

FIGURE 1. (Top) Fundus photograph of the right eye shows disk edema, drusen, and subretinal blood before laser treatment. (Bottom) Angiogram of the right eye, same date as top.

ization and peripapillary hemorrhage,^{3,4} but none had optic disk drusen. It is hypothesized that the cause of the disk edema may be infiltration of the optic nerve or nerve sheath with lymphoid cells, or autoantibodies produced by plasma cells or lymphoid cells may deposit within the nerves of the body, thus, inducing a neuropathy and optic disk edema. Patients with POEMS syndrome may have increased serum levels of vascular endothelial growth factor. Vascular endothelial growth factor may play a role in the development of choroidal neovascularization.

Peripapillary choroidal neovascularization has been described in association with optic disk drusen.⁵ The origin is not completely understood. Enlarging optic nerve drusen may impair the circulation to the peripapillary retina. The consequent retinal ischemia may then stimulate the growth of new vessels beneath the pigment epithelium. Alternatively, enlargement or anterior migration of optic nerve drusen may allow the ingrowth of vessels through this discontinuity, disrupting the integrity of the Bruch membrane.

The most common ocular finding in patients with

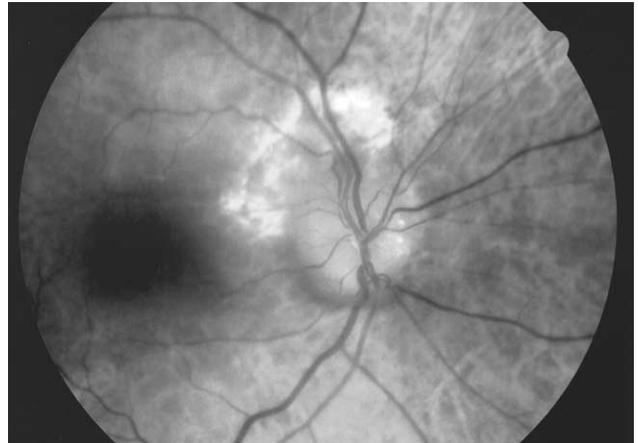


FIGURE 2. Fundus photograph 4 months after laser treatment in patient's right eye; choroidal neovascularization has resolved.

POEMS syndrome is optic disk edema. The possibility of optic disk drusen and peripapillary choroidal neovascularization should be considered in patients with vision loss and POEMS syndrome.

REFERENCES

1. Bardwick PA, Zvaifer NJ, Gill GN, Newman D, Greenway GD, Resnick DL. Plasma cell dyscrasia with polyneuropathy, organomegaly, endocrinopathy, m-protein, and skin changes. The POEMS syndrome. *Medicine* 1980;59:311.
2. Bolling JP, Brazis PW. Optic disk swelling with peripheral neuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS Syndrome). *Am J Ophthalmol* 1990;109:503-510.
3. Brazis PW, Liesegang TJ, Bolling JP. When optic disc edema and peripheral neuropathy constitute poetry? *Surv Ophthalmol* 1990;35:219-225.
4. Arnold PD, Kinyoun JL, Guzak SV. Poems syndrome: an unusual cause of neovascularization. *Retina* 1999;19:166-168.
5. Harris MJ, Fine SL, Owens SL. Hemorrhagic complications of optic nerve drusen. *Am J Ophthalmol* 1981;92:70-76.

Management Options for Early Stages of Acutely Symptomatic Macular Holes

Gregg T. Kokame, MD

Accepted for publication Aug 29, 2001.

From the Division of Ophthalmology, Department of Surgery, University of Hawaii School of Medicine, Honolulu, Hawaii; The Retina Center at Pali Momi, Kapi'olani Health, Aiea, Hawaii. Presented at the Annual Meeting of the Macula Society (3/2/01) in Scottsdale, Arizona.

This study was supported by the Clinical Research Center of the Kapi'olani Research Institute and by a Research Centers in Minority Institutions Award, P2ORR/A111091, from the National Center for Research Resources, National Institutes of Health.

Inquiries to Gregg T. Kokame, MD, The Retina Center at Pali Momi, 98-1079 Moanalua Road, Suite 470, Aiea, Hawaii 96701; fax: (808) 487-3699; e-mail: retinahi@aol.com

