a hydrophilic acrylic intraocular lens was implanted in the capsular bag. The early postoperative period was uneventful, and the patient achieved a best-corrected visual acuity of 20/30. Within the next 2 months, there developed an excessive capsular bag fibrosis with mild upward decentration of the intraocular lens.

Comment. Although hyphema is one of the most common early postoperative complications following trabeculectomy, to our knowledge, intralenticular collection of blood has not been previously reported. Because of the use of an operating microscope and the refinement of surgical techniques, lens injury during trabeculectomy has been infrequently reported. We hypothesized that there had been anterior capsule injury while performing peripheral iridectomy in this case, with seepage of blood into the capsular bag.

The development of a fibrous type of posterior capsule opacification in relation to the presence of blood in the capsular bag, as was evident from the exaggerated postoperative capsular bag fibrosis in this case, has been previously noted. This case highlights the possibility of lens injury during trabeculectomy and provides an insight into the problems encountered while performing phacoemulsification when there is intralenticular blood collection.

G. S. Brar, MS
Jagat Ram, MS
Jaspreet Singh, MS
Ravinder Kaur, MS
Amod Gupta, MS

The authors have no relevant financial interest in this article.

Correspondence: Dr Brar, Department of Ophthalmology, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India (eye@pgi@satyam.net.in).


---

**Episodic Elevations in Intraocular Pressure Associated With Blood in the Schlemm Canal**

Elevated intraocular pressure (IOP) associated with blood in the Schlemm canal may be associated with many different conditions. To our knowledge, we describe a previously unreported case of a patient with recurrent, intermittent spikes in IOP associated with episodic and transient blood in the Schlemm canal. Whenever the patient was examined because of elevated IOP in one or both eyes, gonioscopy disclosed the presence of blood in varying quadrants of the Schlemm canal in the affected eye(s). Whenever IOP was normalized, with or without medical therapy, the blood was no longer visualized.

**Report of a Case.** A 38-year-old woman of Latin American descent sought medical care because of right ocular pain, redness, and decreased visual acuity of approximately 12 hours’ duration. The pain was not relieved by ibuprofen. The patient reported a history of similar ocular episodes that had occurred intermittently during the preceding 2 years. Her medical history was significant for systemic hypertension and left elbow surgery; her blood pressure was 160/90 mm Hg at the time of examination. There was no significant ocular, family, or social history.

On initial examination, visual acuities were 20/40 OD and 20/25 OS, with a mid-dilated, minimally reactive pupil in the left eye. We found no proptosis, orbital bruits, or pulsating exophthalmos. The right conjunctival region had a mild red appearance, and mild corneal edema or haze was noted in the right eye. There was no apparent dilatation of the episcleral veins. The left anterior segment appeared normal. Goldmann applanation tonometry measurements were 55 mm Hg OD and 17 mm Hg OS. In the right eye, gonioscopy showed open angles with blood in the Schlemm canal inferiorly, temporally, and superiorly (Figure). Gonioscopic findings of the left angle structures were unremarkable. Dilated funduscopic examination of the right eye showed mild vascular tortuosity without leakage on fluorescein angiography. Ophthalmodynamometry measurements were 70 mm Hg OD and 60 mm Hg OS (both within normal limits). The elevated IOP was initially treated with an intensive course of topical medications and oral agents (eg, acetazolamide and glycerin). Once the IOP was normalized, the patient was prescribed an antiglaucoma regimen of timolol maleate, latanoprost, and dorzolamide hydrochloride.

During the next 5 months, the patient experienced recurrent, in-
termittent episodes of increased IOP in either or both eyes. These IOP spikes were always associated with the gonioscopic presence of blood in varying quadrants of the Schlemm canal. Whenever IOP was normalized in the affected eye(s), with or without medical therapy, the blood was no longer visualized. The patient also experienced 2 mild episodes of white-colored cells and flare noted in the anterior chamber, both resolving within a few days. At any of the visits, no signs of hyphema, either on slitlamp or gonioscopic examination, were noted.

