Idiopathic Macular Hole Following Vitrectomy: Implications for Pathogenesis

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BACKGROUND AND OBJECTIVE: Attention to the pathogenesis and clinical features of macular hole formation has increased with the advent of therapy. The purpose of this study is to present three cases that occurred in atypical settings that may have important pathogenic implications.

PATIENTS AND METHODS: The authors conducted a retrospective study of three patients who presented with macular holes that developed 10 months to 5.5 years after previous vitrectomy. In each case, the cortical vitreous layer was absent in the region of the macula at the time of operation for the macular hole.

RESULTS: The macular hole was successfully sealed and the visual acuity improved in all patients. All three cases in this report lacked an operculum, occurred long after vitreous removal, and had no evidence of any residual cortical vitreous.

CONCLUSIONS: Macular hole formation in the absence of cortical vitreous suggests the possibility that the etiology of macular holes may involve a spontaneous umbo dehiscence rather than vitreous-induced surface traction. A mechanism involving a cystic degenerative process is proposed as the cause of atypical and typical macular hole formation.


INTRODUCTION

The pathogenesis of idiopathic macular holes has been commonly thought to be mediated by tangential vitreous traction. A classification system proposed by Gass provided a framework for this hypothesis that is consistent with observed clinical features. Subsequent clinical and histopathologic observations have led to a reappraisal of the classification system, but maintain the role of vitreomacular traction as the initiating event.

The purpose of this study is to report three cases of idiopathic macular hole developing after vitrectomy that suggest an etiologic mechanism that is not dependent on vitreomacular traction.

PATIENTS AND METHODS

Case 1

A 73-year-old woman was referred for evaluation of decreased visual acuity in her left eye of 1 month's duration. The patient's history included a macular scar and poor vision following vitrectomies for macular pucker and subsequent retinal detachment in her right eye. Best-corrected visual acuity was 20/200 with a central scotoma in the right eye and 20/80 with central metamorphopsia in the left eye. Funduscopic examination of the left eye showed laser-induced scars surrounding an
inferior retinal break, a temporal vascular malformation with mild intraretinal lipid deposition, and an elevated macula with dense, preretinal tissue demarcated by vitreous attachments at the posterior pole (peripheral vitreous separation).

The diagnosis of vitreomacular traction syndrome was made, and the patient underwent pars plana vitrectomy with membrane peeling (Fig. 1A). The membrane was removed as one large, discrete piece from the macula. Two months after surgery, the center of the macula was undistorted and free of epiretinal membranes, but there was residual distortion at the original superonasal epicenter and, to a mild degree, nasal and temporal to the fovea (Fig. 1B). Best-corrected visual acuity was 20/30 +3. This degree of distortion remained unchanged and was judged to be typical for a vitreomacular traction syndrome case with this degree of preoperative distortion.

Ten months after the vitrectomy, the patient experienced acute visual loss to 20/100 and a full-thickness macular hole was present (Fig. 1C). The morphology of the hole was typical in all respects: there was a cuff of surrounding subretinal fluid, the hole was discrete and round, and no operculum was present. Along the superotemporal arcade was a recurrent epiretinal membrane that was not contiguous with the central macular area. Around the edges of the hole there was a fine, glistening, membrane-like tissue that extended concentrically from the hole for approximately 200 μm.

Intraoperatively, the larger membrane was removed from the superotemporal arcade and across the papillo-macular bundle. Tissue around the edges of the macular hole was dissected and lysed with a sharp, barbed blade, but a confluent membrane was not identified. Long-acting gas and face-down positioning were used.

Three months after macular hole surgery, the best-corrected visual acuity was 20/40 in the left eye with progressive lens opacification (Fig. 1D).
Case 2

A 62-year-old woman first noted the onset of floaters in her left eye on March 16, 1990. A retinal specialist diagnosed and lasered a retinal tear the following day. On March 29, a dense vitreous hemorrhage completely obscured her vision. Ultrasound showed no evidence of a retinal detachment. She was referred for a second opinion regarding management.

Visual acuity was 20/20 in her right eye and hand motions in her left eye. There was an extensive vitreous hemorrhage in the left eye that appeared to be emanating from a laser-treated retinal break located slightly posterior to the equator. The optic nerve head and macula could not be seen due to dense hemorrhage. Initially, observation was recommended. Two months later, with no evidence of resolution of the hemorrhage, a pars plana vitrectomy was performed. Intraoperatively, a vitreoretinal adherence was noted in the superotemporal quadrant at the site of the previously treated retinal break. The traction was relieved and the blood-laden vitreous was removed. The vitreous dynamics were typical for cases with complete hyaloid separation, but an occult residual cortical hyaloid layer was not specifically sought with a silicone-tipped suction needle. No epiretinal membrane was noted.

