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CONGENITAL MACROVESSEL ASSOCIATED WITH CYSTOID MACULAR EDEMA AND AN IPSILATERAL INTRACRANIAL VENOUS MALFORMATION

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Background/Purpose: To report a case of congenital retinal macrovessel associated with cystoid macular edema and an ipsilateral intracranial venous malformation.

Methods: Case report.

Results: A 58-year-old woman with decreased vision was found to have a congenital retinal venous macrovessel associated with cystoid macular edema because of tributary venous occlusion. The patient underwent neuroimaging and an ipsilateral venous malformation of the frontal lobe was discovered.

Conclusion: Congenital retinal macrovessel can occasionally be complicated by vascular occlusion and macular edema. The authors report a case of congenital retinal macrovessel associated with an intracranial venous malformation. Clinicians should be aware of this potential association, and further studies are warranted.

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Congenital retinal macrovessels are large, aberrant blood vessels, which cross the horizontal raphe and are believed to originate from abnormal embryologic development during the 15th and 16th weeks of gestation.[1] Most of these malformations are venous, unilateral, asymptomatic, and incidentally discovered. Rarely, complications may develop resulting in decreased visual acuity.[1–4]

In 1972, Archer et al[5] classified retinal arteriovenous malformations into 3 categories with Type I arteriovenous malformations defined as a retinal macrovessel associated with an abnormal intervening capillary bed between the arterial and venous vasculature. Although Archer et al considered retinal macrovessels to be a variant of racemose angiomatosis or the Wyburn–Mason syndrome, to date there have been no cases reported of intracranial arteriovenous malformations in association with a macrovessel. Congenital retinal macrovessels are therefore considered to be isolated vascular malformations of the eye. Here, we report a case of macrovessel complicated by macular edema and associated with an ipsilateral intracranial venous malformation. To our knowledge, this is the first reported case of a CRM associated with an intracranial vascular malformation.

Case Report

A 58-year-old woman with medical history of hypertension and asthma presented complaining of recent onset decreased visual acuity in the left eye. Her best-corrected visual acuity was 20/20 in the right eye and 20/40 in the left eye. Anterior segment examination was

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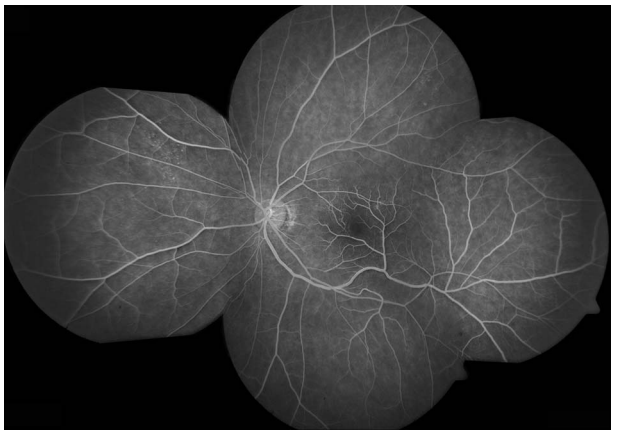

Fig. 1. Fluorescein angiogram montage showing the course of the large abnormal macrovessel as it crosses the horizontal raphe. Note the absence of abnormal arterial–venous communications.

unremarkable. Funduscopic examination of the right eye was also unremarkable. Examination of the macula of the left eye revealed a large inferior macrovessel crossing the horizontal raphe adjacent to the fovea (Figure 1). One of the associated branches appeared sheathed. Fluorescein angiography showed early filling and delayed emptying of the venous macrovessel, and poor filling and possible occlusion of the sheathed tributary consistent with a small branch retinal vein occlusion (Figure 2). Capillary bed abnormalities and late angiographic edema involving the central macula were also noted (Figure 3). Spectral domain optical coherence tomography confirmed the presence of severe cystoid macular edema (Figure 4).

A magnetic resonance imaging of the brain was obtained, which revealed an ipsilateral venous malformation within the left frontal lobe (Figure 5). The patient was observed without treatment for 2 months. At the 2-month follow-up, the patient's vision had returned to 20/20 in the left eye. Repeat fundoscopy, spectral domain optical coherence tomography, and fluorescein angiogram showed resolution of the vascular sheathing and cystoid macular edema and reperfusion of the occluded tributary.

