CONGENITAL RETINAL MACROVESSELS: A DISCOVERY BY CHANCE

MACROVASOS CONGÉNITOS RETINIANOS: UN HALLAZGO CASUAL

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ABSTRACT

Clinical case: Congenital retinal macrovessels, where an aberrant retinal vessel, usually a venule, is present in the posterior pole and may cross the avascular foveal region, are seen rarely.

Discussion: Most of these cases are unilateral and stable with excellent visual prognosis and are detected on routine examination. Foveal cysts, hemorrhages and displaced foveola may also be seen. These entities must be distinguished from racemose angiomas, capillary hemangiomas of the retina and even when associated with neurological symptoms, should be considered as part of the Wyburn-Masson syndrome (Arch Soc Esp Oftalmol 2008; 83: 273-276).

Key words: Congenital retinal macrovessels, aberrant retinal vessels, displaced fovea.

INTRODUCTION

Congenital retinal macrovessels are outstanding and stable vessels which cross the foveal avascular region. This paper presents three cases encountered by chance and their characteristic angiographic findings.

CASE REPORTS

A 17 year-old patient who visits the practice due to midesopsiae in the right eye (RE) with an uncorrected visual acuity (AVSC) of 1 in both eyes (AO). The right eye fundus shows a superior tem-
poral venule which traverses the macula, with the left eye (LE) being normal (fig. 1).

Fluorescein angiography (FAG) showed an early filling in of the aberrant venule (fig. 2a), non-specific alteration thereof and of the perifoveal vascular tree without dilatations or leaks with a normal foveal avascular area (ZAF) (fig. 2b), probable communication between veins and arteries and a delay in venule evacuation in late stages (fig. 2c). All the supplementary tests such as Amsler’s grid, color test and electrophysiological study were normal, as well as the campimetric test (Humphrey 10-2) without alteration of foveal sensitivity in spite of the subjective reduction of light sensitivity. It remained stable after 8 months.

A 48 year-old patient who visited the practice due to a high pressure crisis. An ophthalmological exploration was made to discard organic repercussions. The right eye fundus showed a foveal cyst and a duplicated superior temporal venule going through the macula (fig. 3) and findings compatible with a degree 1 low-high pressure vascular syndrome in the left eye.

Uncorrected VA in RE is of 0.8 and of 1 in LE. The supplementary tests gave normal results.

The RE FAG showed an early filling of the aberrant temporal venule, a ZAF “split in two” (fig. 4a), focal points of macular hyper-fluorescence compatible with retinography image and vision, without late leaks (fig. 4b) and delayed emptying of the venule (fig. 4c). It remains stable after 1 year.
A 33 year-old patient who visited the emergency service due to blurred vision in the RE. Uncorrected VA is of 1 difficult in RE and 1 in LE. The right eye fundus showed an inferior temporal venule crossing the middle rafe through the fovea, with loss of foveal reflex (fig. 5). The LE was normal. FAG confirmed the above findings (fig. 6a and 6b) and the supplementary tests were normal.

Congenital retinal macrovessels (CRM) are lesions first described by Mauthner in 1869 and defined in 1982 by Brown as large aberrant vessels crossing the middle horizontal rafe without symptoms or associated to minimum changes in vision or color perception (1-3).

CRMs are typically unilateral, generally a single venule which drains or supplies the area of both hemiretinae and which, in an aberrant manner, crosses the posterior pole or the fovea itself.

Fig. 4: FAG: A: early filling of the aberrant superior temporal venule, a ZAF «split in two». B: Focal points of macular hyperfluorescence without late leaks. C: delay in venule emptying.

Fig. 5: Ascending inferior temporal venule inferior going through the fovea. Loss of foveal reflex. Case 3.

Fig. 6: AFG: A: Early filling of the venule in arterial phases. B: persistence of contrast in aberrant venule in tissue phases.
For some authors the prevalence of CRM is in the area of 1/200,000 (1) and it is believed that they are formed in week 15-16 of gestation, although the underlying cause is unknown.

The majority of CRM are identifiable and appear in routine explorations. However, a differential diagnostic must be made with other vascular entities such as artery and vein communications, branch-shaped angioma, retinal capillary heman-giomae, pre-papillary vascular loops, congenital venous tortuosity or secondary to venous obstruction (1) and even with tumors such as retinoblastoma and choroidal melanomas (2,3).

In this sense it is important to carry out neuroimaging studies if neurological symptoms occur so as to discard the association of brain vascular anomalies such as the Wyburn-Masson syndrome (1-3).

On some occasions, the anomaly is described with variations, involving eyesight due to foveal ectopia, diminished foveal reflex (3) due to changes in the foveal pigmentary epithelium (1,3), foveal cysts and/or post-valsalva hemorrhage (1) or to the path of the vein through the foveal avascular area as in the instant cases.

The described characteristic angiographic findings are: early filling and delayed evacuation of the venule, dilated surrounding capillary plexus (4), areas with no capillary perfusion, hyperfluorescence due to RPE alterations (1), non-specific leaks or alterations in the vascular wall and even association with vein-artery communication 1,2,4) mentioned above in the study cases.

Some authors have described alterations in Amsler’s grid and in foveal sensitivity in the visual field (5) and even though we found a subjective reduction in sensitivity to light we were unable to prove it with the standard supplementary tests we carried out. The explanation of this phenomenon or angioscotoma involves the presence of hemoglobin circulating in the vessel (5).

In conclusion, the CRM or aberrant vessels are striking and incidental findings which require a differential diagnostic with other retinal entities and which, with rare exceptions, do not cause any alteration of the patient’s vision.

REFERENCES