In this study, we were able to confirm Lincoff's notion of a double layer detachment in 10 of 11 eyes with optic disc pit maculopathy.[5] An outer layer detachment was observed either preoperatively or postoperatively as a secondary phenomenon, developing after presence of an inner layer separation in 3 eyes. In the remaining 7 eyes, the outer layer detachment, along with an outer layer break, were observed at presentation.

Lincoff and colleagues reported on the successful use of intravitreal gas tamponade without vitrectomy to induce pneumatic displacement of the outer layer detachment and improve central vision.[17] However, OCT findings have suggested that fluid may continue to flow from the remaining inner layer separation to the outer layer detachment.[8] Since we believe that vitreoretinal traction is an important factor in the pathogenesis of macular detachment associated with optic disc pit, we feel that vitrectomy with complete PVD induction is essential for the treatment of these eyes. We postulate that vitreous traction may play a critical role in the development of these detachments, although we were unable to detect any vitreomacular or vitreopapillary traction by OCT. The added use of gas tamponade probably helps to push fluid from the inner layer separation into the outer layer detachment. Since we did not apply laser photocoagulation at the optic disc pit margin in our patients, we deduce that release of continued vitreoretinal traction by surgical induction of PVD was sufficient to lead to diminishment of new fluid accumulation in the inner layer separation presumably coming from the optic disc pit. This idea is supported by our postoperative OCT findings in Patient 10, showing that the inner layer separation resolved first before resolution of the outer layer detachment.

Vitrectomy with gas tamponade, with or without laser application to the margin of the pit, for macular detachment associated with optic disc pit has been reported.[12-14] However these previous series did not achieve as good visual acuity results and had

higher rates of re-operation compared to the current study. This difference may be due to differences in surgical technique. Furthermore, the current study did not employ any laser application to achieve good visual and anatomical outcomes, and we believe that assuring a complete PVD induction during surgery played an integral part in achieving these results.

Favorable visual and anatomical outcomes have also been reported using macular buckling in a series reported by Theodossiadis and colleagues of 9 consecutive patients with optic disc pit maculopathy.[16] Recently the same group has also reported on OCT findings in 26 eyes treated with scleral buckling, with resolution of macular detachment noted in 24 eyes. Disappearance of intraretinal fluid over 7 to 9 months postoperatively was documented in 4 of 5 eyes that were evaluated by OCT both before and after surgery.[27] We believe that these favorable results with scleral buckling are due to conversion of the inward perpendicular component of the vector, caused by tangential forces associated with posterior hyaloid traction on the retina at the irregular surface of the optic disc, to an outward perpendicular vector component by the scleral buckle promoting attachment of the retina.[28]

Spontaneous resolution of the macular detachment associated with optic nerve pits is also known to occur in approximately 25% of untreated patients.[4] However, cystic retinal degeneration, macular hole formation, and retinal pigment epithelial atrophy often limits visual recovery in these cases. Over one-half of eyes with macular detachments experience a decrease in VA to 20/100 or less within 5 years.[3] In contrast, although the time to best VA took close to 6 months in most patients, 9 of 11 eyes eventually achieved a postoperative visual acuity of 0.8 or greater. Furthermore, we found that visual acuity started to improve after surgery despite the continued, albeit decreased, presence of inner

layer separation and outer layer detachment in the fovea by OCT. This suggests that mild improvements in foveal contour alone may lead to improved vision, and that longterm observation is essential postoperatively in these eyes. In eyes that have undergone vitrectomy, additional treatments for the macular detachment should not be contemplated too early. Furthermore, given the fact that subretinal fluid persisted for a long duration postoperatively but eventually resolved, gas tamponade may actually not be necessary for successful treatment. Since all eyes in the current study received gas tamponade, we are not able to assess this possibility.

Complications associated with surgery included visual fields defects in 2 of 11 eyes, both of which were early cases in our series. We believe that improved surgical technique allowed us to avoid such complications in later cases.[29,30] Since the induction of PVD in young eyes is particularly difficult due to strong vitreoretinal adhesion, special care must always be taken to avoid excessive mechanical damage when separating the posterior hyaloid from the optic disc and posterior pole. A longer duration of surgery may also lead to an increased risk of light toxicity as has been reported in macular hole surgery.[29-31] However, surgery-related insults must also be distinguished from visual field aberrations that may be present preoperatively in these eyes with optic disc pits.

In conclusion, vitrectomy with modern surgical techniques for creating PVD in young patients and gas tamponade, without laser photocoagulation, can lead to successful reattachment of the macula and improvement in central vision in eyes with optic disc pit maculopathy, although most eyes required almost 1 year to reach this state.

