DOI: 10.4274/tjo.59837
Turk J Ophthalmol 2018;48:52-55

Case Report

Three Cases of Congenital Retinal Macrovessel, One Coexisting with Cilioretinal Artery

Bayram Gülpamuk, Pınar Kaya, Mehmet Yasin Teke
Ulucanlar Eye Training and Research Hospital, Ophthalmology Clinic, Ankara, Turkey

Abstract

In this report, we describe three cases of retinal macrovessel. Two of the three patients presented to our clinic for annual eye exam and had no visual complaints. The third patient presented because of vision loss in the left eye. Two patients had 20/20 best corrected visual acuity in both eyes and the third patient had 20/20 in the right eye, 20/25 in the left eye. Pupillary exams were normal. Slit-lamp examinations of the anterior segment were unremarkable. Fundus examination revealed macrovessels in the left eyes of two patients and in the right eye of one patient. The patients underwent complete ophthalmological examinations including color fundus photography for all three patients and optic coherence tomography, fundus autoflorescence, and fundus fluorescein angiography for two of the patients. Cilioretinal artery coexisting with macrovessel was seen angiography in one case. Congenital retinal macrovessel is a rare vascular condition. It is often unilateral and the vessel is an aberrantly large branch of the retinal arteries or veins. They may cross the fovea and their visual impact is minimal. The coexistence of congenital retinal macrovessel and cilioretinal artery is very rare. Visual impairment may occur in congenital retinal macrovessel due to retinal cavernous hemangioma, foveal cysts, central serous retinopathy, and other retinal vascular abnormalities.

Keywords: Congenital retinal macrovessel, aberrant retinal vessels, cilioretinal artery

Introduction

Mauthner[1] first reported a large aberrant retinal vessel crossing the macula in 1869. In 1982, Brown et al.[2] described the clinical and fluorescein angiographic features of congenital retinal macrovessel (CRM) in seven patients. Impairment of vision in the involved eye is uncommon and is characterized by foveal cyst, macular hemorrhage, serous macular detachment, branch retinal artery occlusion or other vascular abnormalities.[3] CRM occurs mainly in veins but more rarely may stem from an artery or artery and vein together.[4] Beatty et al.[5] in this case report we present a cilioretinal artery connecting with a CRM, suggesting that such patients are at increased risk of retinal vascular decompensation. Herein, we present three cases showing no vision loss in routine ophthalmological examination, and interestingly, one patient had both CRM and cilioretinal artery.

Case Reports

Case 1: An 8-year-old healthy female patient applied to our ophthalmology department for routine ophthalmic evaluation. Her personal and family medical histories were unremarkable. On ophthalmic examination, her best-corrected visual acuity was 20/20 in both eyes. Anterior segment examination of both eyes was normal. Intraocular pressures were within normal limits. Fundus examination was normal in the left eye but revealed a large macrovessel crossing the horizontal raphe adjacent to the fovea in the right eye (Figure 1). The patient was evaluated only with fundus photography because the patient's family did not consent to fundus fluorescein angiography (FFA) and optical coherence tomography (OCT).

Case 2: A 6-year-old female patient was brought to us due to reduced vision in the right eye. In our ophthalmic examination, we detected astigmatism in the right eye but best-corrected

Address for Correspondence: Pınar Kaya MD, Ulucanlar Eye Training and Research Hospital, Ophthalmology Clinic, Ankara, Turkey
Phone: +90 506 538 71 59 E-mail: drpnrcck@gmail.com **ORCID-ID:** orcid.org/0000-0001-9243-6124
Received: 13.02.2017 **Accepted:** 23.05.2017

visual acuity (BCVA) was 20/20 in both eyes using a Snellen chart. Anterior segment and funduscopic examination of the right eye were unremarkable. Examination of the macula of the left eye revealed a large superior macrovessel crossing the horizontal raphe with several tributaries adjacent to the fovea. Furthermore, the abnormal vein was accompanied by a cilioretinal artery (Figure 2a). The patient was evaluated with fundus photograph, FFA, fundus autofluorescence (Figure 2b), and spectral domain (SD)-OCT. FFA showed early filling of the venous macrovessel, accompanied by a cilioretinal artery, crossing the macula and having three tributaries which are surrounding the foveal area (Figure 2c). SD-OCT (Heidelberg Engineering, Heidelberg, Germany) showed normal foveal contour and vessel shadowing at five points (Figure 2d).

