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Eye (2011) **25,** 1512–1514; doi:10.1038/eye.2011.177; published online 5 August 2011

Sir,

Hemoglobin AC retinopathy managed with vitrectomy and adjunctive bevacizumab

Since the first report of hemoglobin (Hb) AC retinopathy,[1] few cases have been described and the pathogenesis remains obscure.[2] Significant intraoperative bleeding may make vitrectomy for proliferative retinopathy from sickling hemoglobinopathies challenging.[3]

Case report

A 33-year-old African American male, with idiopathic hypertension and mild renal failure, complained of decreased vision in his right eye for 6 months and floaters in his left eye for 2 years. Right and left visual acuities (VA) were 6/18 and 6/7.5, respectively. Anterior segments were normal. Fundoscopy showed vitreous hemorrhage (VH), right more than left; sclerotic retinal vessels, more so inferiorly (right more than left); and preretinal fibrosis with localized traction retinal detachments in the midperiphery of both eyes (Figures 1a and b). Fluorescein angiography showed neovascularization, capillary dropout and capillary remodeling (Figures 1c and d). Infective and autoimmune laboratory screen was negative. Hematological assessment revealed Hb AC (A = 56.9%, A2 = 2.8%, F = 0.7%, C = 39.6%), no sickling, and a normal blood count with an Hb level of 15.0 g/dl. Blood sugar was normal and creatinine was mildly raised to 1.5 mg/dl.

Scatter photocoagulation was placed in the anterior retina in both eyes. Three weeks later the left VH worsened (Figure 1e). One month later, VA remained at 6/60, due to nonclearing central VH, and possibly macular traction. After receiving 1.25 mg IVB 5 days before surgery, the VH and preretinal fibrosis were removed and the retina attached, with postvitrectomy VA stable at 6/7.5 (Figure 1f).

The right VH worsened 2 months after laser treatment. Because of nonclearing VH and tractional detachment threatening the macula, this eye was vitrectomized 4 months post laser with VA at 6/45. IVB of 1.25 mg was given 5 days before vitrectomy. The VA improved to 6/18, with persistent inferior macular ischemia (similar to presurgery; Figure 2).

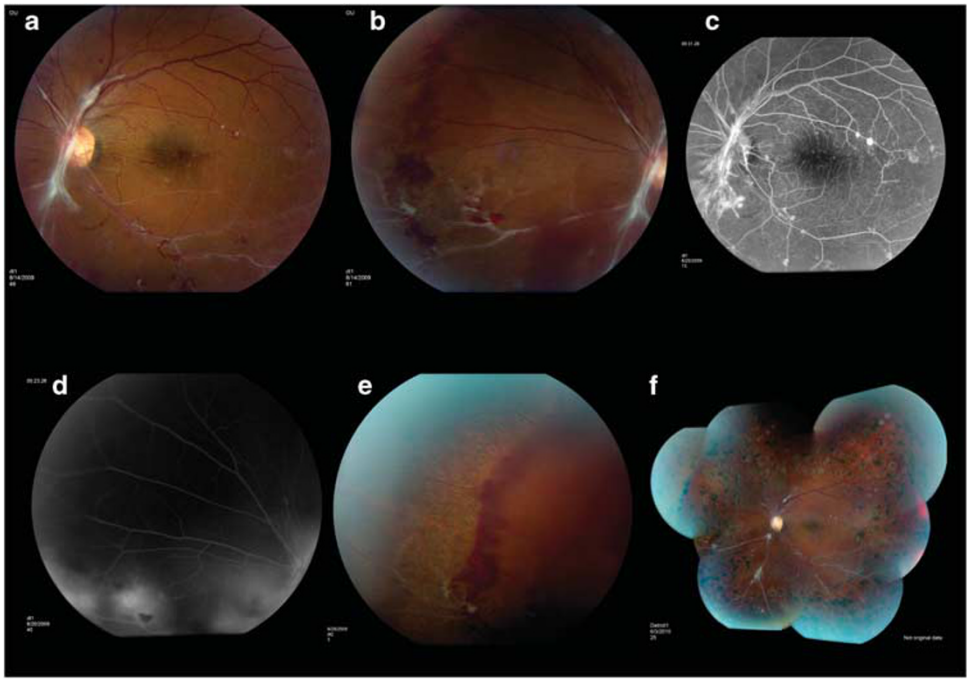

Figure 1 Left eye imaging. (a, b) Pretreatment color photographs show neovascularization, preretinal fibrosis and equatorial hemorrhage. (c, d) Pretreatment fluorescein angiography shows neovascular leakage from the preretinal fibrotic membranes, vascular remodeling of the posterior pole and capillary dropout at the midperiphery. (e) At 3 weeks after scatter laser therapy, color photograph shows a good laser pattern peripheral to preretinal hemorrhage of the superonasal equatorial region. (f) At 8 months post vitrectomy, the retina is fully attached with islands of segmented fibrosis, with visual acuity of 6/7.5.