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Figure Legend

Figure 1. Composite of representative clinical findings from Patients 1 (A-C) and 10 (D-I). All optical coherence tomography (OCT) scan lengths were 10 mm. (A) Photograph of the left fundus of patient 1 preoperatively, showing an inferotemporal optic disc pit associated with an large, oval-shaped area of macular detachment (large arrows), a nerve fiber layer defect leading from the optic disc pit (green arrows) and an area of myelinated nerve fibers in the inferior macula. The outer layer hole (blue arrows) and round, 1.5 disc diameter, outer layer detachment (white open arrowheads) also shown was not present initially but developed after 4 months of follow-up. Preoperatively, the best-corrected visual acuity (VA) was 0.2. (B) Two months after vitrectomy and gas tamponade, an irregularly-shaped break was noted in the outer layer (blue arrows), with the outer layer detachment (white open arrowheads) being slightly enlarged. A new nerve fiber layer defect was also observed leading from the superotemporal edge of the optic disc (green arrows). (C) One year postoperatively, the macula was observed to be completely reattached with a VA of 1.0. The nerve fiber layer defects remained unchanged. (D) Fundus photograph of the left eye of Patient 10 at presentation, showing a shallow retinal elevation extending from the superotemporal to the inferotemporal arcades. A round, 2 disc diameter, outer layer detachment was also observed in the center of the macula and preoperatively the VA was 0.08. (E) OCT at presentation revealed an inner layer separation as well as a hole in the outer layer detachment at the macula and an outer layer detachment surrounding the hole. (F) Three months after vitrectomy and gas tamponade, OCT showed a decrease in the inner layer separation and outer layer detachment. (G) At 9 months OCT showed resolution of the inner layer separation but residual outer layer detachment. (H, I) Fundus photograph and OCT at 12 months

postoperatively, showing the macula completely reattached and the optic disc pit appearing grayer and deeper than preoperatively. The VA was 1.2.

