

# Congenital Retinal MacrovesSEL

## Konjenital Retinal Makrodamar

Cemile ÜÇGÜL ATILGAN<sup>1</sup>, Dilek GÜVEN<sup>2</sup>

### Case Report

### Olgu Sunumu

#### ABSTRACT

Congenital retinal macrovesSEL is a rare condition in which anomalous retinal vessels cross the macula. Visual impairment may result if the anomalous vessel pass across the foveola, if foveolar cysts form, or if hemorrhage occurs. A 13 year-old male patient has an anomalous retinal vein that traversed the horizontal raphe without involving the foveola. In this case report, we present the fundus fluorescein angiography, optical coherence tomography evaluations of this patient with classical characteristics of congenital abnormal macrovesSEL and normal vision.

**Key Words:** Congenital abnormal macrovesSEL, macula, optical coherence tomography.

#### ÖZ

Konjenital retinal makrodamar, maküladan geçen anormal retinal damarlar olarak bilinen nadir görülen bir hastalıktır. Anormal damar eğer foveoladan geçerse, foveolar kist formu oluşturursa, hemoraji meydana gelirse görme bozukluğu ile sonuçlanabilir. Bu makalede foveola hariç, horizontal rafeyi geçen anormal retinal vene sahip 13 yaşındaki bir erkek hastadan bahsedilmektedir. Görmesi etkilenmemiş olan ve konjenital makrodamarın karakteristik özelliklerini sergileyen bu hasta, anjiyografi ve optik koherens tomografi ile değerlendirilmiştir.

**Anahtar Kelimeler:** Konjenital retinal makrodamar, maküla, optik koherens tomografi.

*Ret-Vit 2011;19:274-277*

#### INTRODUCTION

In 1869, Mauthner was one of the first to describe aberrant retinal vessel in the area of the macula.<sup>1</sup> There are several reports in the literature concerned with such anomalous congenital retinal vessels.<sup>2-7</sup> Brown et. al suggested that large congenital vessels that cross the horizontal raphe and have minimal effect on the visual acuity can be referred to as congenital retinal macrovesSEL.<sup>4</sup>

Blood supply of inner retinal layers is provided by central retinal artery other than cilioretinal artery which is present in 15-20% of general population. The distribution and course of retinal blood vessels may be different in every human and even the fellow eye of the same person. Retinal artery with three branches, cross integration of two similar vessels of the retina or abnormal course of a retinal vein are visible retinal vascular abnormalities.<sup>9</sup>

In this article, a patient with vascular anomaly that incidentally was diagnosed during routine eye examination is evaluated with fundus fluorescein angiography and optical coherence tomography.

**Geliş Tarihi : 21/10/2010**

**Kabul Tarihi : 11/03/2011**

**Received : October 21, 2010**

**Accepted : March 11, 2011**

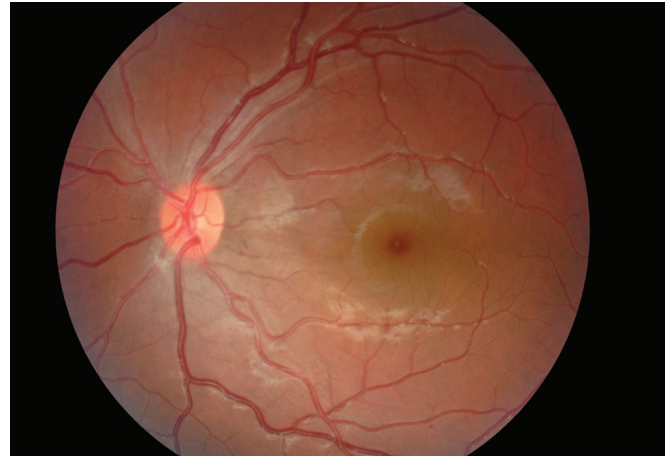
1- Şişli Etfal Eğitim ve Araştırma Hastanesi, 1. Göz Kliniği, İstanbul, Asist. Dr.

2- Şişli Etfal Eğitim ve Araştırma Hastanesi, 1. Göz Kliniği, İstanbul, Doç. Dr.

