorrhage. It took about 2 to 4 weeks for the spontaneous bleeding to build up to a retrolenticular hematoma under normal intraocular pressure. Impaired local hemostasis is responsible for the uncontrolled bleeding. Impaired hemostasis as a predisposing factor for massive hemorrhage complicating macular degeneration is discussed in several reports.<sup>1,4,5,10–15</sup> According to the literature,<sup>1</sup> 19–27% of patients with hemorrhagic complications were taking Coumadin or NSAID.<sup>1,14,15</sup> In another report,<sup>3</sup> 6 of 17 patients with acute angle-closure glaucoma resulting from massive subretinal hemorrhage were using oral anticoagulants. Contrary to reports in the literature, none of our patients was taking any anticoagulants. This discrepancy is either due to the limited cases in our series or due to the fact that coronary or thrombotic diseases are much less prevalent in Asians, so there are fewer patients here taking anticoagulants than in Western countries.

Diabetes mellitus is seldom mentioned as a predisposing factor; however, 3 of the 4 patients in our series had diabetes mellitus. The difficulty of maintaining hemostasis during surgery in diabetes mellitus patients is well-known to surgeons. This might contribute to the impaired hemostasis in massive bleeding. Hypertension is also considered a predisposing factor for massive subretinal hemorrhage, 1,13,14,15 although no definite support could be found from the epidemiologic study. In our cases, 3 of the 4 patients had hypertension. Vascular diseases are other possible risk factors for massive bleeding, which were present in 2 of the 4 patients in our series. One had a cerebral vascular episode and the other had coronary artery disease. Atherosclerotic changes in vessels in patients with hypertension, diabetes mellitus, or cardiovascular diseases would cause the choroidal vessels to be more easily torn in the process of retinal pigment epithelial detachment.

Treatment of angle-closure glaucoma resulting from massive subretinal hemorrhage is frustrating. Medical treatment with anti-glaucomatous agents is rarely effective. Most of the previously reported patients underwent enucleation to relieve the intractable pain. Some patients were suspected having an intraocular tumor.<sup>2,3,5–7,9–11</sup> Others either received retrobulbar injection of alcohol, or cyclodestructive procedures for pain relief.<sup>3</sup> Only 2 eyes underwent the procedures of drainage of blood and reformation of the anterior chamber.<sup>8,11</sup> One eye retained the visual acuity of hand motion with normal ocular appearance and the other eye became phthisical. We performed sclerotomy and reformation of the anterior chamber in 3 of the 4 eyes of our patients. One eye had a final visual acuity of hand motion and the other two had phthisical changes with no light perception. The eye with no surgical intervention became phthisical without light perception. Timing of surgery might explain the different results. In the eye retaining hand-motion vision, we did not wait a long time before initiating drainage. This quickly restored normal intraocular pressure. In the other 2 eyes, the intraocular pressure had been elevated for at least 1 month before the patients came for help.

Prolonged elevated intraocular pressure impairs intraocular blood flow and induces ischemic changes in intraocular structures. It is not possible for the ciliary body to be restored to normal function even though the hematoma is eliminated later. Therefore, we recommend early drainage of hemorrhage as long as anti-glaucomatous agents cannot control intraocular pressure. Although it is not possible for good vision to be restored through this procedure, it is effective for pain relief. Phthisical changes might also be prevented.

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