New Insights Into Progressive Visual Loss in Adult Retinopathy of Prematurity

An abnormal vitreous and aberrant vitreoretinal traction are significant factors in the cataractous changes and poor outcomes in neonates with retinopathy of prematurity. Late complications of retinopathy of prematurity (ie, retinal tears or rhegmatogenous retinal detachment) have also been described and these, too, can be attributed to an abnormal vitreoretinal relationship.1 Tasman and Brown2 observed that progressive visual loss may be noted in adult retinopathy of prematurity without a definitive clinical cause. Tangential vitreoretinal traction was speculated to result in cystoid macular edema or compromised photoreceptors with resulting pigmentary disturbances and visual deterioration.2 Herein we report our observations in a similar case of adult retinopathy of prematurity and with the aid of optical coherence tomography note that subclinical tractional retinal detachment may be the cause of visual loss in at least some of these cases.

Report of a Case. A 29-year-old woman had been delivered at gestational age 28 weeks with a birth weight of 1304 g. She had a history of retinopathy of prematurity. She was referred to our center for examination of a 7-month history of progressive visual loss in her left eye. The patient had previously undergone multiple procedures in the left eye for treatment of glaucoma and subsequent cataract extraction with intraocular lens placement at 16 years of age. Her baseline visual acuity was 20/60 OS before the onset of her recent symptoms. She had not previously undergone cryotherapy or laser treatment in either eye. Her right eye had been enucleated 2 years previously for end-stage glaucoma and complications following rhegmatogenous retinal detachment related to previous intraocular surgeries. On her initial visit, her visual acuity measured 20/400 OS. Biomicroscopic examination results of the left eye revealed a quiet anterior segment with a posterior chamber intraocular lens and a central opening in the posterior capsule. Examination results of the right orbit revealed an ocular prosthesis. Fundusscopic examination results of the left eye showed pulled retinal vessels at the optic nerve (ie, a dragged disc) with temporal dragging of the fovea. A small organized area of central vitreous attached to the optic disc was present. Retinal pigment epithelial change was noted in the macula, but no clinically appreciable cystoid macular edema, epiretinal membrane formation, macular hole, retinal detachment, or other macular pathologic features were noted. The peripheral retina was attached and had an avascular appearance consistent with a history of retinopathy of prematurity. The patient was asked to undergo fluorescein sodium angiography and optical coherence tomography testing.

At a follow-up examination 1 week later, the patient had a further decrease in visual acuity to 20/800 OS. Slitlamp and fundusscopic examination results of the left eye remained unchanged. Fluorescein angiography results of the left eye revealed retinal pigment epithelial clumping without intraretinal edema or other abnormalities of the posterior segment other than dragging.
ence tomography findings, however, did reveal a very shallow but obvious retinal detachment involving the center of the macula (Figure 2).

An uncomplicated vitrectomy with adjuvant autologous plasmin enzyme was subsequently performed. The hyaloid was detached and all lenticular remnants were removed. Her postoperative course was complicated by a filamentary keratitis that required an extended course of topical antibiotic therapy and a therapeutic contact lens. For this reason, her visual acuity remained at light perception to bare hand motions for the first 2 postoperative visits. However, at postoperative week 4, the patient began using a therapeutic contact lens and reported improved vision concomitant with resolution of her filamentary keratitis. Visual acuity at this time measured 20/70 OS at distance. Funduscopic examination results of the left eye revealed a clear vitreous cavity but an otherwise identical-appearing posterior segment and peripheral retina when compared with the preoperative examination findings. However, repeat optical coherence tomography results showed resolution of the macular detachment and restoration of a foveal depression (Figure 3).

Comment. The recent use of optical coherence tomography has demonstrated that visual loss in various vitreoretinal syndromes may result from a subclinical phenomenon. Such conditions include poor vision associated with choroidal nevi, delayed visual recovery after retinal detachment repair, and posterior hyaloidal traction syndrome associated with diabetic macular edema.3-5 Optical coherence tomography findings in these cases show the presence of subretinal fluid with macular detachment not appreciable by clinical examination or with fluorescein angiography. Similarly, in our patient here, with otherwise unremarkable clinical and fluorescein angiographic findings, we were able to document the etiology of her visual loss as subclinical retinal detachment only through optical coherence tomography and not through any other technique.