A computed tomographic scan of the head (with contrast) showed that both superior ophthalmic veins were normal; no mass effect was noted. An open magnetic resonance imaging scan (with gadolinium enhancement) was also read as within normal limits; the cavernous sinus and ophthalmic veins were unremarkable. Automated visual fields (created with the 24-2 SITA program; Humphrey Instruments, Inc, Dublin, Calif) were normal in both eyes. Measurement of the episcleral venous pressure was within normal limits in both eyes. Doppler imaging of the jugular, subclavian, and upper extremity venous system showed normal values for compression and velocity. Extensive laboratory evaluation results (including a full hypercoagulable workup), as well as fluorescent treponemal antibody absorption test results, erythrocyte sedimentation rate, and beta human chorionic gonadotropin, rapid plasma reagin, antinuclear antibody, serum angiotensin-converting enzyme, and Lyme antibody titers, were all within normal limits. The chest x-ray film was also within normal limits. Of note, the patient was found to have a positive sickle cell trait on hemoglobin electrophoresis (HbAS trait; such individuals have 1 copy of the normal hemoglobin A gene and 1 copy of the hemoglobin S gene).

Comment. In our patient, the differential diagnosis included increased episcleral venous pressure due to Sturge-Weber syndrome, dural cavernous sinus fistula, carotid-cavernous sinus fistula, superior vena cava syndrome, orbital arteriovenous fistula, jugular vein obstruction, thyroid ophthalmopathy, and idiopathic familial entities. Other causes of blood in the Schlemm canal include artifact due to the compression of episcleral veins with the Goldmann or Allen-Thorpe gonio-lens, ocular hypotony (following injury, inflammation, or cycloidy-sis), and pathologic deep arterial anastomosis from a deep limbal artery to the Schlemm canal. During our extensive diagnostic evaluation (including normal values on episcleral venous pressure testing), these diagnoses were eventually excluded.

A systematic review of the literature demonstrated one previous case report of a patient with HbAS trait who was first seen because of a single episode of elevated IOP in one eye associated with blood in the Schlemm canal. However, our patient had multiple intermittent episodes of elevated IOP associated with transient episodes of blood in the Schlemm canal, which appeared in either eye. Another recent case report described a patient with a 3-year history of uveitis-glaucoma-hyphema syndrome who was diagnosed as having HbAS trait on the basis of cytopathologic examination findings of the submitted aqueous fluid during posterior-chamber intraocular-lens explantation and anterior-chamber washout.

While patients with sickle cell disease (either hemoglobin SS [HbSS] [individuals having 2 copies of the hemoglobin S gene] or HbSC [individuals having 1 copy of the hemoglobin S gene and 1 copy of the hemoglobin C gene]) are well known to have systemic and ocular vaso-occlusive episodes, little is known about the extent of increased coagulation activity in individuals with HbAS trait. Although HbAS trait disorders are usually asymp-tomatic in nature, we theorize that in our patient, the HbAS trait caused localized sickling of the red blood cells (with subsequent sludging) in the Schlemm canal. In support of this theory, a recent study reports that individuals with HbAS trait have increased coagulation activity (although it is lower than in patients with HbSC or HbSS disease). Moreover, Goldberg has reported that patients with sickle cell hemoglobinopathies (including AS) have a higher percentage of sickled erythrocytes in their anterior chambers than in their circulating venous blood. Injection of sickle cell erythrocytes (AS, SS, SC, and Sthal [consisting of 1 copy of the hemoglobin S gene and 1 copy of the hemoglobin β-thalassemia gene]) into the anterior chambers of living human, monkey, or guinea pig eyes results in sickling of the red blood cells with resultant IOP elevation. Furthermore, a vicious cycle of hypoxia and acidosis is known to contribute to increased sickling of the erythrocytes. In our patient, we suggest that intermittent episodes of sickled red blood cells in the trabecular meshwork (as a result of the HbAS trait) contributed to her transient elevations in IOP.