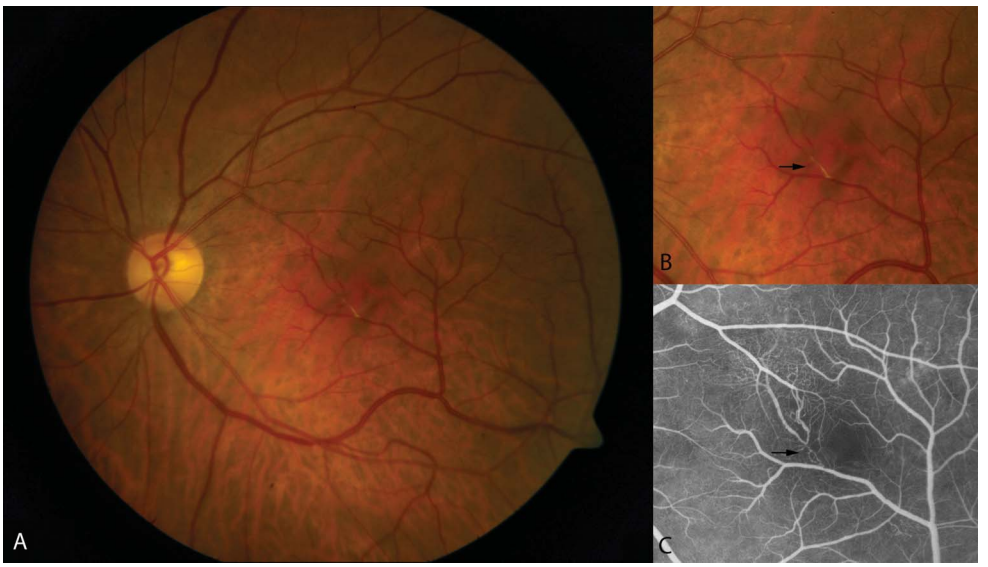

Fig. 2. A. Fundus photograph at presentation (**A**) and a magnified view of the macula (**B**) showing the abnormally sheathed venous tributary (arrow), and magnified fluorescein angiogram of the macula (**C**) showing decreased flow through the sheathed tributary (arrow) and capillary bed abnormalities associated with the macrovessel.

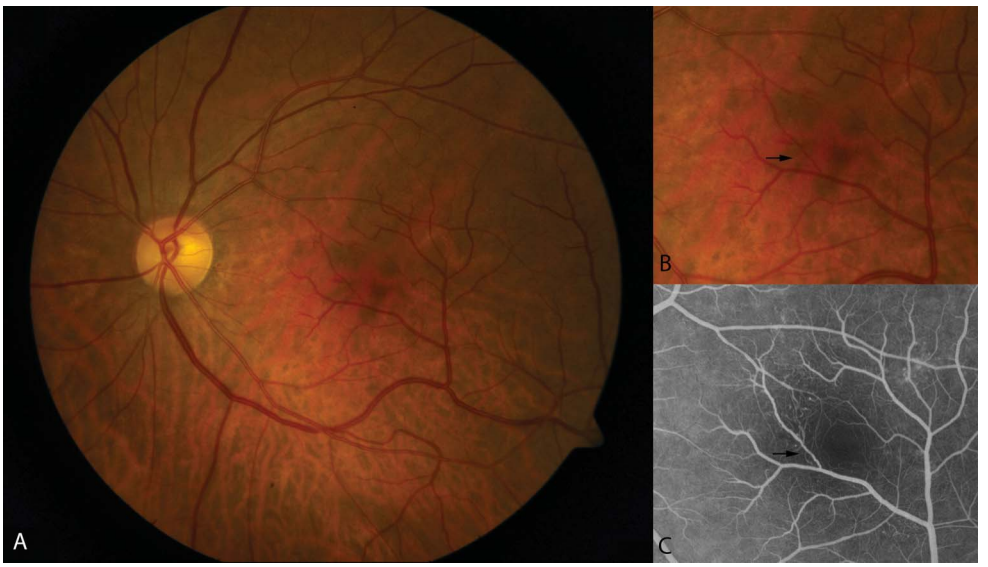

Fig. 3. A. Fundus photograph at the 2-month follow-up visit (**A**) and a magnified view of the macula (**B**) showing resolution of the venous sheathing (arrow) and magnified fluorescein angiogram of the macula (**C**) showing improved flow through the previously sheathed tributary (arrow).

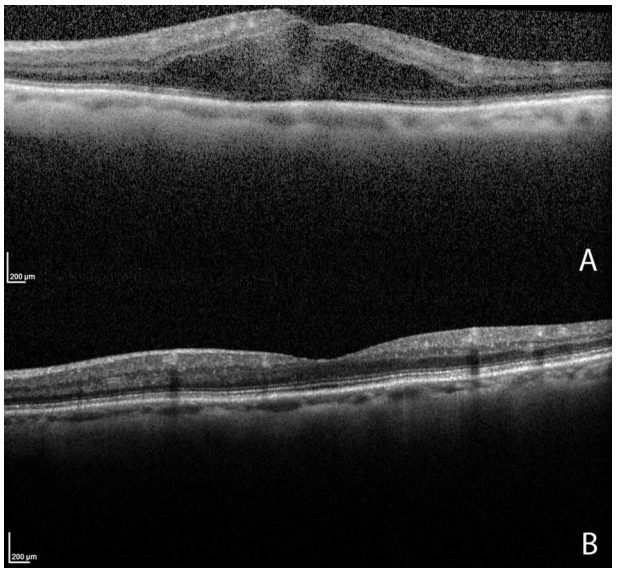

Fig. 4. A. Spectral domain optical coherence tomography at presentation showing severe cystoid macular edema. **B.** Spectral domain optical coherence tomography at 2-month follow-up showing resolution of cystoid macular edema.