Case 3: A 16-year-old male patient was referred to us with a history of blurred vision in the left eye. His BCVA was 20/20 in the right eye and 20/25 in the left eye on Snellen chart. Relative afferent pupillary defects and anisocoria were not present. Intraocular pressures were within normal limits. Slit-lamp examinations of the anterior segments of both eyes were normal. On fundus examination of the left eye, an anomalous large vessel was seen passing through the fovea separated in the optic disc from the inferotemporal vein. The patient was evaluated with colored fundus photograph (Figure 3a), SD-OCT (Figure 3b), FFA (Figure 3c) and fundus autoflorescence (Figure 3d).

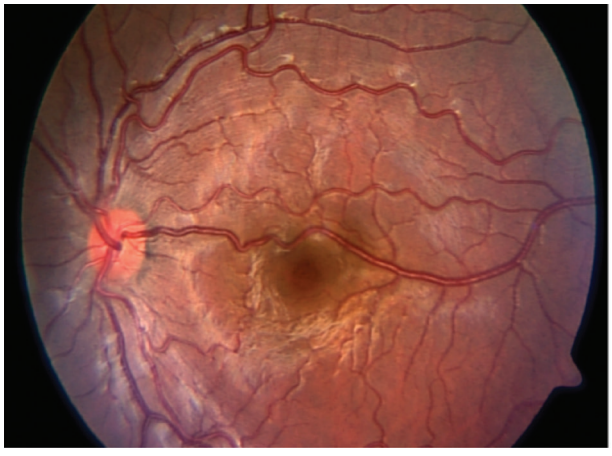

Figure 1. Case 1: fundus photograph at presentation showed a large macrovessel is passing superior to the fovea extending along the papillomacular bundle and showing tortuosity of the vessels

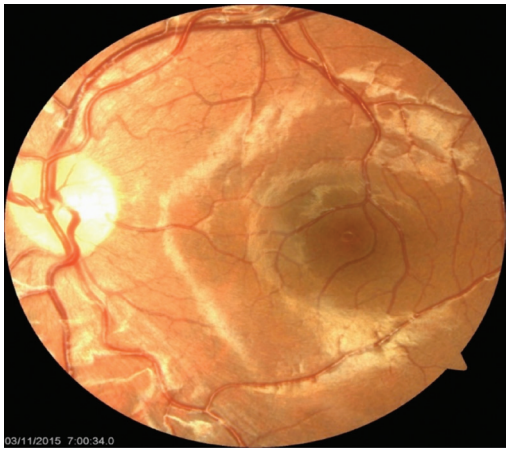

Figure 2a. Case 2: fundus photograph at presentation showed retinal macrovessel was a branch of the superotemporal vein reaching up to the fovea with three tributaries accompanied by a cilioretinal artery

Discussion

Congenital retinal macrovessel is a rare finding and is usually discovered incidentally. CRM are mesenchymal in origin and develop around the first weeks of the second trimester when

