Loss of Reading and Central Vision Due to Macular Diseases - Therapeutic Management, Advances and Limitations

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Remarkable advances have recently developed in the management of macular diseases. There are now treatment options for diseases previously felt to be incurable, and for recovery of central vision for some of these patients. The macula is the central part of the retina (the nerve tissue lining the back of the eye). The macula provides for the fine and discriminating vision, needed for driving, reading and facial recognition. Thus, macular diseases result in loss of some of the most important visual tasks, ie. - the ability to read, the ability to drive, and the ability to recognize others. Vitreoretinal surgery is a rapidly changing subspecialty of ophthalmology, which has recently developed new approaches to macular diseases, such as macular holes, macular degeneration, and subretinal neovascularization. Research in Hawaii has contributed to these advances.

Macular Holes

Age-related macular holes are holes in the central part of the retina, resulting in a missing spot in the center of vision and in distortion of vision (metamorphopsia) (Fig. 1). Macular holes are more common in females and increase in frequency with age, especially after age 55. Prevalence has been estimated to be 3.3 per 1000 in people over 55. This disease was previously felt to be untreatable until 1991, when the first pilot series of vitreous surgery showed closure of the macular hole, sealing of the retinal separation and improvement of central vision (Fig. 2).[1] Marked improvements in surgical techniques have resulted in a significant increase in success rate from 58% in the initial reported series to over 90% in recent series, including series reported from the Retina Center at Pali Momi. To take a disease previously untreatable six years ago to a point of highly successful surgical management is truly a remarkable story in the field of vitreoretinal surgery.

The Retina Center at Pali Momi, (a division of Kapiolani Medical Center at Pali Momi), is one of 15 study centers around the nation participating in the randomized national multi-centered trials on macular hole surgery (The Vitrectomy for Prevention of Macular Hole Study[2,3] and The Vitrectomy for Macular Hole Study).[4-6] These studies have resulted in significant contributions to the literature and represent the only well-controlled randomized studies on macular hole surgery. The Retina Center at Pali Momi is studying the use of serum growth factors to stimulate higher macular hole closure rate and vitrectomy with an intraocular gas bubble to prevent macular hole development in early stages of this disease.[3] In previously published studies ultrasonographic techniques were utilized to demonstrate tangential traction within the vitreous gel causing macular holes[7], and macular hole surgery was shown to allow patients even with chronic macular holes of many years in duration to recover useful central vision following surgery.[8]

Age-Related Macular Degeneration

Age-related macular degeneration (AMD) is a degenerative disease of the central retina, which results in a gradual scarring of support tissues. AMD is the most common causes of reportable blindness in the United States, as well as here in Hawaii. It is most common in Caucasians, especially fair and blue-eyed individuals, and less common, although not infrequently seen in Asians. Presently, preventative treatment is not proven, but studies on mineral, antioxidant, and vitamins supplements are ongoing. Laser treatment in the early stages of the disease is being studied in randomized trials.

AMD has two categories - the dry form and the wet form. The mainstay of treatment is laser surgery for the wet form which is the most common cause of severe vision loss. In the dry form there is gradual scarring with slow and gradual vision loss. In the wet form, choroidal neovascular membranes (CNVM) rapidly proliferate causing bleeding, scarring and leaking and rapid vision loss (Fig. 3). Laser treatment to obliterate the CNVM has been shown in large national multi-centered trials to decrease the risk of further vision loss.[10-11] A fluorescein angiogram (office procedure) is used to visualize the extent of CNVM (Fig. 3). Fluorescein dye is injected into an arm vein and an ophthalmic fundus camera with specific filters for the fluorescent light visualizes the dye in the retinal vessels.

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Fig 1.—Photograph of macular hole in left eye. Note visible hole approximately 800 um in size in center of macula. Visual acuity is 20/200.

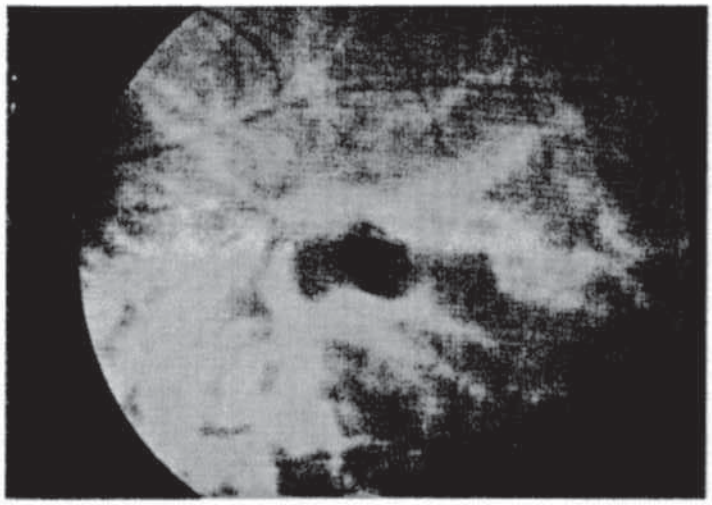


Fig. 2.—Postoperative photograph following macular hole surgery. Note that macular hole has sealed. Visual acuity is 20/40.