Robyn E. Horowitz, MD
Max Forbes, MD
Steven M. Podos, MD
James C. Tsai, MD

The authors have no relevant financial interest in this article.

This study was supported in part by the Homer McK. Rees Scholar Award, New York, NY (Dr Tsai); the Eye Surgery Fund (Dr Tsai), New York; and unrestricted departmental grants from Research to Prevent Blindness, Inc, New York.

This study was presented in part at the Annual Meeting of the Association for Research in Vision and Ophthalmology; May 8, 2002; Fort Lauderdale, Fla.

Correspondence: Dr Tsai, Harkness Eye Institute, Columbia University College of Physicians and Surgeons, 635 W 165th St, New York, NY 10032 (jct2002@columbia.edu).

Coats Disease and VATER Association in a 5-Year-Old Boy

Coats disease is an uncommon exudative retinopathy of unknown origin that may cause blindness. We report the unusual occurrence of Coats disease in a 5-year-old boy with multiple congenital abnormalities due to VATER association. The possibility of a genetic basis for some cases of Coats disease is discussed.

The VATER association comprises congenital defects including vertebral defects, imperforate anus, tracheoesophageal fistula, and radial and renal dysplasia. The underlying cause of VATER association may be a severe embryonic insult during the simultaneous development of the organ systems. Patients with VATER association often require multiple surgical procedures and extensive rehabilitation. We report a unique case of exudative retinopathy (Coats disease) in a case of VATER association.

Report of a Case. A child with a normal prenatal ultrasound at 20 weeks gestation and a negative TORCH (toxoplasmosis, other infections, rubella, cytomegalovirus infection, and herpes simplex) panel was born after an otherwise uncomplicated pregnancy by normal spontaneous vaginal delivery at 33 weeks gestational age. The results of a physical examination at birth revealed an imperforate anus and polydactyly with an extra digit lateral to the right thumb. The results of a radiological examination showed communicating hydrocephalus with grade II bilateral renal reflux and hemivertebra with severe scoliosis (Figure 1A). Corrective surgical procedures included digit amputation, colostomy, and anoplasty. There was no family history of eye or genetic defects, and karyotyping was normal in the child and his parents.

At 5 years of age, the patient was referred for left eye leukocoria. Visual acuity was 20/20 OD and 1/60 OS. There was total exudative retinal detachment with associated tortuous vessels with multiple aneurysmal dilatations compatible with Coats disease, stage 3B (Figure 1B). The lower 2 quadrants of the retina were preferentially affected. There was no evidence of associated anterior chamber or vitreal inflammation. The findings of the retinal examination of the fellow eye, including the periphery area, were normal. Further examination of old photographs showed the patient had had leukocoria since the age of 6 months. The results of B-scan ultrasonography revealed retinal detachment with no calcification or mass. This was confirmed by a computed tomography scan of the orbits. Intraocular pressure was normal with no evidence of neovascularization of the iris. Cryotherapy was offered to the patient to preserve vision and prevent further complications, including neovascular glaucoma, a painful blind eye, and a possible need for enucleation. The parents finally opted for conservative treatment.

Comment. A number of patients with VATER association have been described as having eye defects. The most common associations are coloboma and microphthalmos. However, the retina and posterior segment are seldom involved, and Coats disease was hitherto unreported. The term Coats disease refers to idiopathic retinal telangiectasia with intraretinal or subretinal exudation and without appreciable signs of retinal or vitreous traction. The diagnosis of Coats disease is one of exclusion after careful workup and examination, especially with regard to retinoblastoma, retinopathy of prematurity, retinal capillary angiomas, and toxocara, all of which were not implicated in our case. Reported associations include renal-retinal abnormalities and retinal disorders. Familial forms exist, but no specific gene defect is known. In the largest reported series, the median...