Aged-related macular degeneration (AMD) remains the leading cause of visual loss in adults older than 65 years. Retinal angiomatous proliferation (RAP) is a newly recognized manifestation of exudative AMD that has a poor natural history and demonstrated resistance to treatment with conventional laser photocoagulation. A novel surgical technique has been developed in this pilot series that shows promise in the treatment of this subtype of neovascular lesions. Through specific surgical lysis of the feeding arteriole and draining venule of an RAP lesion, improvement in visual acuity has been noted. This has been correlated with resolution of intraretinal edema and flattening of associated pigment epithelial detachment, which was confirmed by fluorescein angiography, optical coherence tomography, and high-speed indocyanine green angiography.

Retinal angiomatous proliferation (RAP) has recently been described in a subset of patients with exudative age-related macular degeneration (AMD).1 In RAP, the neovascular process originates from the retinal vasculature as opposed to the underlying choriocapillaris, as is seen with typical choroidal neovascularization. On clinical examination, the presence of intraretinal or preretinal hemorrhages associated with retinal vessels that abruptly curve toward the choroid is consistent with RAP. Cystoid macular edema, lipid exudates, and pigment epithelial detachment (PED) may be present.

A classification system has been reported to further subdivide these lesions. Briefly, RAP stage 1 involves the proliferation of abnormal vessels in the intraretinal space, akin to the intraretinal microvascular abnormalities seen in diabetes mellitus. This intraretinal neovascularization often demonstrates retinal-retinal anastomoses and may continue to proliferate and leak. Stage 2 is defined by the presence of intraretinal edema and may be associated with serous PED and/or subretinal neovascularization. In stage 3, retinochoroidal anastomoses are present; associated fibrovascular PED is a late finding and is also seen in RAP stage 3. On fluorescein angiography, RAP appears as purely occult or minimally classic choroidal neovascularization. Standard indocyanine green (ICG) angiography reveals a hot spot in late frames. High-speed ICG angiography allows an accurate identification of the RAP lesion, retinal feeding arterioles, draining venules, and associated retinal-retinal or retinochoroidal anastomoses.1

Unfortunately, patients with RAP have poor visual outcomes after treatment with conventional laser photocoagulation.2-4 Affected individuals often demonstrate a subacute decrease in visual acuity over the course of several months. Because of the inefficacy of laser photocoagulation and the poor natural history of RAP, alternative treatment options warrant further investigation. Patients with RAP stage 1 may be asymptomatic and remain undetected. Patients with RAP stage 3 often already have a disciform scar with irreversible overlying photoreceptor dysfunction. For these reasons, attention is focused on RAP stage 2. In a small series of patients, we describe a new surgical technique for the ablation of these angiomatous lesions.

From the Retina Service, Wills Eye Hospital, Philadelphia, Pa. Dr Borrillo is now with Northern California Retina Vitreous Associates Medical Group, Mountain View. The authors have no relevant financial interest in this article.
Figure 1. A, A preoperative fluorescein angiogram of patient 2 demonstrates foveal leakage in a petaloid pattern consistent with cystoid macular edema. Note the pooled fluorescence centered temporally in the form of a serous pigment epithelial detachment (PED). B, A fluorescein angiogram of patient 2 taken 1 month postoperatively shows an absence of foveal leakage consistent with resolution of cystoid macular edema, with corresponding collapse of the PED.

Figure 2. A, A high-speed indocyanine green angiogram of patient 2 demonstrates a hypercyanescent retinal angiomatous proliferation (RAP) lesion, with a retinal feeder arteriole (short arrow) and draining venules (long arrows). The hypocyanescent area is consistent with serous pigment epithelial detachment (PED). B, A high-speed indocyanine green angiogram of patient 2 taken 1 month postoperatively demonstrates absence of perfusion of the RAP lesion through retinal feeder vessels and resolution of PED. Note the improved visualization of the choroidal vasculature underlying the original lesion site.

Figure 3. A, Intraocular scissors are poised to incise the retinal feeder arteriole and the draining venule (each lysis is performed separately). Intraretinal edema and serous pigment epithelial detachment (PED) are present. B, After lysis of the retinal feeder vessels, resolution of intraretinal edema and flattening of serous PED are noted after several months. Modified with permission from IRIDEX Corp, Mountain View.
METHODS

Four eyes of 3 consecutive patients were identified from the clinical practice of the vitreoretinal surgeon (A.S.) between May and November 2001. All patients underwent clinical examination, fluorescein angiography (Figure 1), ICG angiography, optical coherence tomography, and high-speed ICG video angiography (Figure 2). Patients were included in the study if they had exudative AMD with RAP stage 2 and associated serous PED, as determined by clinical and diagnosti examination. Snellen visual acuities were obtained preoperatively, 4 to 6 weeks postoperatively, and at least 6 months after surgical intervention.

Patients underwent surgical ablation of RAP retinal feeder vessels at the Wills Eye Hospital, Philadelphia, Pa, and were followed up during a 6-month period. The RAP lesions and their corresponding retinal feeder arterioles and draining venules were identified on still frames from the high-speed ICG angiogram. A tracing was made to serve as a guide for surgical lysis. Lower-order vessels were chosen for ablation to avoid significant inner retinal ischemia. After a standard 3-port pars plana vitrectomy was performed, the posterior hyaloid face was detached and trimmed to the far periphery under wide-field viewing. The high-speed ICG angiogram tracing was used to identify the specific retinal feeder vessels targeted for excision. A partial thickness retinotomy with a 22-gauge needle was made superficial to the vessels; this retinotomy was placed at least 0.5 disc diameters away from the fovea. The RAP feeder vessels were then incised with intraocular vertical scissors (Synergetics, St Charles, Mo), with the arteriole lysed prior to the venule (Figure 3). The infusion pressure was elevated temporarily to ensure hemostasis, and endodiathermy was applied to prevent recanalization.