Discussion

Congenital retinal macrovessels are anomalous vessels, which result from abnormal embryogenesis. For the most part, these lesions are considered to be isolated and nonvision threatening. Rarely decreased visual acuity can result from a variety of etiologies including macular hemorrhage or serous detachment, macular ischemia, branch retinal artery occlusion, or angioscotoma.[1–3,6–9] In our case, decreased visual acuity developed from cystoid macular edema likely complicating an occluded venous tributary branching off the macrovessel. Macular edema has previously been reported in association with a macrovessel because of microvascular alterations in the blood-retina barrier.

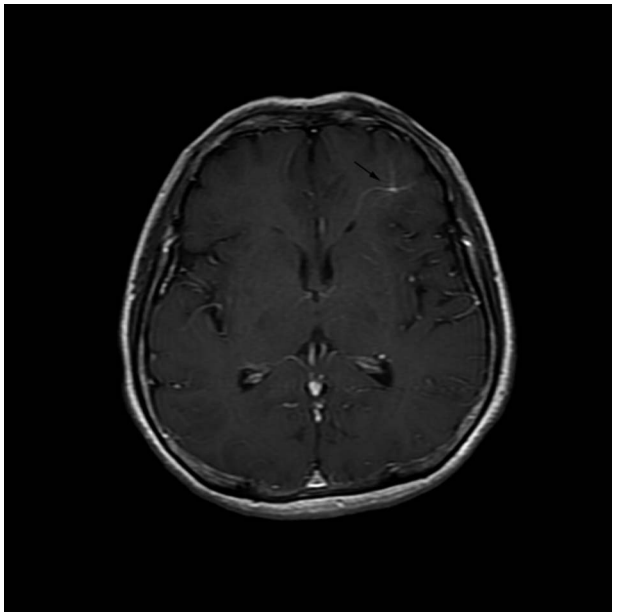

Fig. 5. Magnetic resonance imaging with contrast of the brain showing a venous malformation in the left frontal lobe (arrow).

Our patient's abnormality most closely fits with the Archer classification of Type I arteriovenous malformations. Archer et al[5] considered this to be a mild variant of racemose angiomatosis. Because of this theoretical association, we elected to pursue neuroimaging, which revealed an ipsilateral venous malformation within the left frontal lobe. To our knowledge, this is the first reported case of a congenital retinal macrovessel associated with any type of systemic vascular malformation. Although this association may be a coincidental finding, it should be noted that the retinal and cerebral lesions were on the same left side, were both congenital venous anomalies, and shared a similar structural morphology with imaging. Given the similar anatomical findings of the two lesions in the retina and the brain, one wonders whether the retinal lesion would be better described as a congenital retinal venous malformation.

Key words: congenital retinal macrovessel, macular edema, venous anomaly, retinal arterial–venous malformation.

References

1. de Crecchio G, Alfieri MC, Cennamo G, Forte R. Congenital macular macrovessels. Graefes Arch Clin Exp Ophthalmol 2006;244:1183–1187.
2. Arai J, Kasuga Y, Koketsu M, Yoshimura N. Development and spontaneous resolution of serous retinal detachment in a patient with a congenital retinal macrovessel. Retina 2000;20:674–676.
3. Goel N, Kumar V, Seth A, Ghosh B. Branch retinal artery occlusion associated with congenital retinal macrovessel. Oman J Ophthalmol 2014;7:96–97.
4. Petropoulos IK, Petkou D, Theoulakis PE, et al. Congenital retinal macrovessels: description of three cases and review of the literature. Klin Monbl Augenheilkd 2008;225:469–472.
5. Archer DB, Deutman A, Ernest JT, Krill AE. Arteriovenous communications of the retina. Am J Ophthalmol 1973;75: 224–241.
6. Beatty S, Goodall K, Radford R, Lavin MJ. Decompensation of a congenital retinal macrovessel with arteriovenous communications induced by repetitive rollercoaster rides. Am J Ophthalmol 2000;130:527–528.
7. de Crecchio G, Pappalardo G, Pascotto F, Forte R. Aberrant congenital macular vessel crossing the fovea: evaluation with optical coherence tomography. Eye 2006;20:1464–1466.
8. Makino S, Endoh K, Tampo H. Retinal microvascular abnormalities in neurofibromatosis type 1 associated with congenital retinal macrovessels. Case Rep Ophthalmol Med 2013;2013:604191.
9. Shah VA, Chalam KV. Congenital retinal macrovessel causes reduced retinal sensitivity at the macula. Eur J Ophthalmol 2004;14:341–344.