Laser-Induced Chorioretinal Anastomosis in Neurofibromatosis Type 1

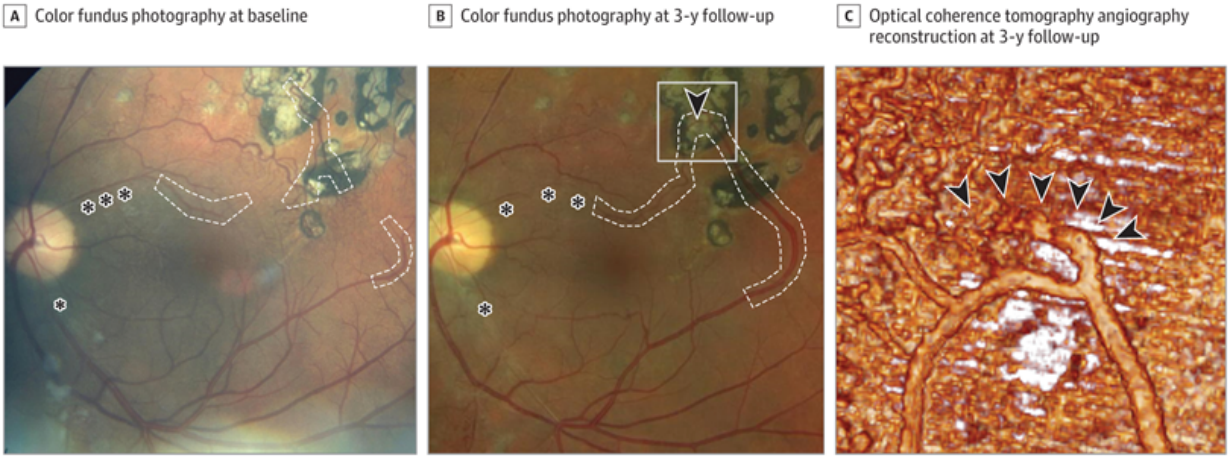
Report of a Case

A 38-year-old woman with neurofibromatosis type 1 (NF1) presented for routine ophthalmic examination. The patient had long-standing peripheral retinal capillary nonperfusion in the left eye, which had been treated years ago with panretinal photocoagulation. Both eyes showed choroidal Yasunari nodules, which are choroidal lesions not easily detected on ophthalmoscopic examination but readily visible by near-infrared photography.[1] The right eye showed no other pathologies. Visual acuity was 20/20 OU with correction. Recent magnetic resonance imaging of the neuraxis and orbits showed no gliomas. The patient was not known to have any systemic vasculopathy.

Compared with a visit 3 years earlier, the central retinal artery and branching arcades had thinned. A chorioretinal anastomosis had formed in the superotemporal macula at the site of a photocoagulation scar; a spiral-shaped choroidal vessel interconnected with both arterial and venous vessels from the superior arcades and a vein from the inferior arcade, which perfused the arterial supply of the central retina, including the fovea (Figure 1). Fluorescein angiography confirmed the new retrograde arterial supply through the anastomosis (Figure 2; Video). The formation may have been associated with a combination of a hydrostatic pressure gradient over the retinal

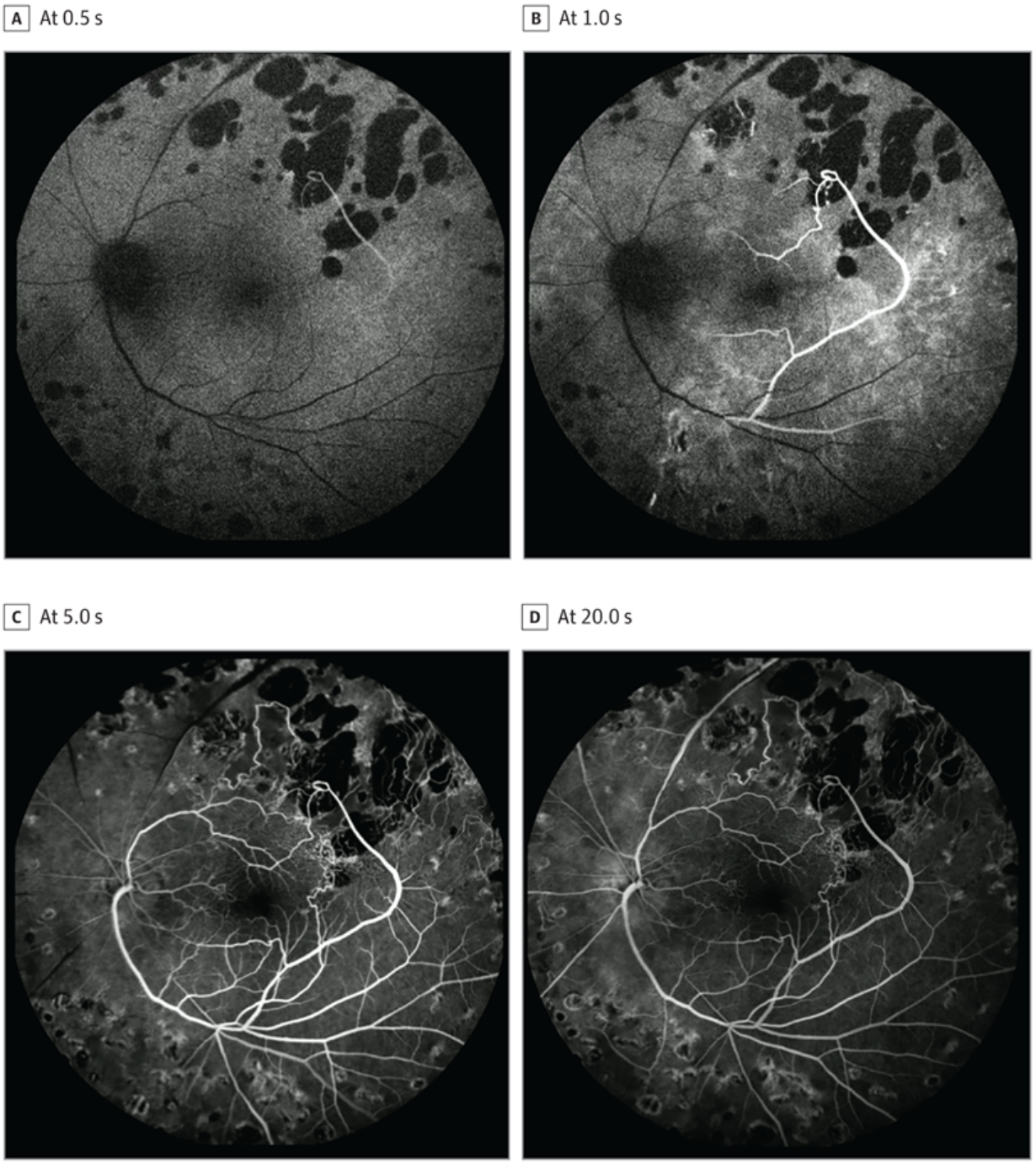
pigment epithelium in the direction of the retina and vascular endothelial growth factor production associated with retinal capillary nonperfusion.

