Congenital Macular Macrovessels:
A Case Report and Review of the Literature

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Abstract

Purpose: To report a case of congenital macular macrovessels

Case report: An 8-year-old girl was referred for decreased vision in her right eye. Her best corrected visual acuity (BCVA) in the right eye was 20/200 and in the left eye 20/20. Anterior segment examinations were normal in both eyes. Posterior segment examination of the right eye showed abnormal macrovessels crossing the macula and passing over the fovea. Left funduscopy finding was insignificant.

Conclusion: Congenital retinal macrovessels (CRM) are rare anomalies. They are aberrant retinal vessels, usually venules, present in the posterior pole and crossing the avascular foveal region. 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 be also associated with this anomaly. 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. In children with decreased visual Acuity, it should be treated promptly to avoid amblyopia. Regular examination and careful follow-up of vascular lesion is mandatory to limit possible complications.

Keywords: Congenital Retinal Macrovessels, Aberrant Retinal Vessels, Amblyopia


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Introduction

Congenital retinal macrovessels (CRM) are outstanding and stable vessels which cross the foveal avascular region. They are rare and may cause visual impairment by crossing the fovea, foveolar cysts formation or hemorrhage.\textsuperscript{1-11}

Since the first report of a large aberrant retinal vessel in the macula by Mauthner in 1869 only a few cases have been reported.\textsuperscript{3}

However congenital macular macrovessels usually are not associated with visual deficit and the visual acuity is usually preserved and remains stable.\textsuperscript{2}

This paper presents a case encountered by routine amblyopia screening service in a school age child.

Case report

An 8-year-old girl was referred to our clinic from amblyopia screening center in Zahedan for a diminished visual acuity in her right eye.

The patient was healthy and her medical history was unremarkable. On ocular examination the best corrected visual acuity (BCVA) were $20/200$ and $20/20$, and its refraction were $+1.5-1.75\times5$ and $+0.25-0.50\times10$ for the right and left eye, respectively. Anterior segment examination of both eyes was unremarkable.

Dilated fundus examination of the right eye revealed a retinal macrovessel branched from the superotemporal vein which crossed the horizontal raphe near the fovea, and there was a prominent branch of this lesion passing superior to the foveal area (Figure 1). Findings for the left fundus were unremarkable.

Fluorescein angiography of the right eye demonstrated rapid filling of the retinal macrovessel tributaries (Figure 2A, 2B, 2C). Foveal avascular zone was involved by these macro vessels.

Evaluation of the right macula using a 3D-scan optical coherence tomography (Topcon OCT) with median mode $3\times3\times1$ was performed, it demonstrated the increase in retinal thickness of the macula with hyper reflectivity of the inner retina corresponding to the course of the macrovessels and showing associated posterior shadowing (Figure 3).

Figure 1. Fundus photo of the right eye showing superotemporal macrovessel
Figure 2. Fundus fluorescein angiography of the right eye showing rapid filling of the macrovesel without any leakage.
Discussion

CRM are the lesions that were 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 the blood from both hemiretinas, and in an aberrant manner crosses the posterior pole or the fovea itself. For some authors the prevalence of CRM has been estimated to be around 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 development of retinal neural and vascular components seem to be independent of each other.\(^2\)\(^,\)^\(^8\) Visual acuity even though may be reduced but it seems to remain stable.\(^1\)\(^,\)^\(^4\) In most cases there is no visual complaint if the abnormal vessel is not covering the foveola.\(^1\)

The majority of CRM’s are not identifiable and are discovered in a routine exploration. However, a differential diagnosis must be made with other vascular entities such as artery and vein communications, branch-shaped angioma, retinal capillary hemangiomas, prepapillary vascular loops, congenital venous tortuosity or secondary to venous obstruction\(^1\) and even with tumors such as retinoblastoma and choroidal melanomas.\(^2\)\(^,\)^\(^3\)

In a sense, it is important to carry out neuroimaging studies to look for neurological malformations to discard the association of brain vascular anomalies such as the Wyburn-Masson syndrome.\(^1\)\(^-\)\(^3\)

In some cases the vision can be threatened due to foveal ectopia,\(^3\) changes in the foveal pigmented epithelium,\(^1\)\(^,\)^\(^3\) foveal cysts, postvalsalva hemorrhage\(^1\) or passage of the vein through the foveal avascular zone, as in our present case.

The 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,\(^1\) nonspecific leaks or alterations in the vascular walls and even association of vein-artery communication.\(^1\)\(^,\)^\(^2\)\(^,\)^\(^4\)

**Figure 3.** OCT of the right eye showing retinal thickening in macular area, hyper reflectivity of inner retina and posterior shadowing
Some authors have reported the visual loss after heavy physical stress or activity probably due to a valsalva maneuver. The visual impairment was probably due to her amblyopia because with amblyo-therapy her visual acuity improved significantly.

Alterations in Amsler’s grid and in foveal sensitivity have been also reported in literature.\textsuperscript{4,11}

**Conclusion**

The CRM or aberrant vessels are striking and incidental findings which require a differential diagnosis with other retinal entities and rarely cause any alteration of the patient’s vision.

This lesion can reduce vision by inducing hyperopic shift and making amblyopia specially in young children such as our case.

**References**