Severe Serous Macular Detachment in the setting of hypotony and Complex Hypercoagulability Syndrome

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Abstract
We report the occurrence and management of a massive serous detachment of the macula, which followed trabeculectomy and lowering of the intraocular pressure (IOP) in a patient with central retinal vein occlusion (CRVO) and a previously undiagnosed complex coagulopathy with elevated plasma fibrinogen and homocysteine levels, as well as prothrombin 20210 and factor V Leiden mutations. Our case illustrates prompt resolution of the serous detachment with elevation of the IOP and acute recurrence of the detachment following subsequent recurrence of hypotony after aqueous tube shunt surgery. Residual cystoid macular edema (CME) in the right eye, as well as hemiretinal vein occlusion with serous macular detachment and CME in the fellow left eye responded to bevacizumab. The occurrence of severe macular edema following lowering of intraocular pressure may warrant further evaluation for possible underlying venous occlusive disease or systemic coagulopathy.

Key words: macula, detachment, glaucoma, hypotony, coagulopathy.

Case report
A 26 year-old Caucasian male presented with a serous macular detachment in the setting of a prior central retinal vein occlusion (CRVO) of the right eye. His prior treatment history consisted of the following: he had previously received intravitreal triamcinolone (IVTA) for macular edema and developed steroid-induced glaucoma. His glaucoma was poorly controlled despite maximum tolerated medical therapy (latanaprost 0.05%, dorzolamide 2%-timolol 0.5% and brimonidine purite 0.15%) and he underwent trabeculectomy surgery. Postoperatively, his visual acuity declined from 20/40 to 20/200 and he underwent cataract extraction with intraocular lens implantation. After the cataract surgery, massive subretinal macular fluid developed and did not respond to additional IVTA. The patient then presented to us for a second opinion regarding further management.

Past medical history was significant for severe hypertension and end-stage renal disease secondary to glomerulonephritis. The patient had undergone a successful cadaveric renal transplant five years prior and was therefore on immunosuppressive medications including prednisone 20 mg/day, sirolimus (Rapamune, Wyeth) and mycophenolate mofetil (Cellcept, Roche). In addition he was on epoetin alpha (Procrit, Ortho biotech), omeprazole (Prilosec, Procter and Gamble), furosemide, hydralazine, metoprolol and minoxidil.

On initial presentation, his visual acuity was 20/800 in the right and 20/20 in the left eye. There was a functioning bleb in the right eye,
and intraocular pressures were 6 OD and 19 OS. Fundus examination showed a hyperemic optic nerve head, massive serous detachment of the macula, and dilated tortuous vessels, OD. (Figure 1A) The macular OCT (optical coherence tomography, Zeiss Stratus) scan showed the massive serous detachment (Figure 2A).

The patient underwent revision of the trabeculectomy with scleral flap closure, with placement of a Baerveldt glaucoma implant (AMO, Santa Ana CA), with a vicryl ligature to delay function. Postoperatively, the intraocular pressure rose to the high teens with improvement of VA to 20/400 and objective improvement in the macular edema. (Figure 1B)

The intraocular pressure gradually decreased, and with the onset of Baerveldt function, four weeks later, the patient developed sudden recurrent hypotony. The visual acuity decreased to hand motions. The anterior chamber was shallow and there was a recurrent massive macular detachment, which extended beyond the inferior arcade.

The patient underwent prompt Baerveldt revision to remove the tube from the anterior chamber and place it under the plate (conversion to stage 1) and close the tube entry site. Subsequently, the intraocular pressure rose to the mid-teens levels. The macular detachment resolved and visual acuity improved to 20/400. During follow up over the next 4 months the patient’s visual acuity and macular appearance continued to improve, with stabilization of the VA at 20/200. The macula OCT scan showed significant improvement in the macular detachment, with some residual cystoid edema remaining. (Figure 2B)

In June 2005, the patient developed a hemiretinal vein occlusion in the fellow left eye. Two weeks later, diffuse macular edema caused the visual acuity to decrease to 20/60. A hematologic work-up revealed elevated plasma fibrinogen and homocysteine levels, as well as prothrombin 20210 and factor V Leiden mutations. He was started on enoxaparin sodium (Lovenox, Sanofi Aventis) and coumadin. Over the following four weeks the VA in the left eye declined to 20/200, and in view of the complicated history of steroid response in the fellow eye, the patient was offered intravitreal bevacizumab injection in his left eye.

One week following intravitreal bevacizumab injection in his left eye, prompt resolution of macular edema occurred. VA
improved to 20/30 OS. The response was sustained for about 10 weeks until his VA dropped to 20/70 due to recurrent edema. He received a repeat injection with resolution of the edema and return of his vision to 20/30. One month after the initial injection in the left eye, the patient received an injection in his right eye, which was followed by resolution of the retinal edema, but without improvement in VA. Six weeks following the injection in the right eye, there was mild recurrence of retinal edema, but due to lack of improvement of VA in this eye despite the anatomic response to the injection, further injections were deferred.

**Discussion:**

Serous macular detachment following glaucoma surgery has rarely been reported as a manifestation of hypotony.\(^1\)\(^2\) Kokame and associates described moderate serous macular detachment in the setting of hypotony following Baerveldt implant in a patient with uveitis that resolved with reversal of the hypotony.\(^3\) Serous detachment in the setting of CRVO is a rare occurrence and is mostly associated with severe ischemia and poor visual outcome.\(^4\) More recently, 82% of patients with CRVO were found to have serous macular detachments that averaged 567 microns in height on OCT.\(^5\) In contrast, our patient developed localized massive serous detachment (>2000 microns in height) in the macular area of the right eye, without any evidence of ischemic changes, and only following trabeculectomy and lowering of the IOP. Moreover, the retinal detachment showed prompt resolution following elevation of the IOP, and reappeared with recurrence of the hypotony. We believe that mild hypotony, in the face of chronic underlying venous stasis secondary to the complex coagulopathy, caused an imbalance in the retinal interstitial fluid pressure that favored massive serous exudation.

There is an association between primary open angle glaucoma and ocular hypertension, as well as steroid induced elevation of IOP and retinal vein occlusion.\(^6\) With the increased popularity of intravitreal triamcinolone acetonide (IVTA) as a therapeutic modality for macular edema secondary to vascular occlusion,\(^6\) serious considerations must be given to the management of glaucoma in these patients. When surgery is performed to lower the IOP in patients with venous stasis, attention should be paid to the occurrence of persistent postoperative hypotony, which even if mild in nature, could precipitate this unusual complication of massive serous macular detachment. In young patients with CRVO, especially in the setting of underlying coagulopathy, or suspicion thereof, surgical intervention for glaucoma requires a carefully planned, graded or staged approach, which avoids sudden or severe lowering of the intraocular pressure, to prevent the occurrence of massive exudative detachments as in our patient. The use of bevacizumab as an alternative treatment approach for the macular edema in patients with vascular occlusions, who have a history of glaucoma or are known to have steroid response glaucoma,\(^7\) may avoid the need for glaucoma surgery in these patients.

The occurrence of massive serous macular detachment in the setting of hypotony should alert ophthalmologists to an underlying venous stasis and should prompt a systemic work-up for an underlying coagulopathy. Moreover, in patients with known steroid response glaucoma as in our patient, bevacizumab may offer an effective and safe alternative to IVTA for CME in retinal vascular occlusions.

Recognizing the role of hypotony in precipitating massive macular serous detachment in the setting of venous stasis, with immediate treatment directed to elevating the IOP, combined with the use of bevacizumab as an adjuvant, might offer a safe and effective option to prevent permanent vision loss in affected patients.

**REFERENCES**