Short communication

Congenital retinal macrovessel: An atypical presentation with low vision and macular thickening


Departamento de Retina, Hospital General Universitario Gregorio Marañón, Madrid, Spain

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A B S T R A C T
Case report: We report a case of a 35-year-old male patient with no medical history, who experienced decreased vision in his left eye that he noticed by chance. After a complete ophthalmic examination, he was diagnosed with congenital retinal macrovessel with macular thickening.

Discussion: Congenital retinal macrovessels are rare vascular anomalies, in which the diagnosis is usually incidental as their visual impact is minimal. In the rare cases where there is a significant visual impairment, this is due to macular hemorrhages, foveal cysts, serous macular detachment, or the course of the vessel itself through the foveal avascular zone.

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Macrovaso congénito retiniano: presentación atípica con baja visión y engrosamiento macular

R E S U M E N
Caso clínico: Se presenta el caso de un varón de 35 años sin antecedentes de interés que consulta por mala visión del ojo izquierdo objetivada de forma fortuita, en el cual tras un completo estudio se llega al diagnóstico de macrovaso congénito retiniano con engrosamiento macular.


E-mail address: ire_blanco@hotmail.com (I. Blanco Domínguez).

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Introduction

Congenital retinal macrovessels are a rare vascular anomaly of unknown etiopathogenicity. It has been proposed that their formation occurs between gestation week 15 and 16.¹ One of the current hypotheses is that they could originate in differentiation errors of mesenchymal cell cords.² Said macrovessels were first defined in 1982 by Brown et al., although the first documented case goes back to 1869 in a report by Mauthner.

At the funduscopic level, macrovessels appear as a large aberrant vessel, normally a vein or venule and in rare cases an artery which crosses the posterior pole and in some cases the fovea.

In angiography, early filling of the aberrant vessel with emptying in late stages is characteristic. Other findings which could be present comprise a dilated surrounding capillary plexus, areas void of capillary perfusion, leaks or non-specific vessel wall alterations or associated arteriovenous communications.³

Typically, macrovessels present unilaterally and are diagnosed fortuitously as in general visual repercussion is very low and the visual prognosis is excellent.¹,³

Clinic case

Male, 35, without relevant history who visited due to poor vision in left eye (LE) which was identified fortuitously.

Uncorrected visual acuity (VA) was of one unit for the right eye (RE) and 1/6 for the left eye (LE).

Exploration revealed microstrabismus in LE (2° exotropia), with normal duction, version and convergence.

Refraction without cycloplegia (RE: +0.5 – 0.5 to 25 / LE: 0.25 – 0.5 to 52°) and under cycloplegia (RE: +1.5 – 0.5 to 32° / LE: +1.25 – 0.25 to 51°) discarded unknown anisometropia.

The anterior pole was normal in both eyes. Under funduscopy the RE did not exhibit alterations but the LE posterior pole exhibited increased superior temporal vein diameter with thickened walls which bifurcated crossing the median raphe, with one of its branches going through the fovea (Fig. 1).

Optic coherence tomography (Cirrus HD-OCT; Carl Zeiss Meditec, Dublin, CA) evidenced foveal profile loss with 360 μm central macular thickening, without intraretinal fluid aggregation (Fig. 2).

Fluorescein angiography (Topcon TRC 50 IX; Topcon Medical Systems, Inc.) enhanced the early macrovessels filling with delayed emptying in late stages. The avascular foveal zone (AFZ) was traversed by an aberrant branch. Areas void of capillary perfusion or leak zones were not observed (Fig. 3).

The macular visual field (Octopus Perimeter 1-2-3; Interzeg AG, Schlieren, Switzerland) was reliable and compatible with normality in both eyes (Fig. 4).

Cerebral facial nuclear magnetic resonance (NMR) discarded the presence of other associated vascular anomalies.

The study reached a diagnosis of LE retinal congenital macrovessels. After one-year follow-up, the patient remains stable.

Discussion

In the majority of cases, retinal congenital macrovessels do not impact VA, although it has been seen that retinal sensitivity to light is reduced throughout the pathological vessels (relative angioscotoma). It is believed that this is due to the circulating hemoglobin,¹,³ but this was not observed in the present case.

On the rare occasions involving significant visual impairment, it is caused by macular hemorrhage (generally due to valsalva maneuvers), foveal cysts, macular serous detachment or the passage of vessels through the AFZ.¹,³

In the present case, VA reduction is explained by the passage of the venule through the AFZ with macular thickening and foveal profile loss, without intraretinal fluid as demonstrated by OCT. Taken together, this would produce LE VA reduction due to congenital organic cause, with secondary microstrabismus in said eye. However, associated functional components cannot be discarded but would be untreatable due to the patient’s age.

Even though this case presents characteristic lesions, it is important to carry out differential diagnostic by means of funduscopy and angiography with other vascular anomalies such as arteriovenous communications, racemose angioma, retina capillary hemangioma, venous tortuosity of congenital origin or secondary to venous obstruction and choroidal tumors.¹

Fig. 1 – Superior temporal macrovessel crossing the fovea.
Fig. 2 – Cirrus HD-OCT with central macular thickening and loss of foveal profile, without intraretinal fluid, (A) at diagnostic time, (B) after 13 months follow-up.

Fig. 3 – AFG: (A) early filling of vein and its collaterals in arterial phases. (B) AFZ crossed by aberrant vessels. (C) Delayed emptying of superior temporal vein.

Fig. 4 – Macular visual field without alterations. (A) Right eye and (B) left eye.
In some cases, above all when arteriovenous communications are observed or in the presence of neurological symptoms, NMR is important to discard association with cerebral vascular anomalies such as the Wyburn–Mason or Bonnet–Dechaume–Blanc syndromes, phakomatosis characterized by the association of ipsilateral arteriovenous malformations in the maxillary, retina, optic nerve, thalamus, hypothalamus and brain cortex which appear successively, sometimes in the course of several decades.

In what concerns the present patient, the decision to discard said process was mainly due to the presentation of the unilateral poor vision in an adult male without ophthalmological history, which led the authors to consider the congenital origin of said anomaly.

Conflict of interests

The authors declare no conflict of interest.

REFERENCES