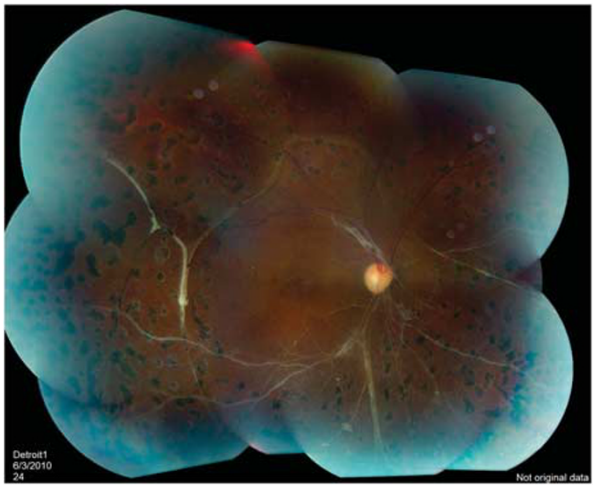

Figure 2 Right eye color photograph 4 months postvitrectomy. The retina is fully attached with islands of segmented preretinal fibrosis. Bands of subretinal fibrosis are seen temporal to the macula. Inferior macula and disk are ischemic and atrophic, as they were presurgery, with visual acuity of 6/18.

Comment

The present understanding of Hb C trait retinopathy is that it is extremely rare and, like Hb S trait retinopathy, only occurs with comorbidities.[4] We theorize that the increased blood viscosity (due to Hb AC), and the narrowed vessels (due to hypertension), acted synergistically to cause occlusive microangiopathy, which lead to proliferative neovascularization. Our case demonstrates how two non-neovasculogenic diseases, if coexisting, can cause sight-threatening retinopathy.

Because the preretinal fibrosis leaked fluorescein (Figures 1c and d), we used preoperative IVB to decrease bleeding when cutting these fibrovascular membranes. The right eye VA did not improve beyond 6/18 after surgery because of preexisting disease-related ischemic maculopathy. As reported by others,[5] we did not see IVB-related complications, such as worsening of the traction retinal detachment and macular ischemia.

Conflict of interest

The authors declare no conflict of interest.

Acknowledgements

Russell Pokroy received fellowship grants from the American Physicians Fellowship for Medicine in Israel, and from the Israel Ophthalmic Society.

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Eye (2011) **25,** 1514–1515; doi:10.1038/eye.2011.189; published online 5 August 2011

Sir,

Nerve fiber layer irregularity after internal limiting membrane peeling, seen by spectral domain optical coherence tomography

Tadayoni *et al*[1] reported that the retina acquired a particular appearance, featuring 'arcuate striae', after pars plana vitrectomy (PPV) for treatment of a macular hole. They considered that the feature was attributable to the presence of a dissociated optic nerve fiber layer (DONFL). Ito *et al*[2] found that a DONFL resulted in the development of numerous arcuate retinal striae mimicking a retinal nerve fiber layer (RNFL) defect. Earlier conventional time-domain (TD) OCT showed only focal dimples on two-dimensional imaging.

We performed a PPV with internal limiting membrane (ILM) peeling to treat macular hole. No frank retinal trauma was observed during PPV or the ILM peeling procedure. Preoperative fundus photograph showed no RNFL defect (Figure 1). On 12-months postoperative review, multiple dark round lesions were detected in the superior-temporal perimacular area. We performed high-resolution spectral-domain (SD) OCT with three-dimensional imaging (Cirrus OCT, Carl Zeiss Meditec, Inc., Dublin, CA, USA). SD OCT showed deep focal dimples, with clear margins, in the perimacular area, compatible with the presence of a DONFL. Three-dimensional SD OCT images, which reveal the inner retinal surface, confirmed the presence of multiple round, cobblestone-shaped, 'beaten bronze' dimples in the superior temporal perimacular area. DONFL did not present as the arcuate striae described in many previous reports.[2–4] Many reports have described RNFL morphological changes after ILM peeling during vitrectomy with TD OCT.[1–4] Removal of the ILM is a common procedure in macular hole surgery, as it increases the probability that the macular hole will close. In our case, we used SD OCT, which provides images of greater clarity and higher resolution than afforded by TD OCT. SD OCT clearly revealed the dimple margins and the exact depth of each DONFL. The shape and size of a DONFL could be visualized using the new three-dimensional SD OCT imaging facility. Our patient had