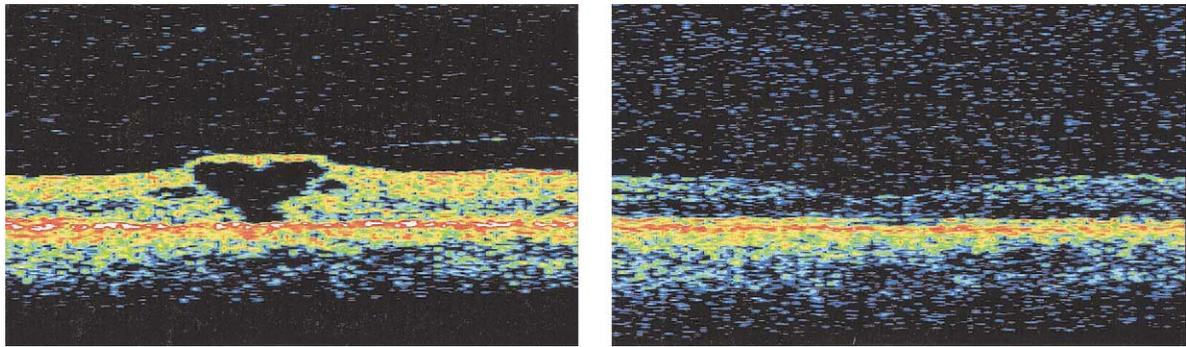


FIGURE 1. (Left) Optical coherence tomography of advanced stage I macular hole with poor vision of 20/70 showing tangential traction on the fovea with a pseudocyst, a hole in the outer layer or photoreceptors, and an intact inner layer. (Right) Postoperative optical coherence tomography of the same eye after vitrectomy, posterior cortical vitreous separation and fluid–sulfur hexafluoride (SF₆) gas exchange shows resolution of the pseudocyst, closure of the outer layer hole, and a foveal depression.

PURPOSE: To evaluate management options for acutely symptomatic stage I macular holes, and vitrectomy with intraocular gas tamponade for advanced stage I macular holes.

METHODS: Retrospective noncomparative study. **Setting:** Clinical practice. **Study population:** Acutely symptomatic stage I macular holes. **Management options:** (I) spontaneous resolution with observation –7 eyes; (II) stage I holes developing poor visual acuity of 20/50 or worse during observation –9 eyes undergoing pars plana vitrectomy, posterior vitreous separation, fluid –20% sulfur hexafluoride gas exchange; (III) stage I holes acutely progressing to stage II or III during observation –9 eyes with vitrectomy, posterior vitreous separation, membrane peel, fluid –18% perfluoropropane gas exchange. **Main Outcome Measures:** visual acuity of 20/40 or better, prevention, or closure of macular hole.

RESULTS: Spontaneous resolution developed in 7 eyes with 20/40 or better vision (group I). 8/9 eyes undergoing vitrectomy for advanced stage I holes (group II) did not progress and recovered 20/40 or better vision. 1/9 recovered 20/40 vision after further surgery. 9/9 eyes following acute progression to full-thickness holes (group III) had closed macular holes with recovery of 20/40 vision after vitrectomy.

CONCLUSIONS: Stage I macular holes can initially be observed. However, excellent visual and surgical results can be obtained in stage I holes with poor vision, or with acute progression to full-thickness holes. (Am J Ophthalmol 2002;133:276–278. © 2002 by Elsevier Science Inc. All rights reserved.)

THE INITIAL INTEREST IN SURGICAL MANAGEMENT FOR idiopathic macular holes was in the prevention of macular hole development by vitrectomy.¹ Management in the acute stages of macular hole was initially theorized to have the best potential for prevention of visual loss.² The Vitrectomy for Prevention of Macular Hole Study did not

show a significant benefit to vitrectomy without intraocular gas tamponade in the early stages of macular hole development.¹ Furthermore, full thickness macular hole surgery resulted in recovery of vision, which has remarkably improved since the initial study by Kelly and Wendel.³

The risk of progression of stage I macular hole increases as best corrected visual acuity worsens with a 66% progression with 20/50 or worse vision.⁴ Stage II macular holes have a similar progression rate of 71%.⁵ These similar progression rates are explained by optical coherence tomography of advanced stage I macular hole, which shows tangential traction on the fovea and a foveal pseudocyst (Figure 1, left). The decrease in visual acuity in advanced stage I macular hole is explained by a hole in the outer photoreceptor layer, which develops often before the development of a visible break in the inner layer (stage II or III hole). Because of similar risks of progression, it is reasonable to study intervention in both acutely symptomatic stage I holes with poor vision, and stage I holes acutely progressing to full-thickness holes.

Retrospectively, patients with acutely symptomatic stage I macular holes presenting to The Retina Center at Pali Momi, a division of Kapi'olani Health, were reviewed over a 10-year period from 1990 to 2000. All patients gave consent that medical records and photographs could be utilized for research purposes, and understood that they would not be identified individually. The study was reviewed by the Kapi'olani Research Institute. The purpose of this study is to review the results of management options for stage I macular holes, and to evaluate the role of vitrectomy with intraocular gas tamponade for stage I macular holes with poor vision.

All 25 patients presented with stage I macular holes as defined in the original Gass classification² with a yellow spot (stage Ia) or yellow ring (stage Ib), but without a visible full-thickness break biomicroscopically. All patients were acutely symptomatic, defined as symptoms of

three months or less with focal central distortion noted on the Amsler grid. All patients did not see a break in a thin slit beam placed across the lesion (negative Watzke-Allen sign) on initial evaluation. Patients underwent a complete ophthalmologic examination, best corrected Snellen visual acuity, fundus photographs, and fluorescein angiography. Exclusion factors included high myopia over -6.5 diopters, myopic macular degeneration, diabetic retinopathy, or age related macular degeneration. Most stage I holes were initially observed, but then managed in three groups: (I) observation only, if spontaneous resolution developed -7 eyes; (II) pars plana vitrectomy, posterior cortical vitreous separation, and fluid -20% sulfur hexafluoride gas exchange if vision worsened to 20/50 or worse with a stage I hole configuration -9 eyes; (III) pars plana vitrectomy, posterior cortical vitreous peeling, membrane peeling without internal limiting membrane peeling, fluid -18% perfluoropropane gas exchange for acute full-thickness hole (stage II or III) -9 eyes. All patients had at least six months and up to nine years of follow-up.