1- M.D. Asistant, Şişli Etfal Training and Research Hospital, 1st Eye Clinic, İstanbul/TURKEY ÜÇGÜL ATILGAN C., cemileucgul@gmail.com

2- M.D. Associate Professor, Şişli Etfal Training and Research Hospital, 1st Eye Clinic, İstanbul/TURKEY GÜVEN D., dkguven@hotmail.com

**Correspondence:** M.D. Assistant Cemile ÜÇGÜL ATILGAN Şişli Etfal Training and Research Hospital, 1st Eye Clinic, İstanbul/TURKEY



**Figure 1a,b:** Fundus examination of right eye revealed an anomalous inferior retinal vein. It was traversing the horizontal raphe just superior to fovea towards equator after giving secondary branches, one at the optic nerve margin and another over the papillomacular bundle. Cilioretinal artery was present with multiple dilated branches. There were tortuosity and saccular dilatations of small vessels near the superior and inferior temporal arcades. There were no signs of hemorrhage, exudate, edema or foveolar cysts (a). Fundus examination of left eye was normal (b).

### CASE REPORT

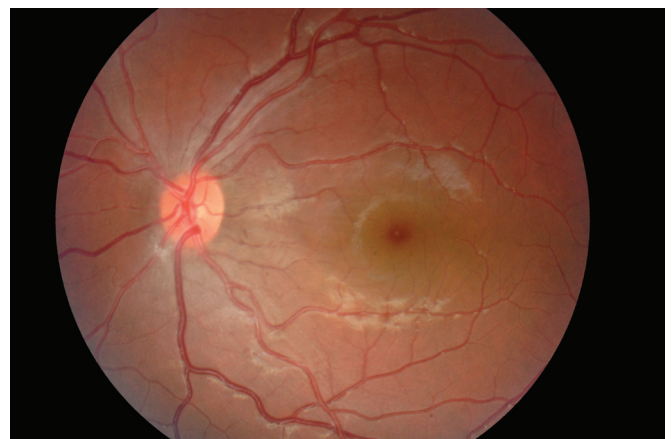
A 13-year-old patient with complaining of headache and television watching closely admitted to our clinic. Best corrected visual acuities were 20/20 in both eyes. Examination of the anterior segment was unremarkable in both eyes. There was no afferent pupillary defect or anisocoria. There was orthophoria with normal extraocular muscle movements. Applanation Goldmann tonometry disclosed intraocular pressure of 16 mmHg OD and 17 mmHg OS.

Fundus examination of the right eye revealed an anomalous inferior retinal vein which was traversing the horizontal raphe just superior to fovea towards equator after giving secondary branches, one at the optic nerve margin and another over the papillomacular bundle. Cilioretinal artery was present with multiple dilated branches. There were tortuosity and saccular dilatations of small vessels near the superior and inferior temporal arcades. There were no signs of hemorrhage, exudate, edema or foveolar cysts.

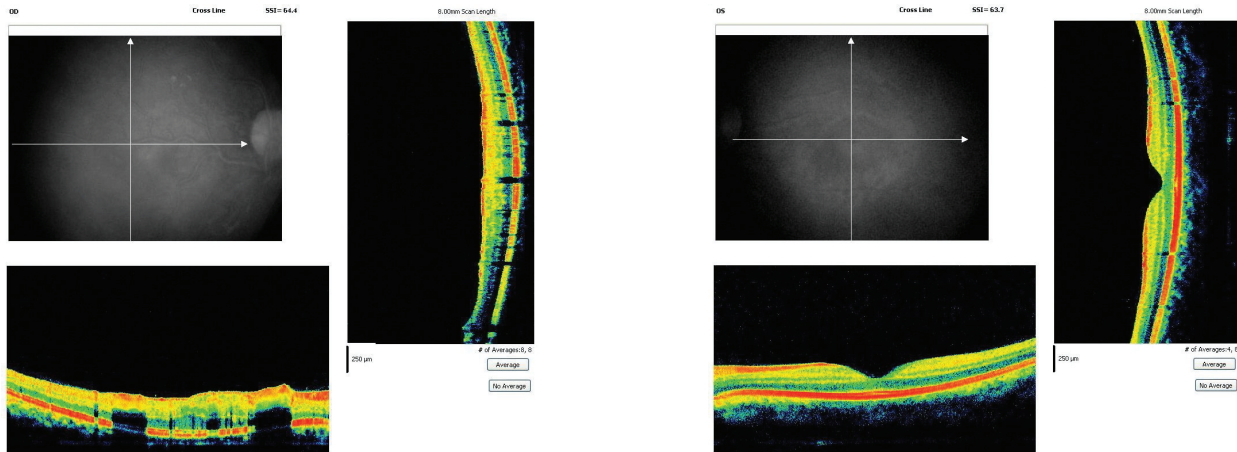
Fundus examination of the left eye was normal (Figure 1a,b). Our patient had no neurological complaint in accordance with his normal neuromuscular examination. Fundus fluorescein angiography of the right eye revealed a venous congenital retinal macrovessel with multiple tiny abnormal arteriovenous communications and showed early filling and delayed emptying (Figure 2a). There was no leakage or ischemia. In the left eye there were twining of tiny branches on the retinal veins (Figure 2b). Spectral-domain optical coherence tomography (RTVue Model-RT100 Version 2.0 Optovue Inc. Fremont, CA) of the right eye revealed shadow effects of abnormal macrovessels. There were no cysts or edema, and fovea could not be visualized (Figure 3a,4a) Left eye revealed normal foveal contour (Figure 3b,4b).