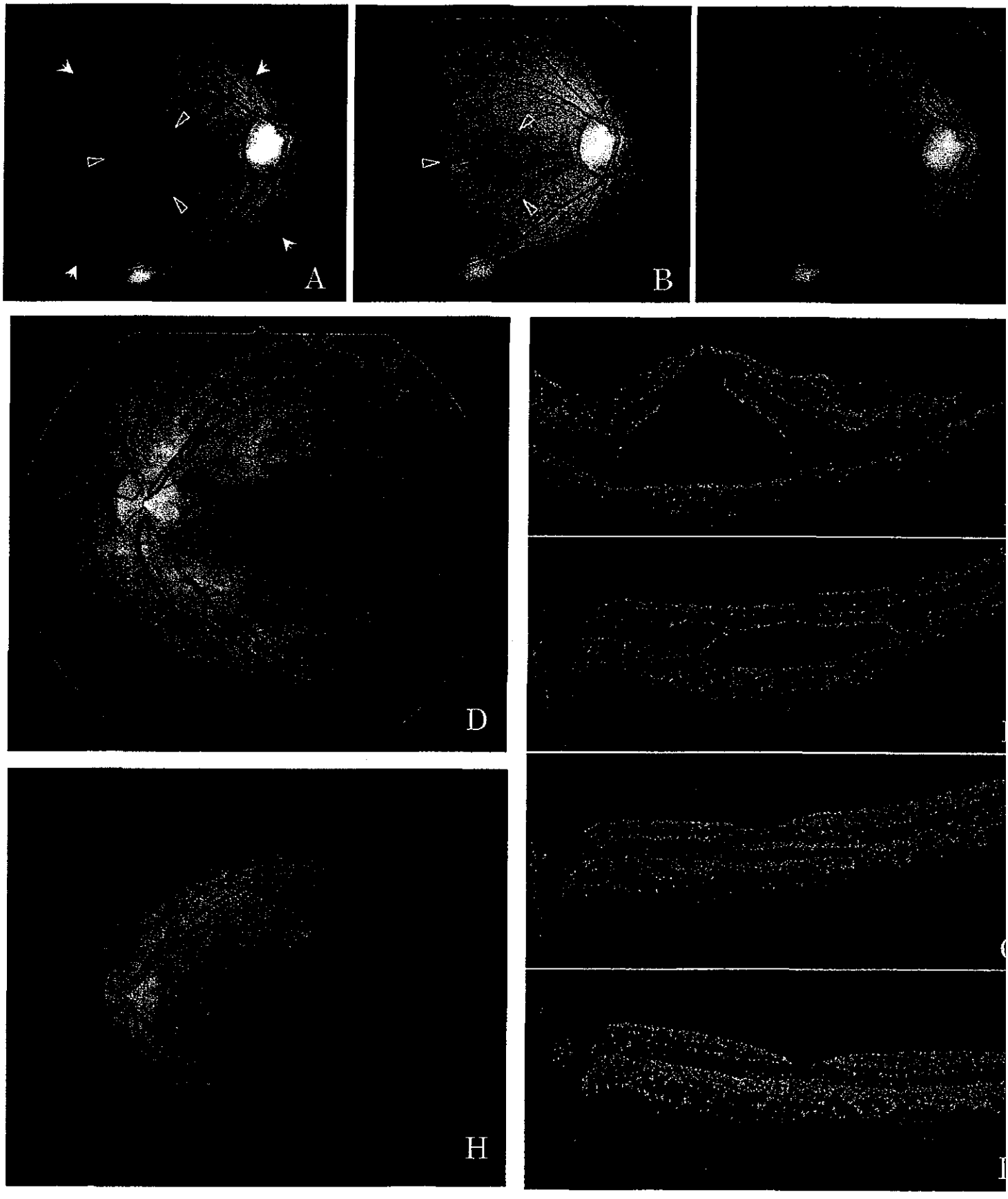
Précis

Vitrectomy and gas tamponade, without laser photocoagulation to the margin of the optic nerve, is sufficient for successful treatment in most eyes with macular detachment associated with optic disc pit.

Table 1. Clinical Characteristics

Patient	Age	Sex	Eye	Ref (D)	Symptom	Pre-op VA	Pit Location	Duration of Symptoms#	Double Layer RD	Outer layer hole at fovea	PVD	Duration Until Macular Attachment#	Final VA	Complications	Follow-up#
1	40	F	R	-2.0	central scotoma	0.3	inferotemporal	5.0	+	+	−	12	1.2	peripheral VF defect	80
2	15	F	L	-1.0	decreased VA	0.2	inferotemporal	36.0 <	+	+	−	2	0.04	retinal damage	91
3	43	F	L	-1.5	central scotoma	0.08	temporal	6.0	+	+	−	5	0.1	−	98
4	30	M	R	-0.5	central scotoma	0.6	temporal	6.0	+	+	−	8	0.8	retinal break	24
5	22	M	L	-1.0	metamorphopsia	0.5	inside in coloboma	2.0	−	−	−	12*	1.2	−	73
6	19	M	R	0.0	central scotoma	0.4	temporal	9.0	+	+	−	12*	1.0	−	37
7	24	M	L	0.0	central scotoma	0.6	inferior	24.0	+	−	−	12*	1.2	−	15
8	42	F	R	-1.0	metamorphopsia	1.0	central	2.0	+	+	−	15	1.0	−	57
9	47	M	L	0.0	central scotoma	0.2	inferotemporal	3.0	+	+	−	10	1.2	−	14
10	8	F	L	-1.0	decreased VA	0.08	temporal	5.0	+	+	−	12	1.2	−	12
11	44	F	R	0.0	decreased VA	0.4	temporal	24.0	+	+	−	flatter	1.0	−	10

* Displacement of subretinal fluid immediately after surgery
# duration in months
PVD = posterior vitreous detachment; RD = retinal detachment; Ref = refractive error; VA = best corrected visual acuity; VF = visual field.


A
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Multi-layered Retinoschisis associated with Optic Disc Pit

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Abstract

Background: Recent optical coherence tomography (OCT) analysis has contributed greatly to the detection of posterior retinoschisis associated with optic disc pits. We report an unusual case of optic disc pit maculopathy.

Case: A 37-year-old woman with unusual posterior retinoschisis associated with an unusually tiny, shallow optic disc pit was observed.

Observations: The patient presented with a few weeks' history of decreased vision in her right eye. Fundus examination showed a shallow macular detachment with a tiny shallow pit at the temporal edge of the optic disc. OCT revealed an unusual multi-layered structure of retinoschisis connected to the optic disc. We performed vitrectomy with induction of the posterior hyaloid separation and gas tamponade. After vitrectomy, OCT showed a marked fluid resolution in the retinoschisis, and the patient's vision improved rapidly.

Conclusion: The multi-layered separation of retinoschisis can be caused by the traction of the optic disc with a tiny shallow pit at the temporal edge of the optic disc.

Key words:

Macular detachment

Optical coherence tomography (OCT)

Optic disc pit

Retinoschisis

Vitrectomy

Introduction

Lincoff and co-authors proposed that the retinal elevation that communicates with optic disc pits is a schisis-like separation of the internal layers of the retina, based on a study of stereoscopic transparencies and visual fields performed in 1988.[1] Outer layer detachment centered over the macula was suggested to be a secondary phenomenon. More recently, several authors have confirmed the two-layered structure of optic disc pit maculopathy using optical coherence tomography (OCT).[2,3] However, the source of the fluid in the retinoschisis cavity remains unclear.