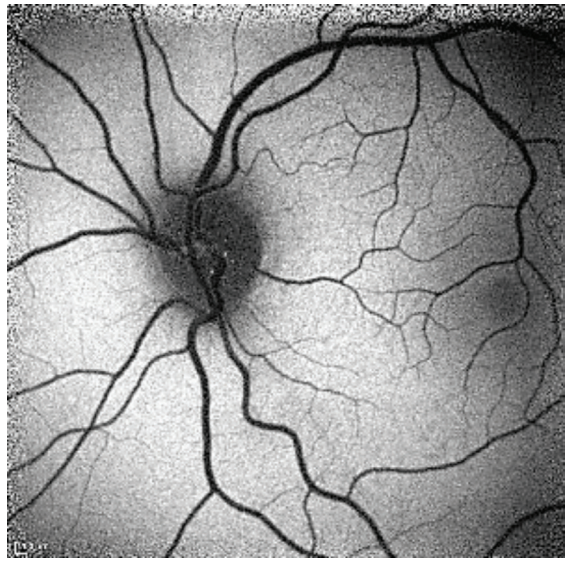

Figure 2b. Case 2: fundus autofluorescence at presentation

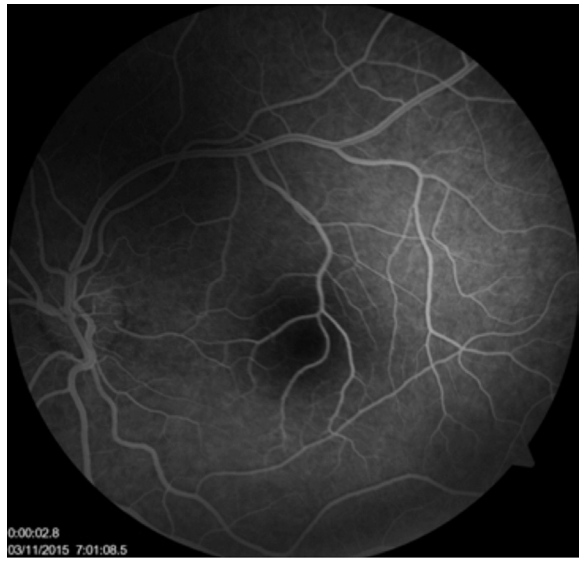

Figure 2c. Case 2: farly filling of the aberrant retinal macrovein was observed on fundus fluorescein angiography

differentiation of arteries and veins occurs.[6] They are generally asymptomatic, and vision is not affected in most cases. Archer et al.[7] classified congenital retinal arteriovenous communications into three groups. Group 1 arteriovenous communications are the mildest variant, and clinically, can be very subtle. Group 2 are larger than those of group 1. Our case 2 was compatible with group 1 and our cases 1 and 3 were compatible with group 2 of the Archer classification. To our knowledge, a congenital retinal venous macrovessel that communicates with a cilioretinal artery is very rare. This condition was first described by Beatty et al.[5] Most of the cases of CRM that have been documented to date exhibited normal visual acuity.[2] When macrovessel is associated with reduced vision, one of the rare conditions should be considered: foveal cyst, macular hemorrhage or serous detachment, macular ischemia, branch retinal artery occlusion, and Valsalva retinopathy.[8] For this reason, clinicians should be vigilant and follow these patients regularly.

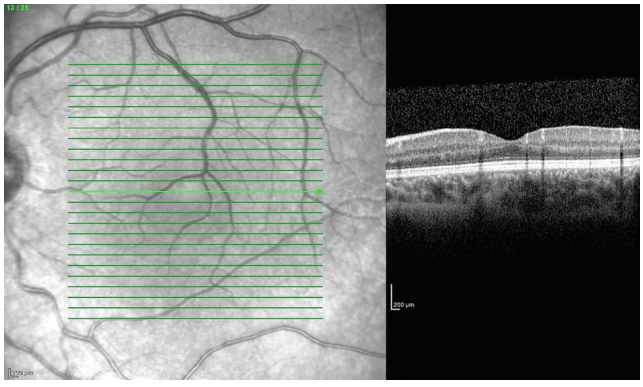

Figure 2d. Case 2: spectral domain optical coherence tomography showing normal foveal contour and vascular shadowing at five points (arrows)

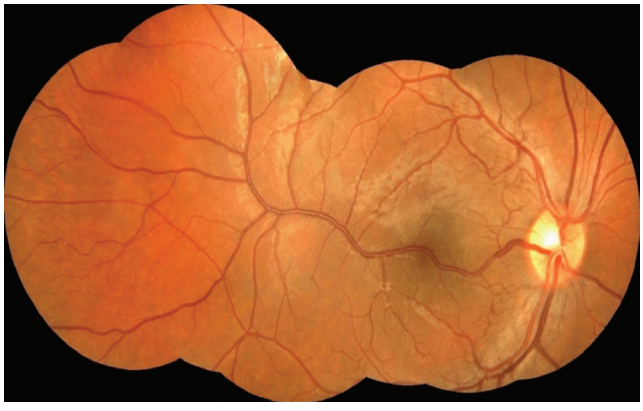

Figure 3a. Case 3: montage colored fundus image showing a congenital retinal macrovessel crossing the macula horizontally in the left eye and separating branches of the vein peripherally

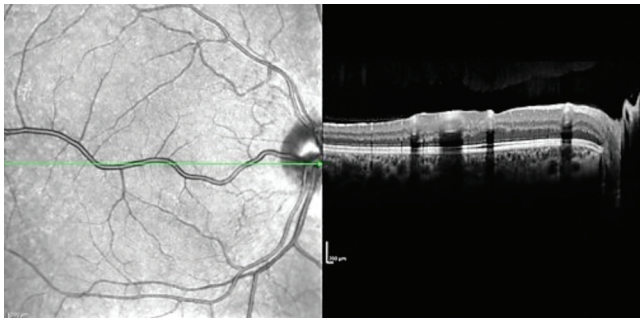

Figure 3b. Case 3: spectral domain ocular coherence tomography at presentation in the left eye with horizontal section passing through the macrovessel