### DISCUSSION

Congenital retinal macrovessel is a rare condition that can be an artery alone, a vein alone or an artery and a vein travelling together.<sup>4</sup>



**Figure 2a,b:** Fundus fluorescein angiography of right eye revealed a venous congenital retinal macrovessel with multiple tiny abnormal arteriovenous communications and showed early filling and delayed emptying. There was no leakage or ischemia (a). In the left eye there were twining of tiny branches on the retinal veins (b).



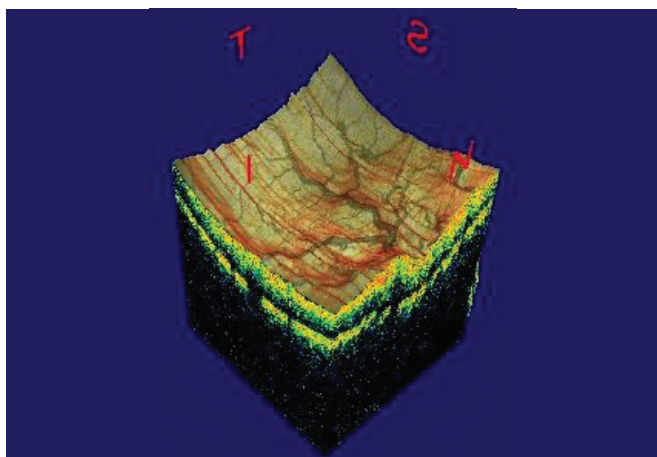
**Figure 3a,b:** Spectral-domain optical coherence tomography (RTVue Model-RT100 Version 2.0 Optovue Inc. Fremont, CA) of right eye revealed shadow effects of abnormal macrovessels. There were no cysts or edema, and fovea could not be visualized (a). Left eye revealed normal foveal contour (b).

In majority of the cases, the anomalous vessel is a vein and is found in one eye only. Congenital retinal macrovessels present usually as one or more large veins that traverse the central macula and have large tributaries that extend beyond the horizontal raphe.

Congenital retinal macrovessels are usually identified and are seen during routine examination. Typically visual acuity is unaffected but when the vessel crosses over the fovea or there are foveolar cysts or haemorrhage visual impairment may be encountered.<sup>3,4,8</sup> In our patient there was no foveolar cyst, edema, exudate or hemorrhage and the vision was unaffected.

Archer et al., proposed a classification system for congenital vessels that exhibited arteriovenous communications in which there were three categories based on severity of the vascular disorders.<sup>10</sup>

In group 1; anomalous arteriovenous communications are localized to one sector of the retina, most commonly the macula. The site of arteriovenous communication is virtually impossible to be located without fluorescein angiography where these vessels show full perfusion with rare decompensation.

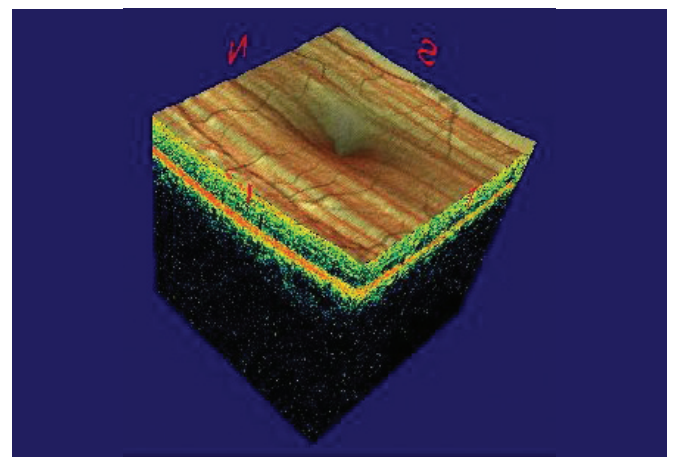


**Figure 4a:** Three dimensional view of abnormal macrovessels of right eye with Spectral-domain optical coherence tomography.