RESULTS

Patient data are summarized in the Table. The mean age of the patients was 82 years. All patients were female and had a history of hypertension controlled with medication. Preoperatively, the mean initial Snellen visual acuity was 20/200 as determined by conversion to the logarithm of the minimal angle of resolution. Postoperatively, the mean final visual acuity improved to 20/70 at a follow-up interval of 6 to 12 months. Macular thickening resolved in all eyes that underwent RAP ablation (Figure 4), with collapse of associated PED. No eyes developed surgical complications, such as endophthalmitis or retinal detachment, in the postoperative period.

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Eye</th>
<th>Age, y</th>
<th>Initial</th>
<th>6-8 Weeks Postoperatively</th>
<th>Final</th>
<th>Follow-up, mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>OD</td>
<td>89</td>
<td>20/200</td>
<td>20/40</td>
<td>20/40</td>
<td>12</td>
</tr>
<tr>
<td>2</td>
<td>OS</td>
<td>89</td>
<td>20/400</td>
<td>20/80</td>
<td>20/200</td>
<td>12</td>
</tr>
<tr>
<td>3</td>
<td>OD</td>
<td>82</td>
<td>20/50</td>
<td>20/80</td>
<td>20/60</td>
<td>6</td>
</tr>
<tr>
<td>4</td>
<td>OD</td>
<td>67</td>
<td>20/400</td>
<td>20/100</td>
<td>20/30</td>
<td>8</td>
</tr>
</tbody>
</table>

Figure 4. A, A preoperative optical coherence tomograph of patient 3 demonstrates a decreased intraretinal signal in cystoid spaces consistent with cystoid macular edema, overlying a serous elevation of the retinal pigment epithelium (RPE)–choriocapillaris layer consistent with serous pigment epithelial detachment (PED). B, An optical coherence tomograph of patient 3 taken 3 months postoperatively demonstrates resolution of both intraretinal edema and serous PED, with restoration of a normal foveal contour; there is some residual parafoveal thickening of the RPE-choriocapillaris layer, with a normalized contour, as well.

COMMENT

Retinal angiomatous proliferation is a distinct clinical entity in the spectrum of patients with exudative AMD. The presence of an RAP lesion is suggested by preretinal or intraretinal hemorrhage at the point where the macular edema is thickest with or without associated PED. To date, there are no effective treatments for the subset of AMD patients with RAP. We present a novel surgical approach that may benefit patients with this manifestation.

Because RAP lesions, by definition, originate from the retinal vasculature, the focus has been on surgical lysis of the retinal feeding arterioles and draining venules associated with angiomatous proliferation. Patients with RAP stage 2 appear to be the best candidates for this procedure. In this small series of patients who underwent surgical ablation of an RAP lesion, improvement in clinical, angiographic, and tomographic appearance was noted in all cases. Upon successful lysis of the feeding and draining vessels, complete resolution of retinal fluid and PED was generally noted within 7 to 10 days. This translates into a trend toward corresponding improvement in visual acuity. In the current series, the lesions remained closed without recurrence of the RAP.

One theoretical advantage of this surgical approach is that it spares the retinal pigment epithelium (RPE), unlike laser photocoagulation. The reduction of macular thickening with preservation of the underlying RPE may allow for better functioning of the overlying outer
neurosensory retina. In contrast, it can be argued that localized inner retinal ischemia from retinal vascular ablation may prevent optimal functioning in the distal retina supplied by the ablated vessels. For this reason, lower-order vessels are chosen for ablation to avoid significant inner retinal ischemia. Also, by sparing the RPE, there may be a decreased incidence of typical choroidal neovascularization, which often manifests after conventional laser therapy. Finally, surgical ablation may induce a more permanent closure of feeder vessels with a trend toward decreased recanalization.

Patients with advanced RAP stage 3 may not be good surgical candidates because these lesions have developed anastomoses with the choroidal circulation. The high flow rates through such connections and the depth of vascular ingrowth would likely make this subtype of RAP lesions resistant to surgical lysis.

We acknowledge that the lack of a control group, relatively short follow-up period, and small number of patients are inherent flaws of this study. Nonetheless, surgical lysis of RAP stage 2 lesions appears to be a promising approach to a condition that currently has a poor natural history and no effective treatment. No complications were encountered in this small-group study. A large controlled, prospective trial is needed to ultimately determine the efficacy and safety of surgical RAP ablation.

Submitted for publication July 26, 2002; final revision received December 2, 2002; accepted December 26, 2002.

Call for Papers

AMA and the ARCHIVES journals will be participating in the 2003 global theme issue, “Pain Management.” We invite all investigators to submit their best research addressing issues concerning pain management in ophthalmology for consideration for the November 2003 issue. Manuscripts received by May 1, 2003, have the best chance for acceptance for the global theme issue. Manuscripts not accepted for the theme issue may be considered for other issues of the ARCHIVES. All manuscripts are subject to our usual peer review. Please note in the cover letter that the submission is for the November 2003 “Pain Management” issue.

REFERENCES


Corresponding author: Arunan Sivalingam, MD, Retina Service, Wills Eye Hospital, 840 Walnut St, 10th Floor, Philadelphia, PA 19107 (e-mail: asivalinga@aol.com).