Ethics

Informed Consent: It was taken.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Mehmet Yasin Teke, Concept: Bayram Gülpamuk, Design: Bayram Gülpamuk,

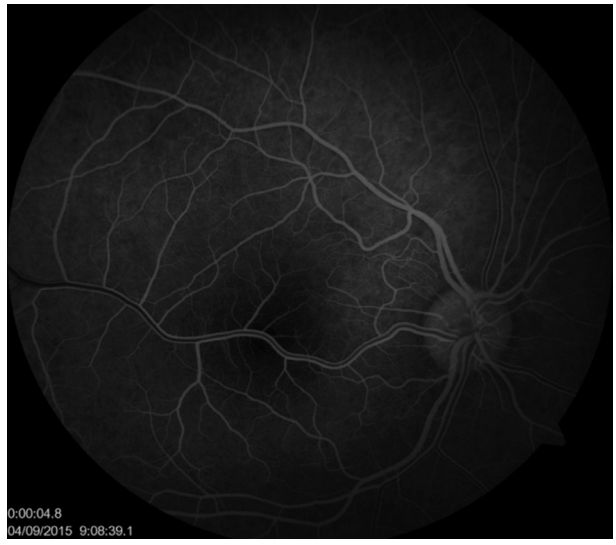

Figure 3c. Case 3: fundus fluorescein angiography at presentation showed late filling of macrovessel simultaneously with inferotemporal vein

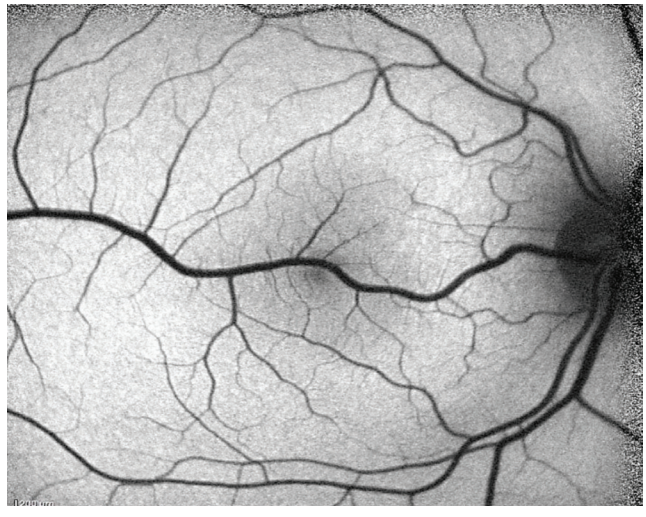

Figure 3d. Case 3: left eye fundus autofluorescence image

Data Collection or Processing: Bayram Gülpamuk, Pınar Kaya, Analysis or Interpretation: Bayram Gülpamuk, Pınar Kaya, Literature Search: Bayram Gülpamuk, Pınar Kaya, Writing: Bayram Gülpamuk, Pınar Kaya.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Mauthner L. Lehrbuch der ophthalmoscopie. Tendler; Vienna;1869:249.
2. Brown GC, Donoso LA, Magargal LE, Goldberg RE, Sarin LK. Congenital retinal macrovessels. Arch Ophthalmol. 1982;100:1430-1436.
3. de Crecchio G, Alfieri MC, Cennamo G, Forte R. Congenital macular macrovessels. Graefes Arch Clin Exp Ophthalmol. 2006;244:1183-1187.
4. Sanfilippo CJ, Sarraf D. Congenıtal macrovessel associated wıth cystoid macular edema and an ipsilateral intracranial venous malformation. Retin Cases Brief Rep. 2015;9:357-359.
5. 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.
6. Bhatia HK, Sharma S, Laxminarayana P. Congenital Retinal Macrovessel with Normal Visual Acuity: A Case Report. Int J Ophthalmol Clin Res 2015;2:2-4.
7. Archer DB, Deutman A, Ernest JT, Krill AE. Arteriovenous communications of the retina. Am J Ophthalmol. 1973;75:224-241.
8. Goel N, Kumar V, Seth A, Ghosh B. Branch retinal artery occlusion associated with congenital retinal macrovessel. Oman J Ophthalmol. 2014;7:96-97.