In group 2 the anomalous vessels are larger than in group 1. The arteriovenous malformations exhibit direct communication without interposition of capillary or arteriolar elements. Decompensation of these arteriovenous communication can occur and photocoagulation may be needed. Group 3 contains the most severe forms with largest caliber vessels that can result in retinal complications that lead to severe vision impairment. The anastomosing channels are of large caliber, they are intertwined and convoluted so that separation into arterial and venous components becomes almost impossible.

Finding in our patient can be classified into group 1. Anomalous retinal vessel was a vein that crossed the horizontal raphe over the macula. The vein did not cause any signs of decompensation or any visual impairment. We could not show the normal foveal contour of the patient's right eye with optical coherence tomography, but examination of central visual field that will be done when the patient is cooperated may be helpful to us.

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 congenital retinal macrovessels.<sup>11</sup>



**Figure 4b:** Three dimensional view of macula of left eye with Spectral-domain optical coherence tomography.

During 15<sup>th</sup> to 16<sup>th</sup> prenatal week, mesodermal mesenchymal cells appear near the hyaloid artery on the disc. These cells later form the vascular system that will replace the embryonic blood supply of the hyaloid system.

Mesenchymal cells invade the nerve fiber layer of the retina and eventually differentiate into endothelial cells and form cords that canalize to become capillaries. Initially, no arteries or veins are recognizable. After differentiation, arteries, veins and capillaries are formed.

Eventually blood is diverted into these arterioles, venules, capillaries. 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.

Congenital retinal macrovessels sometimes have a relation with abnormal vascular anomaly of conjunctiva and mouth.<sup>12</sup> Retinal macrovessels must be differentiated from acquired venous collaterals due to retinal occlusion vein, racemose angioma, capillary hemangioma, retinoblastoma and malignant melanoma.

Visual acuity evaluation, fundus examination, photography, fluorescein angiography, optic coherence tomography are important for both diagnosis of retinal macrovessels and follow-up for decompensation.

## REFERENCES/KAYNAKLAR

1. Mauthner L.: Lehrbuch der Ophthalmoscopie. Vienna: Tendler, Co. 1869:249.
2. Tamler E.: Large aberrant macular vessels. Am J. Ophthalmol. 1958;45:106-108.
3. Chronister CL, Nyman NN, Meccariella AF.: Congenital Retinal Macrovesel. Ophthalm Vis Sci. 1991;68-69:747-749.
4. Brown GC, Donoso LA, Magargal LE, et al.: Congenital retinal macrovessels. Arch Ophthalmol. 1982;100:1430-1436.
5. Hayasaka S, Kanamari M, Marihira K-I, Setogawa T.: Anomalous macular vessels: case report and review of recent Japanese Literature Ann Ophthalmol. 1990;22:454-456.
6. Nakashima M.: Anomalous artery in the region of the macula lutea, abstracted. Klin Monatsbl Augenheilkd. 1922;69:159.
7. Kornzweig AL.: Anomalous retinal vein crossing the macula. Arch. Ophthalmol. 1940;24:362-366.
8. De Crecchio G, Mastursi B, Alfieri MC, et al.: Congenital retinal macrovesel. Ophthalmologica. 1986;193:143-145.
9. Awan KJ.: Arterial vascular anomalies of the retina. Arch Ophthalmol. 1977;95:1197-1202.
10. Archer DB, Deutman A, Ernest JT, et al.: Arteriovenous communications of the Retina. Am J Ophthalmol. 1973;75:224-241
11. Ashton N.: The mode of development of the retinal vessels in man. In : Cant JS, ed. The William MacKenzie Centenary Symposium on the Ocular Circulation in Health and Disease. St Louis: CV Mosby Co. 1969;7-17.
12. Gass JDM.: Stereoscopic Atlas of Macular Diseases, diagnosis and treatment. 4<sup>th</sup> edition, St. Louis. 1997;440-442.