During observation (group I) spontaneous resolution developed in 7 eyes with all maintaining 20/40 or better vision. Presenting vision in group I was 20/40 or better in 5 eyes. Two patients with presenting vision of 20/60 to 20/70 elected to observe and spontaneously resolved. Spontaneously resolved holes often showed a focal foveal facet, similar to postoperative cases. In group II following vitrectomy and fluid-gas exchange with sulfur hexafluoride, 8 of 9 eyes did not progress and recovered 20/40 or better vision (Figure 1, right). One of 9 eyes required a subsequent surgery with perfluoropropane after macular hole progression, but recovered 20/40 vision with hole closure. In 9 eyes which acutely progressed from stage I to a full-thickness hole (group III), pars plana vitrectomy, posterior cortical vitreous separation, membrane peeling, and fluid-gas exchange with perfluoropropane resulted in hole closure in all eyes, and recovery of 20/40 or better vision. Of the 9 eyes in group II, one was previously pseudophakic, two developed moderate cataracts, and six underwent cataract surgery. Of the 9 eyes in group III, two were previously pseudophakic, one had a mild cataract, and six underwent cataract surgery. One of 18 operated eyes (5%), an eye in group II, developed retinal detachment with reopening of the macular hole, but recovered 20/20 vision with surgical repair.

Resolution of macular hole in the acute stages of development allowed excellent anatomic and visual results in all three groups. All 25 patients recovered 20/40 or better vision at last follow-up. Stage I holes with poor vision (group II) progressed in 12% (1/9 eyes) following vitrectomy with gas tamponade, which compares favorably with the natural history of stage I macular holes with vision of 20/50 or worse (66% progression rate).⁴ This also compares favorably with the vitrectomy group in the Vitrectomy for Prevention of Macular Hole Study (37% progression rate),¹ which did not include intraocular gas

tamponade and whose progression rate may have been even higher if limited to high risk eyes with poor vision.⁴ However, this present study is a retrospective and nonrandomized study with small numbers in each group. In addition, the risks of post-vitrectomy cataract formation and occasional retinal detachment must be weighed against the potential benefit of better vision potential with earlier intervention. Observation was recommended initially in most cases, and is especially recommended for stage I holes with best corrected visual acuity of 20/40 or better due to the low progression rate.⁴ A randomized clinical trial studying vitrectomy with intraocular gas tamponade in stage I holes with poor vision versus intervention only after full-thickness hole development is indicated and may be aided by the use of optical coherence tomography for patient selection.

REFERENCES

1. De Bustros S. The Vitrectomy for Prevention of Macular Hole Study Group. Vitrectomy for prevention of macular holes—Results of a randomized multicenter clinical trial. *Ophthalmology* 1994;101:1055–1059.
2. Gass JD. Idiopathic senile macular hole: its early stages and pathogenesis. *Arch Ophthalmol* 1988;106:629–639.
3. Kelly NE, Wendel RT. Vitreous surgery for idiopathic macular holes: results of a pilot study. *Arch Ophthalmol* 1991;109:654–659.
4. Kokame GT, de Bustros S. The Vitrectomy for Prevention of Macular Hole Study Group. Visual acuity as a prognostic indicator in stage I macular holes. *Am J Ophthalmol* 1995;120:112–114.
5. Kim JW, Freeman WR, Azen SP, El-Haig W, Klein DJ, Bailey IL. The Vitrectomy for Macular Hole Study Group. Prospective randomized trial of vitrectomy or observation for stage 2 macular holes. *Am J Ophthalmol* 1996;121:605–614.

Macular Dystrophy in a 9-year-old Boy With Fundus Albipunctatus

Makoto Nakamura, MD, and Yozo Miyake, MD

PURPOSE: To report a 9-year-old boy with fundus albipunctatus and macular dystrophy.

DESIGN: Observational case report.

METHODS: A complete ophthalmic examination was performed. The 11-cis retinol dehydrogenase gene (*RDH5*) was examined by direct genomic sequencing.

Accepted for publication Sep 22, 2001.

From the Department of Ophthalmology, Nagoya University School of Medicine, 65-Tsuruma-cho, Showa-ku, Nagoya 466-8550, Japan.

Supported by Grants for Research Committee on Chorioretinal Degenerations from The Ministry of Health and Welfare of Japan and Grant-in-Aid for Scientific Research (Dr. Miyake, A13307048, Dr. Nakamura, C12671703) from the Ministry of Education, Science, Sports and Culture, Japan.

Reprint requests to Makoto Nakamura, MD, Department of Ophthalmology, Nagoya University School of Medicine, 65 Tsuruma-cho, Showa-ku, Nagoya 466-8550, Japan; Fax +81-52-744-2278; e-mail: makonaka@med.nagoya-u.ac.jp