One month postoperatively, the patient’s vision was 20/25. Best-corrected visual acuity had worsened to 20/100 in the left eye due to progressive nuclear sclerosis. Cataract extraction with posterior chamber intraocular lens implantation was performed approximately 1 year after vitrectomy, and best-corrected visual acuity returned to 20/20 in the left eye. A YAG-laser capsulotomy was performed 6 months later, and her vision returned to 20/20. She consulted her ophthalmologist on August 15, 1995, because of a 2-week history of decreasing vision in her left eye. On examination, a macular hole was noted, more than 5 years after the previous vitrectomy (Fig. 2). Visual acuity had declined to 20/80. She underwent macular hole surgery 1 week later.

Intraoperatively, no residual cortical vitreous could be demonstrated with the silicone-tipped suction needle. A discrete, confluent piece of epiretinal tissue was removed from the surface of the edges of the hole using a sharp, barbed blade. At the nasal margin of the macular hole there appeared to be a tiny crescent-shaped hole in a thin layer of tissue that was overlying what happened to be the true macular hole. There was a cuff of subretinal fluid surrounding the macular hole. Long-acting gas was used at the end of the case for intraocular tamponade. The visual acuity was 20/25 at the 6-week and 3-month postoperative examinations.

Case 3

A 70-year-old woman presented with slowly progressive loss of vision in the left eye of 1 year's duration. She was referred on October 19, 1993, for evaluation of an epiretinal membrane in her left eye. Cataract surgery had been performed in the left eye in 1991. Best-corrected visual acuity was 20/25 in the right eye and 20/60 in the left. There was mild central metamorphopsia in the right eye with marked metamorphopsia present over the entire Amsler grid pattern as viewed by the left eye. The left macula was covered with a thin, sheet-like epiretinal membrane. She elected to proceed with surgery the following week. There was a typical posterior vitreous detachment, and a discrete, continuous epiretinal membrane was removed. Two months postoperatively, best-corrected visual acuity was 20/40 with an undistorted macula.

Vision remained stable until April 1995 when she experienced a sudden, painless loss of vision 2 months before reexamination. Best-corrected visual acuity was 20/200 in the left eye. The macular examination showed what appeared to be a coalescence of three perifoveal macrocysts. Two months later, a typical, round contour indicated the presence of a full-thickness macular hole. Visual acuity was 20/300, and macular hole surgery was performed.

Intraoperatively, a discrete, thin epiretinal membrane was elevated from the retina around the circumference of the hole using a sharp, barbed blade and vitreoretinal pick, but extended only approxi-
mately 500 µm from the edges. There was a firm annular attachment at the inside edge of the hole. Intraocular forceps were used to rip the elevated preretinal tissue from this firm attachment.

Three months postoperatively, the vision was 20/100 with a closed hole. There was also a cystic appearance to the central retina.

RESULTS/DISCUSSION

Macular holes may be classified as traumatic or idiopathic. Traumatic macular holes appear to be mediated by anterior-to-posterior vitreoretinal traction occurring at the time of traumatically induced posterior vitreous detachment. Idiopathic macular holes are thought to arise secondary to tangential vitreofoveal traction\(^1\)\(^-\)\(^6\) generated during the course of posterior cortical vitreous separation. This hypothesis has been based on the finding of an overlying operculum that appears clinically to correspond to a plug of retinal tissue occurring in association with posterior cortical vitreous separation. The classification system initially proposed by Gass seemed to explain these findings and the clinical course.\(^6\)

Unsatisfactory explanations remained, however, for at least four observations. First, how could visual acuity improve so dramatically after macular hole surgery if a substantial amount of foveal tissue was lost? Second, what specifically initiates traction on the fovea? Third, the original\(^6\) and subsequent classification schemes\(^7\) suggest that macular holes are initially small and subsequently enlarge due to the elasticity of the retinal tissue, but why should this take several months? Fourth, what causes macular holes to develop in atypical settings, such as after apparent posterior vitreous detachment or vitrectomy?