Figure 1. Color Fundus Photography of the Left Eye Before and After Formation of the Chorioretinal Anastomosis



A and B, Marked thinning of the retinal arteries (asterisks) was observed compared with a visit 3 years earlier. A chorioretinal anastomosis had formed in a photocoagulation scar (arrowheads) interconnecting both arterial and venous vessels from the superior arcade and a venous vessel from the inferior arcade (dashed line markings). C, Corresponding to the white square in panel B, a volume-rendered optical coherence tomography angiography reconstruction revealed a spiral shape of the chorioretinal anastomosis (arrowheads).

Figure 2. Fluorescein Angiography Filling Sequence in the Left Eye



Fluorescein angiography sequence at 0.5, 1, 5, and 20 seconds showing a conspicuous, rapid initial filling of the temporal macular vasculature by retrograde circulation through the laser-induced chorioretinal anastomosis in the superotemporal macula (0.5-second and 1-second frames), which is sequentially followed by anterograde filling of the central retinal artery branches that brings perfusion to the nasal retina (5-second and

20-second frames).

Video. Fluorescein Angiography of the Left Eye at Baseline and at 3-Year Follow-Up

A, Fluorescein angiography at baseline had shown signs of chronic vascular abnormalities primarily in the superior retina, but an expected anterograde flow through the central retinal artery was preserved. B, Fluorescein angiography 3 years later revealed that the central and temporal macula was now being perfused by retrograde flow through a newly formed chorioretinal anastomosis located in a photocoagulation scar.

Discussion

Recent studies of larger cohorts have found the prevalence of retinal vascular abnormalities in individuals with NF1 to be 17% to 37%, according to various definitions.[2,3] Severe retinal vascular abnormalities in NF1 are considered rare,[4] but at least 13 previous case reports describe peripheral or central retinal vascular occlusion, neovascular glaucoma, or occlusion of the central retinal artery or ophthalmic artery. Like this case, mostly young patients with unilateral impaired arterial inflow and progressive retinal capillary nonperfusion are described. Preservation of choroidal perfusion in this patient suggests that vasculopathy was confined to arteries distally to the branching of the posterior ciliary arteries from the ophthalmic artery.

High-energy photocoagulation treatment can cause iatrogenic breaks in the Bruch membrane and facilitate the formation of choroidal neovascularization, which, under certain conditions, can develop into a laser-induced chorioretinal anastomosis (LICRA). McAllister[5] has proposed the use of this phenomenon

as a treatment for nonischemic central retinal vein occlusion (CRVO). The intended mode of action is to create auxiliary flow between the obstructed high-pressure retinal venous circulation and the unobstructed low-pressure choroidal venous circulation, thus normalizing retinal venous pressure. A trial including 58 selected patients with nonischemic CRVO who received intravitreal ranibizumab injections and were randomized to a LICRA procedure or sham found that the LICRA group had superior visual acuity and required fewer anti–vascular endothelial growth factor injections than controls after 2 years. However, only 2 in 3 attempts to create an anastomosis were successful, and procedure-related complications requiring secondary surgery occurred for every third patient.[5] Attempts to create LICRAs in eyes with ischemic CRVO have been mostly unsuccessful, perhaps due to severe endothelial cell damage secondary to ischemia and venous thrombosis across the retinal circulation.[6]

In this patient with NF1, an unintended, iatrogenic LICRA potentially salvaged the retina and preserved 20/20 visual acuity in an eye with severe, progressive retinal arterial occlusive disease. Here, the mechanistic mode of action of the LICRA differed from that of eyes with CRVO, in that the chorioretinal anastomosis causes inflow of oxygenated blood to the retina rather than outflow from congested retinal veins to the choroid. However, due to the year-long, extensive vascular remodeling seen in this case, more evidence is needed to suggest that therapeutic LICRAs can be created in patients with severe, progressive retinal arterial occlusive vasculopathy where choroidal perfusion is still intact.

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