As mentioned previously, abnormal vitreous traction plays a causative role in both neonatal and adult manifestations of retinopathy of prematurity. When present, vitrectomy is indicated to relieve vitreoretinal traction and restore the normal anatomic configuration of the macula. Plasmin enzyme may be useful in these patients in particular because hyaloidal separation is routinely difficult in young adults and especially in patients with retinopathy of prematurity who have an abnormal vitreoretinal interface.6 The posterior hyaloid in these cases is uncommonly adherent and when contracted can result in tractional detachment of the retina and even vascular stasis.7 In our case, con-

Figure 2. Preoperative horizontal optical coherence tomography section results reveal a very shallow but obvious macular detachment with subretinal fluid.

Figure 3. Postoperative horizontal optical coherence tomography results show resolution of the macular detachment with restoration of normal macular anatomy.
traction of the vitreous and hyaloid as noted by organization of the vitreous overlying the optic nerve presumably resulted in macular detachment and visual loss. Plasmin-assisted vitrectomy was successful in reestablishing the patient’s normal macular anatomy and restoring her baseline visual acuity. When present and confirmed by careful clinical examination, including adjunct methods such as optical coherence tomography, subclinical tractional retinal detachment in patients with adult retinopathy of prematurity with otherwise unexplained visual loss can be successfully treated by vitrectomy. The results from using optical coherence tomography in our case explain a circumstance that for many years had been observed yet was inadequately understood by physicians managing adult patients with retinopathy of prematurity.

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Transretinal Pigment Migration: An Optical Coherence Tomographic Study

Cells of the retinal pigment epithelium (RPE) play an important role in the formation of epiretinal membranes after retinal detachment. The RPE cells may access the inner retinal surface through the retinal break that caused the detachment. Subsequent proliferation of these cells on the retinal surface then contributes to the formation of the epiretinal membranes, which typically consists of a variety of different cell types.

A second mechanism by which the RPE cells might access the inner retinal surface is by migration through the retina. The ability of proliferating RPE cells to migrate is well documented and can be altered by the cells’ microenvironment. Proliferation of RPE cells occurs not infrequently after trauma and after retinal detachment repair. Experimental models of retinal detachment show that the RPE cell proliferation begins early in the course of the detachment and is confined to the region of the detachment.

Translocation of RPE pigment into the retina occurs in some diseases such as retinitis pigmentosa (“bone spicula”) as well as in experimental models of subretinal pigment clumping. The advent of optical coherence tomography (OCT) has provided an opportunity to obtain high-resolution, cross-sectional images of the retina and RPE in a variety of disease states. Optical coherence tomography provides an indirect image of the retina and RPE based on the reflective properties of the various cellular layers. The varying intensities seen on the OCT scan correlate well with the different levels of the retina and RPE. The RPE provides the most highly reflective surface, and usually appears as a bright orange-red layer on the OCT scan. In this article we report 2 cases of retinal detachment with subsequent migration of RPE cells into the retina. Optical coherence tomographic scans in both of these patients showed highly reflective material that was consistent with RPE proliferation and migration through the retina into the region of secondary epiretinal membranes. These findings suggest that the transretinal migration of proliferating RPE cells may have played a role in the formation of epiretinal membranes in these patients.

Report of Cases. Case 1. A 52-year-old man was initially seen with progressive loss of vision in the right eye over the previous 6 days. Ophthalmic examination was significant for visual acuity of counting fingers OD at 1 ft and 20/20 OS. Dilated fundus examination of the pseudophakic right eye showed a near-total retinal detachment with a single horseshoe retinal tear at the 12:30 o’clock position. The detachment was repaired with pars plana vitrectomy, retinotomy with internal drainage of subretinal fluid, endolaser photocoagulation, and a perfluoropropane fluid–air gas exchange (12%) followed by facedown positioning. At the 1-month postoperative visit, visual acuity was 20/200 OD and subretinal pigment clumping was evident in the macula. Three months after the retinal detachment repair, the visual acuity had improved to 20/40 OD. There was consolidation of the subretinal pigment clumps (Figure 1A). The OCT scan showed highly reflective spots emerging from the RPE and extending anteriorly into the retina (Figure 1B). These intraretinal spots had the same reflective intensity as the RPE and were found in the same location as the RPE pigment seen on the fundus photograph. Mild patches of glistening on the retinal surface suggested early epiretinal membrane formation.

Case 2. A 44-year-old man was initially seen in our clinic 6 weeks after primary repair of an open-globe injury with a nonclearing vitreous hemorrhage and retinal detachment. The patient underwent pars plana vitrectomy. During perfluoropropane fluid–air gas exchange, a large mobile pigment plaque was observed in the subretinal space in the superior midperiphery. Over the next 7 months, the patient developed an epiretinal membrane over the macula that extended well beyond the vascular ar-