unusual systemic locations, such as the eye. Our case is, to our knowledge, both the most severe that has been reported and the first noting retinal vascular tortuosity. The corkscrew vascular pattern is reminiscent of that in Fabry disease, a related disorder of sphingolipid metabolism. In our case, there were Gaucher cells in the vitreous cavity, while the vitreous gel contained large amounts of glucosylceramide.

The pathophysiologic mechanism of glucosylceramide deposition in the vitreous cavity is unclear. The material is a by-product of breakdown of myelin, leukocytes, red blood cells, and endothelial cells; myelin may be deposited within the eye when oligodendroglial cells migrate through the lamina cribrosa. Alternatively, deposition may occur by leakage from the vasculature. The latter is less likely, as glucosylceramide is not deposited within neurons in type 1 Gaucher disease, although Gaucher cells do accumulate in the perivascular macrophages of the brain.1

We have shown that vision may be improved with vitrectomy in patients with Gaucher disease. It is unknown whether vitreal opacities will recur in our patient, as has been reported in patients who have undergone vitrectomy for vitreous opacities in familial amyloidotic polyneuropathy syndrome.2 The relationship of long-term enzyme replacement therapy to the vitreous opacities in our patient is speculative.

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Choroidal Neovascularization After Central Retinal Vein Occlusion in a 47-Year-Old Man

Report of a Case. A 46-year-old white man was referred to The Wilmer Ophthalmological Institute, Baltimore, Md, for evaluation of macular edema. He had been diagnosed as having a nonperfused central retinal vein occlusion (CVO) in the right eye 5 months previously, as documented by photographs and a fluorescein angiogram from his referring ophthalmologist (Figure 1). Medical history was positive for hypercholesterolemia. Medications included aspirin and a lipid-lowering agent.

Visual acuity at the time of referral was 20/160 in the right eye and 20/20 in the left eye. There was no iris or angle neovascularization. Intraocular pressure was 16 mm Hg in each eye. Ophthalmoscopic examination showed resolving intraretinal hemorrhages in all 4 quadrants in the right eye, with foveal thickening and cystic edema (Figure 2). The patient was diagnosed as having a nonperfused CVO with macular edema. Observation by his referring ophthalmologist for development of any neovascularization was recommended. He was told to return to The
Wilmer Ophthalmological Institute in approximately 6 months to determine whether he still had macular edema with perfusion of macular capillaries. If edema persisted, laser photocoagulation might be considered, because a trend toward an increased chance of stable visual acuity after photocoagulation, compared with observation, had been demonstrated in younger patients with perfused macular edema in the Central Vein Occlusion Study.1

Six months later, 11 months after the onset of the CVO, the patient reported a slight improvement in vision, although measured visual acuity remained unchanged at 20/160 in the right eye. No neovascularization was seen in the anterior segment. Ophthalmoscopic examination showed resolution of the intraretinal hemorrhages. However, a new subretinal hemorrhage was seen in the macula with fibrovascular tissue that just extended into the fovea from the superonasal aspect of the macula. There was cystoid macular edema at the infero-temporal aspect of this lesion (Figure 3A). An early-phase frame from a fluorescein angiogram (Figure 3B) showed an anastomosis between a retinal arteriole and the fibrovascular lesion, as well as an anastomosis between a retinal venule and the lesion. Within the fibrovascular tissue, early, intense, and well-circumscribed hyperfluorescence with late leakage was seen. There also was late leakage of dye in a petalloid pattern at the inferotemporal aspect of the fibrovascular lesion corresponding to the cystoid macular edema (Figure 3C). A diagnosis of subfoveal, predominantly classic choroidal neovascularization (CNV) was made. Although the cause of the CNV was not clear, photodynamic therapy (PDT) was recommended because the lesion was subfoveal and because it was predominantly classic.

Extrapolation from the Treatment of Age-Related Macular Degeneration With Photodynamic Therapy Investigation2 was judged applicable to this patient to reduce the risk of moderate and severe visual acuity loss compared with no treatment. The patient underwent PDT with verteporfin (Visudyne; Novartis Pharma AG, Basel, Switzerland), without complications.

Three months later, visual acuity was 20/250 in the right eye. Ophthalmoscopic examination showed resolution of much of the subretinal hemorrhage but some subretinal fibrosis (Figure 4A). Fluorescein angiography showed continued leakage from CNV and cystoid macular edema (Figure 4B and C). Because of the continued fluorescein leakage from the CNV, a second course of PDT was performed. Seven months after the patient’s first course of PDT and 18 months after the recognition of his CVO, the visual acuity remained 20/250 in the right eye. Ophthalmoscopic examination showed a decreased amount of subretinal fluid and more subretinal fibrosis. Some intraretinal hemorrhages overlay the subretinal fibrosis (Figure 5A). Angiography showed minimal fluorescein leakage from CNV, resolution of the leakage from the cystoid macular edema, and staining around the fibrovascular lesion. The anastomoses between the fibrovascular tissue and the retina showed little flow of fluorescein (Figure 5B and C). With only little fluorescein leakage from the CNV, PDT was not repeated.

Comment. Documentation of development of macular CNV after CVO, with retinal arteriole or venous anastomosis to the CNV, is of interest because of the recent attention given to retinal vascular anastomoses to CNV lesions.3-7 Our literature search disclosed no previous reports of CNV occurring in young patients within a short time after a CVO. One report documented CNV occurring in an older individual years after a vein occlusion.8 All-
though these 2 diseases (CNV and CVO) could occur in the same eye, independent of each other, a causal relationship appears to exist in this case. This is supported by the following: (1) the absence of any other known causes of CNV in this young patient; (2) the temporal relationship of developing CNV within 6 months after documentation of the CVO; and (3) the retinal anastomoses to the CNV that were observed at the initial presentation of the CNV.

What could be the relationship of the CVO to the CNV? One could postulate that chronic edema of the retina led to progressive damage of the retinal pigment epithelium, with disruption to the Bruch's membrane, that was associated with the growth of CNV. It is also possible that some CNV lesions begin in the retina and extend to the choroid, as has been documented previously in idiopathic paravascular telangetasias and more recently suggested in age-related macular degeneration. Increased hydrostatic pressure in venous collaterals, secondary to the CVO, could have either initiated or potentiated the development of such anastomoses. As shown in Figure 3A, the retinal hemorrhages caused by the CVO largely disappeared at about the same time that anastomoses between the fibrovascular lesion and the retina were first observed. This suggests the possibility that these spontaneously occurring anastomoses decompressed the occluded retinal venous vasculature into the fibrovascular lesion and into the choroid. Such retinal venous decompression has been documented therapeutically, after intense laser photocoagulation designed to rupture the Bruch's membrane and create anastomoses between the retina and choroid.
Whatever the cause of the CNV, the PDT was associated with a diminution of fluorescein leakage from CNV, despite the presence of anastomoses between the retina and the fibrovascular tissue. The PDT appeared to have had little immediate effect on the retinal vessels, although the anastomotic vessels became nonperfused over time. Regardless of the cause of the CNV after CVO, whether or not there is a causal relationship, we believe that this is the first well-documented case of CNV developing in a relatively young patient with a preexisting CVO. However, retinal anastomoses and cystoid macular edema were observed. In addition, PDT appeared to have a typical involutorial effect on the fibrovascular lesion.

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