A Patient with Congenital Retinal Macrovessel and Prepapillary Vascular Loop Together at The Same Eye

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ABSTRACT

In this article, we reported a patient with two rare retinal vascular anomalies together; congenital retinal macrovessel (CRM) and prepapillary vascular loop. We diagnosed this two rare anomalies incidentally during an eye examination in the right eye of a 70 years old male patient. Best-corrected visual acuity (BCVA) was 20/20 in both eyes. There were no signs of complications. Fundus fluorescein angiography (FFA) of the right eye revealed an abnormal artery. At optic coherence tomography (OCT) of the right eye, abnormal vessel was seen at fovea. Neurological examination, brain and orbital magnetic resonance imaging (MRI) and MRI angiography of the brain and the orbit were normal. CRMs, where an aberrant retinal vessel is present in the posterior pole and may cross the avascular foveal region and prepapillary vascular loop formations are seen rarely. Before our case only one case in the literature has been reported in the same condition.

Keywords: Congenital retinal macrovessel, Prepapillary vascular loop, Retina.

INTRODUCTION

Congenital retinal anomalies of the vessels are rare developmental vascular anomalies. Hereditary retinal artery tortuosity, inherited retinal venous beading, hereditary hemorrhagic telangiectasis, congenital prepapillary vascular loops, congenital retinal macrovessels (CRMs), and arteriovenous communications are various types of congenital anomalies of retinal vessels.[1] Some of these anomalies involve the disc and the surrounding area; others may involve the entire vascular tree.

We reported 70 years old male patient who has a peculiar type of retinal vascular anomaly in which the involved retinal vessel is the main branch of central retinal vessels that show looped or coiled patterns around or near the optic disc and another vascular anomaly, CRM, at the same eye together.

CASE REPORT

We detected 70 years old male patient who has CRM at foveal avascular zone and prepapillary vascular loop at optic disc area in the right eye during routine eye examination.

Best corrected visual acuity (BCVA) was 20/20 in both eyes. Examination of anterior segment was normal in both eyes. There was no afferent pupillary defect or anisocoria.

Intraocular pressure was 13 mmHg in the right eye and 15 mmHg in the left eye.

Examination of posterior segment revealed an abnormal superior retinal artery which was exiting from the root of central retinal artery and crossing foveal avascular zone (Image 1). In this patient, we also detected a coiled peripapillary retinal vessel, prepapillary vascular loop, which was located at the inferior branch of the central retinal artery (Image 1). Both of this vascular anomalies were abnormal retinal arteries. There was tortuosity of small vessels near prepapillary vascular loop at the temporal region of the optic disc. There were no signs of hemorrhage, intraretinal exudate, oedema or foveolar cysts. Fundus Fluorescein Angiography (FFA) of the right eye revealed an abnormal artery which showed early

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filling and delayed emptying (Image 1 and Image 2). There was no leakage or ischemia due to the abnormal vessel, only retinal pigment epithelium (RPE) alterations were detected (Image 1 and Image 2). At Spectral Domain Ocular Coherence Tomography (OCT) of the right eye, we detected this abnormal vessel at fovea on a few sections. (Image 3).

Fundus examination of the left eye was normal. FFA and OCT images of the left eye were normal except RPE alterations (Image 4 and Image 5).

Our patient had no neurological complaint and neurological examination was normal. Both the brain and the orbital Magnetic Resonance Imaging (MRI) were normal, also there were no vascular anomalies detected at MRI angiography of the brain and the orbit. There was no systemic disease accompanying these anomalies except systemic hypertension which was under control with medical therapy for ten years.

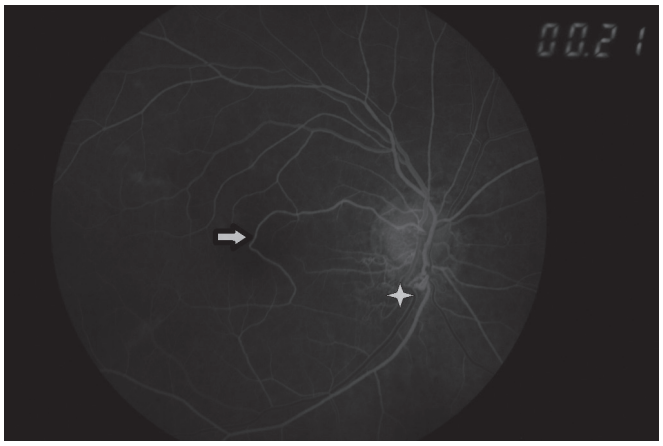

Image 1. *Early Stages of FFA. Early filling of macrovessel and prepapillary vascular loop, RPE alterations' window defects.*