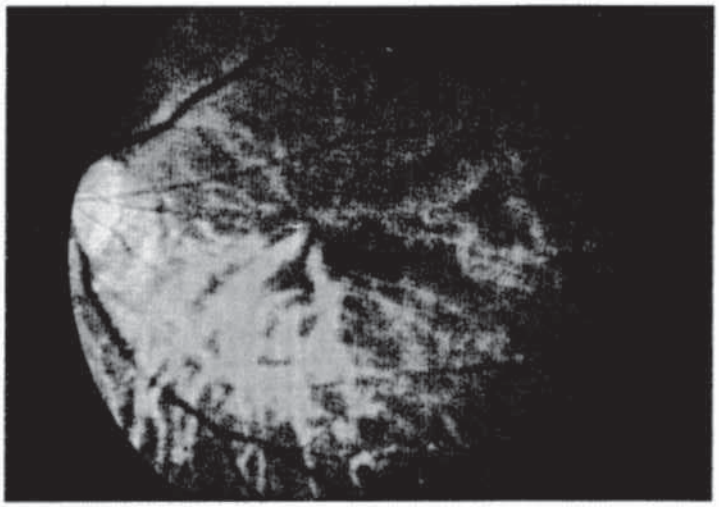


Fig. 3.—Fluorescein angiogram of retinal pigment epithelial detachment (RPED) or blister-like formation in pigment epithelium due to subretinal neovascularization and AMD. Note bright spot at superonasal edge of blister consistent with SRNVM, but partially obscured by fluorescence within the RPED.

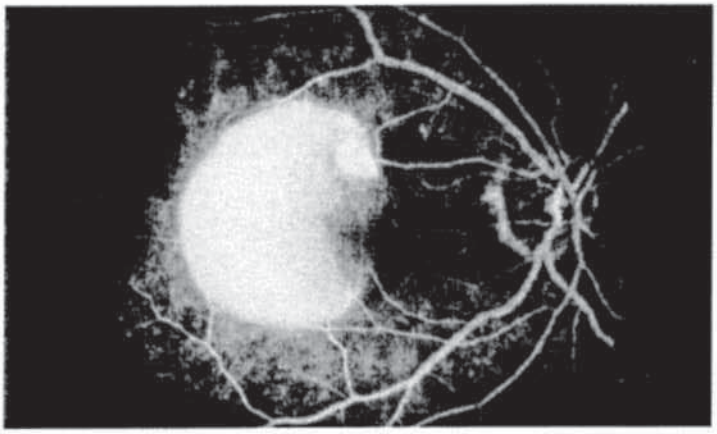


Fig. 4.—ICG angiogram demonstrating bright, focal area of hyperfluorescence consistent with SRNVM in same eye as in Figure 3. Note absence of fluorescence obscuring SRNVM within the RPED.

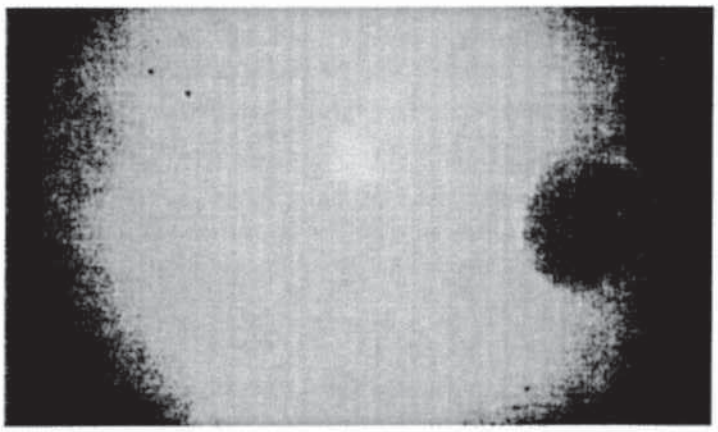


Fig. 5.—Photograph of SRNVM in patient with idiopathic inflammatory disease of retina, called punctate inner choroidopathy. Note the grey subretinal lesion extending into central fovea, as well as serous retinal detachment. Visual acuity is 20/200.