The observation that fibroglial elements (rather than retinal elements) are present in the operculum\(^8\) suggests that the etiologic mechanism does not involve substantial retinal tissue loss, indicating that more viable retinal tissue remains than might be judged intuitively from clinical examination. Fibroglial elements appear to reapproximate the edges of the hole in surgically\(^9\)\(^,\)\(^10\) and spontaneously\(^11\) sealed cases, contributing to recovery of vision. The revised classification\(^7\) scheme for idiopathic macular hole formation postulates a spontaneous umbo dehiscence (rather than a retinal operculum formation) with a variable degree of fibroglial proliferation yielding the substratum for the operculum. These hypotheses are consistent with the histopathologic observations and appear to satisfactorily explain why visual acuity may improve dramatically in some cases.

The stimulus initiating the spontaneous umbo dehiscence is still considered to be tangential vitreoretinal traction.\(^7\) Although some contractile cells have been found in the vitreous of eyes undergoing surgery for pre–macular hole lesions,\(^12\) no activating stimulus has been identified and it is also unclear whether they are the stimulus or the result of hole formation. Preretinal membrane tissue has been documented frequently in association with a macular hole,\(^13\) but has been thought to be a secondary change because it seems to be more apparent with older, later-stage holes.

Once the umbo dehiscence occurs (through whatever mechanism), its enlargement is postulated to be mediated by the inherent elasticity of the central macular tissue.\(^7\) An epiretinal specimen consisting of glial cells was removed from the edges of the hole in a case of a recurrent macular hole.\(^14\) This suggests the possibility that centripetal glial cell proliferation may be causing the progressive enlargement—a hypothesis that seems more consistent with the time course of the enlargement.

A final concern raised above is especially pertinent to the current report: What explains the presentation of atypical cases? Patients have been reported to have macular holes after what appeared to have been well-documented posterior vitreous detachments\(^15\) and after rhegmatogenous retinal detachment.\(^15\)\(^-\)\(^18\) A hypothesis that only segmental posterior vitreous separation occurs with persistent attachment at the time of the peripheral retinal break was offered,\(^16\) but this inadequately explains the clinical observations. One case of macular hole formation occurring 2 weeks after vitrectomy and epiretinal membrane peeling was reported, but similarly was not satisfactorily explained.\(^16\)

The cases in this report and the previously reported atypical cases\(^15\)\(^-\)\(^18\) have important pathogenic implications, at least for these atypical cases and possibly for typical cases. The three cases in this report lacked an operculum, occurred long after vitreous removal, and had no evidence of any residual cortical vitreous. Although it is possible that residual epiretinal membrane proliferation could have played an etiologic role in the formation of the macular hole in case 1, the circumferential, typical appearance of the hole makes this less likely. Also, an occult residual cortical hyaloid layer was not sought at the time of the original vitrectomy in case 2. The vitreous dynamics during initial
removal were typical of a case with full hyaloid separation, and it was not present at the macular hole surgery. Vitreofoveal traction, therefore, does not seem to have played a role in macular hole formation in these cases.

We suggest that these observations can be reconciled with Gass's revised hypothesis, but with a couple modifications. First, we propose that a degenerative process initiates the spontaneous umbo dehiscence (Fig. 3). This may merely represent an atrophic attenuation of the thin foveal center, much the same as peripheral atrophic holes form. The authors have observed a macrocystic change in some cases that may be involved in the process whereby a rupture of the internal and external borders of such cysts may be the final step in completing the umbo dehiscence. This mechanism involving the breakdown of a foveal cyst has been previously suggested^{13,19,20} and appeared to be the cause of a reported case of lamellar macular hole.\(^{21}\)

We postulate that a similar degenerative process is also consistent with the changes observed in typical cases. Although this process may occur in the absence of a supra-adjacent vitreous cortex layer, as in the cases reported here, it probably usually occurs underlying it in typical cases. In that more typical situation, the layer is available as a template on which a spontaneous reparative process involving glial elements may form, leading to the occult stage 1B macular hole configuration described by Gass\(^7\) (Fig. 4). If, instead, the glial proliferation spreads along the retinal surface and subsequently contracts, the edges of the umbo dehiscence are pulled outward and the standard macular hole morphology results (Fig. 5). Such glial proliferation may represent the fine epiretinal membranes that are surgically dissected or removed from the margins of the hole during surgery. The firmer adherence of that layer to the margin of the hole routinely observed by the authors intraoperatively could be due to its continuity with the intraretinal source cells.

Subsequently, with cortical vitreous separation in
This study reports three cases of atypical macular holes that, similar to previously reported atypical cases, do not appear to involve vitrefoveal traction. We propose a pathogenic theory that postulates that the spontaneous foveal dehiscence observed by Gass may be the result of a degenerative process independent of the presence of the cortical vitreous. Furthermore, we suggest that this process may also be the mechanism of typical macular hole formation.

REFERENCES