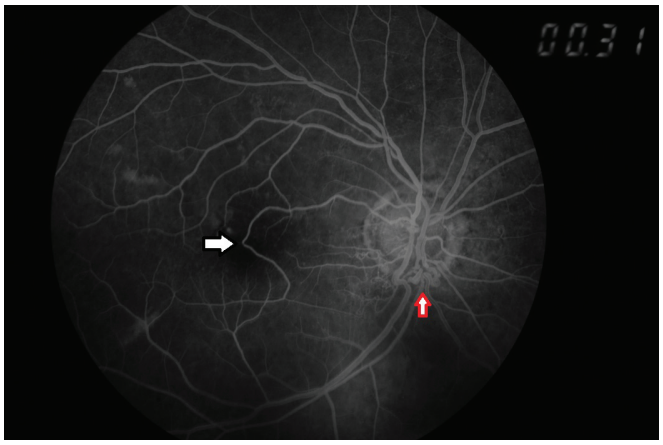

Image 2. *FFA of the right eye, the venous phase of angiography started, macrovessel's delayed emptying, hyperfluorescent areas of window defects due to RPE alterations.*

All the supplementary tests such as Amsler's grid, Ishihara color test and Humprey 10-2 visual field test were normal in both eye.

DISCUSSION

Mauthner was first to describe abnormal retinal vessel in the area of the macula at 1869.[2] The presence of CRMs, a phenomenon first described by Brown et al in 1982.[3] CRMs are rare with an estimated prevalence of one in 200.000 individuals.[4] The diagnosis of them is usually incidental because their visual impact is minimal. Visual impairment may result if the anomalous vessel pass across the foveola,