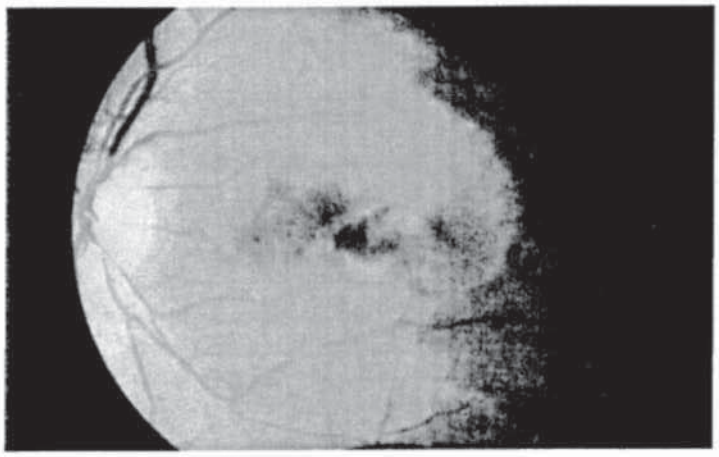
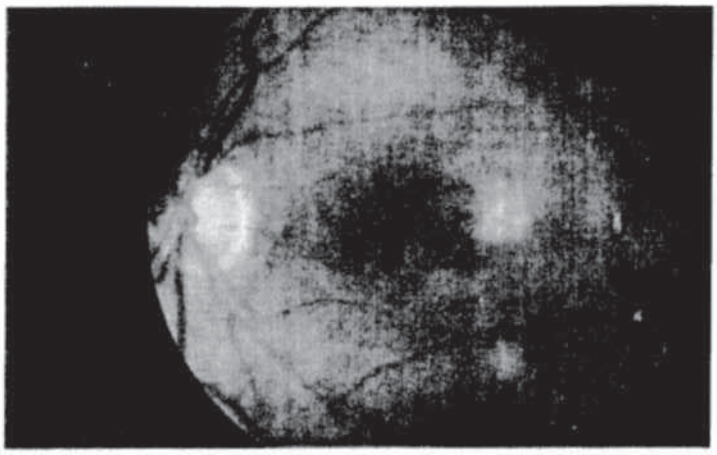


Fig. 6.—Postoperative photograph following subretinal surgery to remove SRNVM showing focal hypopigmented scar in temporal macula. Note absence of grey membrane and serous retinal detachment. Visual acuity has improved to 20/30.



The images are captured on film or on digital camera with computer interface, and laser treatment can be guided to the area of CNVM. Unfortunately, the percentage of patients with the wet form treatable with fluorescein angiographic criteria is less than 15%. A new imaging technology utilizing indocyanine green (ICG) may allow more precise localization of CNVM in select cases (Fig. 4). This imaging technology uses digital imaging, because of the higher sensitivity of digital cameras or video cameras to the fluorescence of the ICG dye in the infrared range. This imaging technique has become available in Hawaii for just over a year. With further experience and research with this modality, more patients may become eligible for laser surgery, although we have often found the two imaging modalities are complimentary. For patients not treatable with laser surgery, the prognosis for central vision is poor, although the majority of patients will have normal peripheral vision. Ongoing studies are evaluating medical treatments of CNVM by anti-angiogenic modalities, such as oral thalidomide, vascular endothelial growth factor antibodies, and radiation therapy.

Choroidal neovascular membrances (CNVM) are abnormal blood vessels growing into the subretinal space through retinal pigment epithelium. The most common disease is AMD, but there are other entities, which cause retinal scars and CNVM ingrowth. These include ocular histoplasmosis syndrome (a scarring disease associated with the fungus, Histoplasma capsulatum, common in the midwest), high myopia, angioid streaks (associated most commonly with pseudoxanthoma elasticum), trauma with choroidal ruptures, and multiple viral or idiopathic inflammatory diseases of the retina. Because CNVM is subretinal under the nerve tissue thus limiting surgical access, surgical removal was not considered until the first successful surgical series of CNVM removal through a small retinotomy in 1991.[12] Subretinal surgery has since develeped remarkably with improved techniques and instrumentation. Most promising visual results have been in patients with ocular histoplasmosis or idiopathic causes (Figs 5-6).[13] The initial results of surgical removal of CNVM in AMD were dissappointing, but research is ongoing, including exciting possible new treatment avenues of neuroretinal and retinal pigment epithelial cell transplantation. Subretinal surgery to remove hemorrhage using tissue plasminogen activator has successfully restored vision in subretinal hemorrhage due to retinal macroaneurysms and AMD.

Macular diseases cause loss of some of the most important visual abilities. Recent diagnostic, therapeutic and surgical advances have resulted in recovery of central vision for many patients with these diseases. Ongoing research has led to advances in this rapidly changing field, much occurring within this decade.

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