Slowly Progressive Retinal Arteriovenous Malformation and Relative Amblyopia

Congenital retinal arteriovenous malformation is a rare, unilateral, nonhereditary disorder with variable visual impairment depending on the severity of the vascular anomaly. Most cases do not show any anatomical progression. Venous occlusion, intraretinal or vitreal hemorrhage, or optic atrophy are rare causes of sudden or gradual visual loss. We describe the case of a child with visual loss caused by slow progression of a foveal vascular loop and relative amblyopia.

Report of a Case. A 6-year-old boy had deteriorating vision 1 year after the incidental detection of a retinal arteriovenous malformation in his right eye (Figure 1). At the time of the initial detection, his visual acuity was 20/20 OU. One year later, his visual acuity was 20/60 OD and 20/20 OS. Multiple dilated and tortuous vessels emerged from the right optic disc (Figure 2) with 1 supertemporal loop reaching the fovea. No retinal thickening, exudates, bleeding, or relative afferent pupillary defects were present. Scanning laser ophthalmoscope–fundus perimetry showed an absolute scotoma overlying the foveal

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**Figure 1.** Fundus photograph taken in 1998. The central vascular loop was well separated from the foveola, and visual acuity reached 20/20 OU.

**Figure 2.** Fundus photograph taken in November 1999. The loop has reached the foveola. The overlay shows the retinal fixation areas when first seen in our clinic (March 1999; visual acuity of 20/60 OD), after successful occlusion therapy (November 1999; visual acuity of 20/30 OD), and at the final visit after therapy had been discontinued (October 2000; visual acuity 20/400 OD).
vascular loop. Fixation was unstable and 5° inferotemporal to the fovea (Figure 2). Comparison with the fundus photograph taken 1 year earlier showed some advancement of the loop.

The left eye was healthy. There was no strabismus, and stereopsis (Lang test) was normal. Cycloplegic refraction was +2.00 diopters spherical OU. The medical history, systemic history, and examination results were unremarkable. Recent magnetic resonance tomography results did not show any intracranial anomaly of Bonnet-Dechamp-Blanc or Wyburn–Mason syndrome.

Because the location of the loop itself did not account for the total amount of visual acuity loss, we suspected relative amblyopia and recommended occlusion of the left eye for 4 hours per day.

Eight months later, visual acuity had improved to 20/30 OD and scanning laser ophthalmoscope–fundus perimetry showed a definite central shift of the retinal fixation area (Figure 2). Esotropia of +10° without correction and +3° with correction was now present.

Afterward, occlusion therapy was difficult and finally abandoned by the parents. One year later, visual acuity was 20/400 OD. A further progression of the vascular loop and a 13° temporal shift of the fixation area were present (Figure 2). Because of that and the lack of success to maintain useful vision, we stopped any treatment.

Comment. The initial drop in visual acuity may have been due to 2 factors, a slow progression of the loop approaching the fovea and relative amblyopia. The progression of the loop toward the fovea was gradual and totaled 500 µm in 33 months (180 µm/y). Although most cases of arteriovenous malformation are thought to be stable,2 anatomical changes of arteriovenous malformations have previously been described3 but not linked to visual loss.

On the other hand, relative amblyopia is likely to have been a contributing factor. It is defined as a reversible functional deficit superimposed on a congenital organic abnormality or pathologic changes acquired in early childhood. Our hypothesis was supported by the increase in visual acuity with patching and the shift of fixation. When manifest esotropia was present, strabismic amblyopia may have contributed to the final visual loss.

Not all retinal AV malformations are stationary, and small changes can induce a visual loss that may be amplified by relative amblyopia in patients during their sensitive period of visual development. Unlike this patient, most cases do not progress. A trial of occlusion therapy may be warranted in young children if visual acuity is worse than expected from the location of a central loop.

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Report of a Case. A 20-year-old woman had complex retinal detachment and proliferative vitreoretinopathy in her right eye. She noted a deterioration in the vision of her right eye 1 day prior to hospital admission. Her ocular history was remarkable for a progressive impairment in the peripheral visual field during the past 18 months, but she had had no dilated eye examinations in the meantime. The function of the right eye had always been worse compared with the left eye, and it was thought to be amblyopic. On examination, visual acuity was hand movements OD and 20/20 OS. Slitlamp examination was unremarkable except for anterior vitreous opacities in the right eye. Funduscopy disclosed a subtotal retinal detachment with macular involvement in the right eye. There were subretinal fibrous strands in the inferior 2 quadrants and epiretinal membrane formation with star folds peripheral to the major vascular arcades consistent with a long-standing detachment and advanced proliferative vitreoretinopathy. While the optic disc in the left eye appeared healthy, there were 2 pits noted in the disc of the right eye. A small, rather subtle depression was located at the 8-o’clock position. A larger, well-developed, obvious declivity, the bottom of which was not visible, was at the nasal disc margin (Figure 1). No diaphanous tissue was identified overlying any of the pits.

Subsequently, pars plana vitrectomy, removal of subretinal and