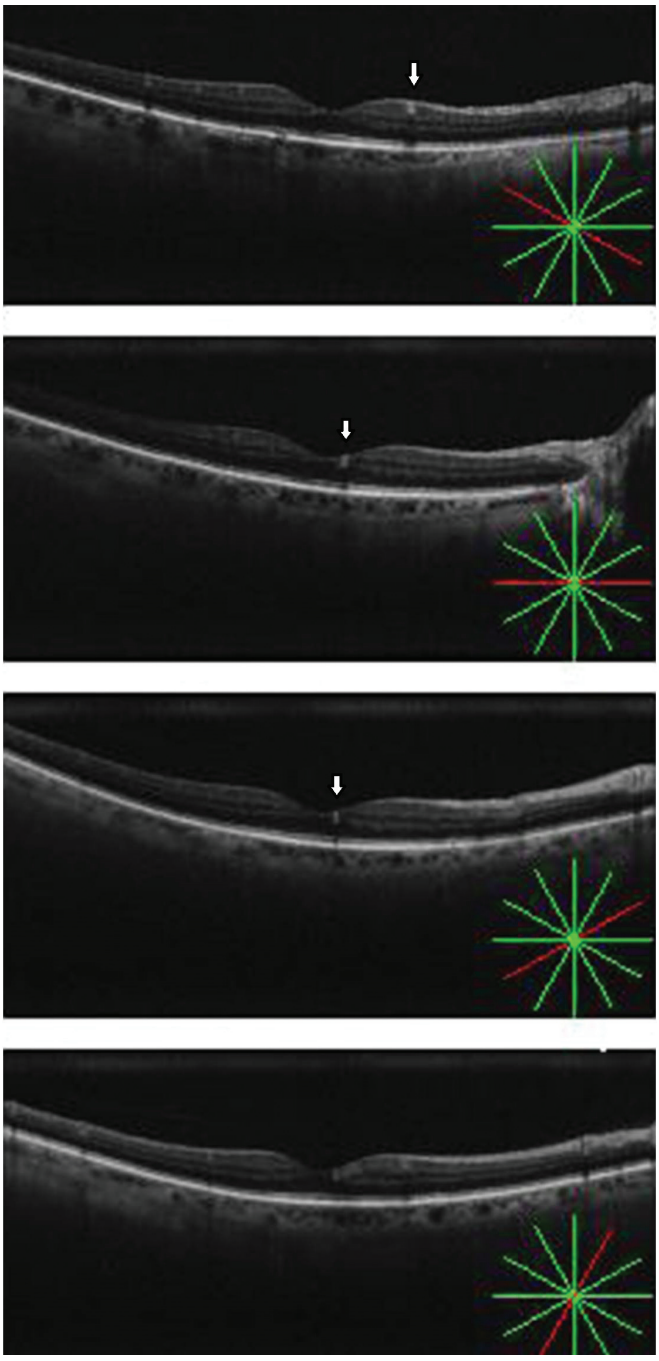

Image 3. *OD macular OCT sections.*

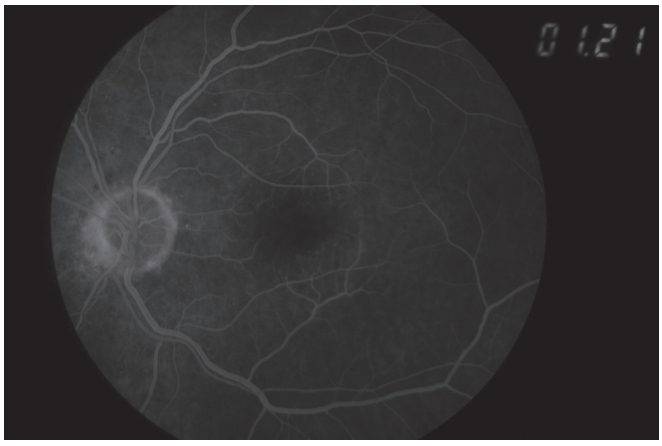

Image 4. F*FA of the left eye.*

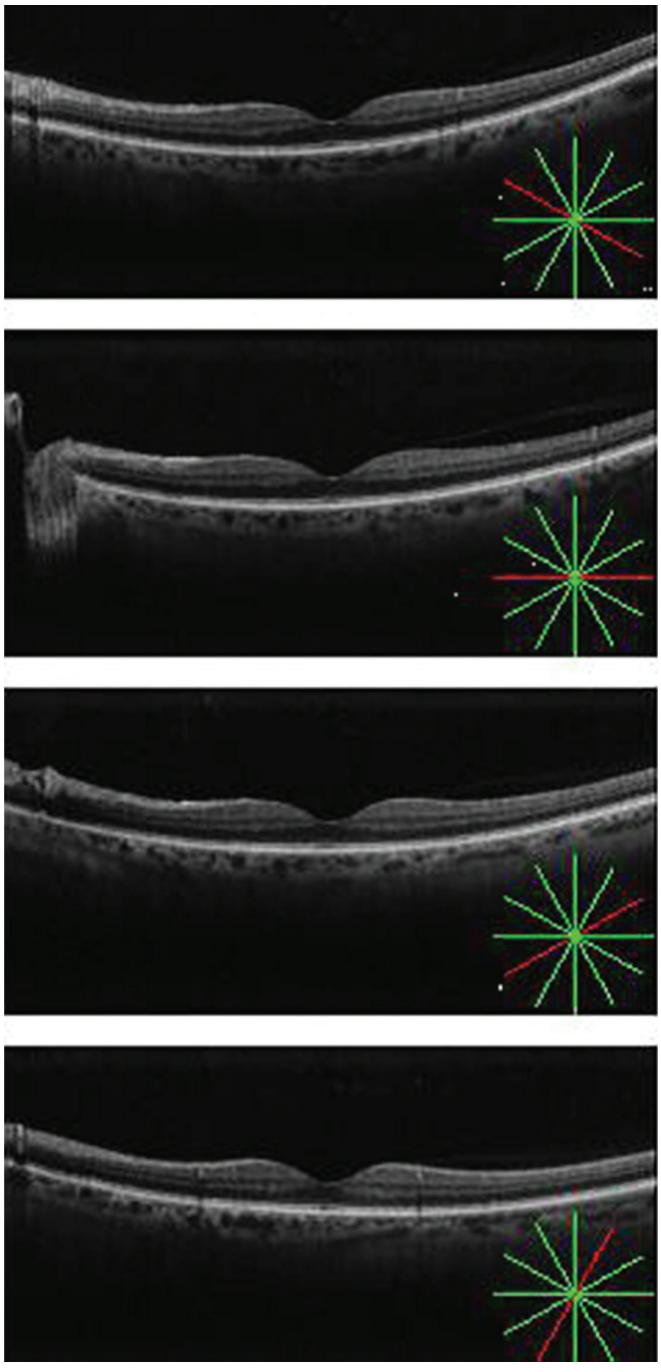

Image 5. *OS macular OCT sections.*

foveolar cysts form and if macular hemorrhages or serous macular detachments happen because of the abnormal vessel.[3,5,6] In our case, we detected anomalous vessel passing across the foveola at some sections of macular OCT but there was no visual impairment detected.

CRMs are mostly unilateral, single veins.[3] These vessels may supply or drain blood from both inferior and superior retina. Fewer cases have been reported of arteriolar origin as in our case.

The occurrence of central serous chorioretinopathy with CRM has been reported in few studies.[7,8] In our case, we detected multiple RPE alterations without subretinal fluid and that may be resulted from multiple attacks of central serous chorioretinopathy in the past.

The decompensation of macrovessel has been associated with the alteration of pressure (Valsalva maneuver) or gravitational forces (roller-coaster rides or bungee jumping).[9,10] Branch retinal artery occlusion occurring as a result of ischemic decompensation of CRM has also been discussed.[11] Amblyopia can occur in children by these vessels crossing fovea.[12] None of them detected in our case.

There also exists a possibility of brain vascular anomalies like WyburnMasson syndrome. In our case neurological examination, structural and vascular MRI were normal. Cavernous hemangiomas have been found to coexist with macrovessels.[13] Microvascular anomalies with CRMs have been reported in a case of neurofibromatosis.[14] In our case we found CRM coexist with prepapillary vascular loop.

Prepapillary vascular loop formations are uncommon congenital vascular malformations, like CRMs they are usually detected incidentally at routine fundus examination. The majority of them are of arterioral origin.[15] The incidence of prepapillary vascular loops ranges from 1 in 2000 to 1 in 9000 patients and most of these cases are unilateral.[16] In our case, it was also unilateral.