We report a 37-year-old woman with an unusual multi-layered structure of retinoschisis associated with a tiny shallow pit at the temporal edge of the optic disc.

Case Report

A 37-year-old Japanese woman was referred to our hospital with a few weeks' history of acute decreased vision in her right eye in April 2004. Her best-corrected visual acuity (BCVA) was 0.5 OD and 1.2 OS. Her medical history was unremarkable. Fundus examination of the right eye revealed a serous detachment of the macula linked to the optic disc (Fig.1A). The optic disc was slightly enlarged and exhibited a relatively large cupping with a tiny shallow pit at the temporal edge of the optic disc (Fig.1A). Optical coherence tomography (OCT; Humphery Instruments, San Leandro, CA) disclosed a multi-layered separation of the neurosensory retina between the optic disc and fovea (Fig.1B,1C). The irregular split in several layers of the papillomacular area was unusual in its appearance and

association with the optic disc pit. A lamellar break of inner layer and retinoschisis in the outer layer of sensory retina were present around the center of the macula. Fluorescein angiography showed a weak leakage from the retinal small vessels beside the optic disc and late staining of the optic disc. After 2 months of observation, she complained of a worsening in her central vision and metamorphopsia in her right eye. We decided to perform vitrectomy to improve her visual acuity. Advanced vitreous syneresis, but no posterior vitreous detachment, was observed during the operation. Triamcinolone acetonide was used intraoperatively to highlight the posterior hyaloid membrane. The separation of the posterior hyaloid was smoothly performed, even though the adhesion of the posterior hyaloid was very tight at the anterior border of the pit at the temporal edge of the optic disc. Fluid-air exchange was performed using a gas tamponade with 20% sulfur hexafluoride (SF_6). The patient was maintained in a postoperative face-down position. Two weeks postoperatively, the patient's BCVA was 0.9, and the macular elevation was markedly reduced. OCT showed the marked reduction of the irregular split in several layers of the superficial neurosensory retina at the papillomacular area. Two months postoperatively, the BCVA improved to 1.2, with decreased residual retinoschisis of the posterior neurosensory retina (Fig.2A, B).

Discussion

Recent OCT findings support the concept of a bilaminar structure in which a macular detachment develops secondarily to a pre-existing retinoschisis, consisting of severe outer retinal edema.[2,3] In previous reports, the retinoschisis cavities have

been located deeper, at the outer plexiform layer, compared to that in the present case. The present patient exhibited a multi-layered separation or severe edema. The separation in the more superficial part, seemed to be at the neurofiber layers between the optic disc and fovea. The shape and position of the retinoschisis in the outer layer of sensory retina around the center of the macula were similar in appearance to that of retinoschisis usually seen in optic disc pit maculopathy. The difference in present case might be caused by the shape and position of the pit. The pit was atypically small and seemed to be a partial defect of the neurofiber layer inside the optic disc. Todokoro and Kishi reported a case with a split in several layers of the papillomacular area associated with an optic disc pit.[4] Using OCT analysis, they observed that the optic disk pit was not a true pit, but a cystic cavity covered with a superficial layer of the optic disc. Several variations in the structure or position of the pit at the optic disc may lead to the different OCT appearances of the maculopathy. The vision in the present case relatively acutely decrease and was restored soon after the surgery. This clinical course of the visual acuity in the present patient was also different from those usually observed in patients with optic disc pit maculopathy.

Several authors have suggested that posterior vitreous traction may have an important role in the pathogenesis of optic pit maculopathy, based on OCT examinations or experience in performing vitrectomies.[4] In this case, the vitrectomy reduced the serous detachment, enabling an improvement in the patient's BCVA. During the vitrectomy, the posterior hyaloid was tightly attached to the anterior edge of the tiny shallow pit at the temporal edge of the optic disc. The late staining at the optic disc and the weak leakage from the small retinal

vessels during the fluorescein angiography may have been caused by vitreous traction at the optic disc. Posterior vitreous traction may have introduced the fluid from the shallow pit at the temporal edge of the optic disc to the neurosensory retina, causing the irregular split in several layers of the papillomacular area.

Spaide and co-authors reported a 29-year-old patient with a macular schisis without an optic disc pit.[5] OCT findings showed a thin sheet of tissue with a small break or fenestration on the optic nerve. The abnormal structure on the optic disc and/or the pre-existing tight adhesion between the posterior vitreous, including the abnormal membrane-like tissue, and the optic disc might have produced the retinoschisis, even if an optic disc pit was not present. If the fluid in the unique retinoschisis in the present case originated from the unusual small and shallow pit at the edge of the optic disc, the fluid in the two-layered structure of typical optic disc pit maculopathy may arise from a much deeper level, possibly the subarachnoid space.

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