Prepapillary vascular loops have been reported to be associated with branch retinal artery occlusion (BRAO), amaurosis fugax, recurrent vitreous hemorrhage, subretinal hemorrhage, and hyphema.[15] In our case, there were no signs of this conditions.

Abnormal vessel development is the most popular theory to be the cause of congenital macrovessels. In 1969, Ashton described vessel development and related it to the formation of CRMs.[17] During 15th to 16th prenatal week, mesodermal mesenchymal cells appear near the hyaloid artery on the disc. These cells form the vascular system that will replace the embryonic blood supply of the hyaloid system. During the migration, formation of

cords and canalization process it is possible that a vessel might enlarge to a greater extent than others and also assume an abnormal retinal position. The exact time and triggering mechanisms for this phenomenon are unknown. The embryologic origin of prepapillary vascular loops is uncertain like CRMs. For some of the authors the origin of prepapillary vascular loops is the hyaloid system. Shakin et al. described an anatomic study of prepapillary vascular loop.[18] The loop communicated with the retinal arterial system and did not have an internal elastic lamina and also the connective tissue matrix of the vascular loop contained less hyaluronic acid than the vitreous. Their findings seem to support the embryologic deviation of prepapillary vascular loops from the retinal arterial system instead of the hyaloid system.

Our case report is strengthening the hypothesis of Shakin and Ashton. These two rare anomalies detected together in our case and the same embryological origin-retinal arterial system is possible. To our knowledge before our case, prepapillary vascular loop and CRM together has been reported only in one case by Saltou et al.[19]

In conclusion, careful observation and monitoring of patients with prepapillary vascular loops is necessary because of the possibility of complications such as macroaneurysm rupture, BRAO, and vitreous hemorrhage. Visual acuity evaluation, fundus examination and photography, FFA, OCT are important for both diagnosis and follow-up of congenital retinal macrovessels for decompensation.

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