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Macroaneurysms Associated with Congenital Retinal Macrovasculars

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Abstract

PURPOSE—Congenital retinal macrovasculars (CRM) are large aberrant retinal blood vessels that cross the horizontal raphe and can traverse the central macula. Using multimodal imaging and optical coherence tomography angiography (OCTA), we describe two cases of CRM associated with macroaneurysms.

METHODS—Two patients presented for evaluation and were found to have CRMs associated with macroaneurysms. Color photography, optical coherence tomography (OCT), fundus autofluorescence (FAF) fluorescein angiography (FA) and OCTA were performed and used to establish the diagnosis and monitor resolution at follow-up visits.

RESULTS—The first patient presented with central vision loss in the right eye and was noted to have a ruptured macroaneurysm and scattered microaneurysms along the course of a venous macrovessel. After 3 months of observation, the patient's vision improved. The second patient presented for evaluation of a cataract in her left eye and was incidentally found to have an arterial macrovessel in her right eye with an associated macroaneurysm. Both cases demonstrated an intricate capillary network in the central macula best visualized on OCTA.

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CONCLUSIONS—Macroaneurysms can occur on both arterial and venous macrovessels. Following rupture of these lesions, hemorrhage and exudation can resolve with observation alone. Macrovasculars can also present with microaneurysms. OCTA can effectively image the complex capillary network associated with these vascular anomalies.

Keywords

Congenital Retinal Macrovascular; Macroaneurysm; OCT Angiography

Introduction

Congenital retinal macrovasculars (CRM) are large aberrant blood vessels that result from abnormal embryologic development and are categorized as type 1 arteriovenous communications.¹ They are usually venous in origin, asymptomatic and cross the horizontal raphe.^{2,3}

Retinal macroaneurysms, usually arterial, are seen primarily in the elderly and are associated with hypertension and atherosclerotic vascular disease. Retinal venous macroaneurysms, typically originating from the capillary bed, are rare and have been described in Coats disease and following branch retinal vein occlusion.^{4,5}

Previous reports have documented macroaneurysms occurring in eyes with retinal macrovasculars^{6,7}, although none have used multimodal imaging including optical coherence tomography angiography (OCTA) to aid in the diagnosis, management and follow-up of these vascular abnormalities.

Herein, we present two cases of CRM with associated macroaneurysms. Multimodal retinal imaging, including OCTA, was performed to further characterize the vascular abnormalities in these eyes.

Case Report

Case 1

A 53-year-old woman noticed sudden, painless central vision loss in her right eye 30 days prior to presentation. She had systemic hypertension that was being treated with oral medications as well as a recent diagnosis of acute myelogenous leukemia (AML) for which she was receiving chemotherapy.

On presentation, visual acuity was 20/400 in the right eye and 20/20 in the left eye. The anterior segment examination and intraocular pressure were normal in both eyes. Ophthalmoscopic examination of the right eye revealed intraretinal and subretinal hemorrhage in the central macula. Some of the subretinal blood was dehemoglobinized.

There was an aberrant vein consistent with a congenital retinal macrovascular coursing through the central macula and area of hemorrhage (Figure 1). Ophthalmoscopic examination of the left eye was normal. Fundus autofluorescence (FAF) of the right eye showed central hypoautofluorescence with a surrounding ring of hyperautofluorescence

corresponding to the dehemoglobinized portion of the hemorrhage (Fig 1). FAF of the left eye showed no abnormalities.

Fluorescein angiography (FA) of the right eye confirmed the venous origin of the macrovessel and showed a large saccular dilatation along the vessel wall with surrounding hypofluorescence corresponding to the location of the central macular hemorrhage. These findings were consistent with a spontaneous rupture of a macroaneurysm originating from the CRM. There were also several smaller isolated saccular dilatations along the distal course of the anomalous vessel with no associated dye leakage (Figure 1). Spectral domain optical coherence tomography (SD-OCT) showed foveal and parafoveal retinal thickening overlying a large amount of subretinal hyperreflective material (SHRM) producing posterior shadowing. The adjacent retina showed disruption of the ellipsoid zone and a small amount of subretinal fluid (Fig 1). SD-OCT of the left eye revealed no abnormalities.

After observation for 6 months, the hemorrhages steadily resolved. Visual acuity in the affected eye improved to 20/50. SD-OCT showed persistent SHRM with surrounding ellipsoid zone disruption (Fig. 2). The flow associated with the macrovessel was well visualized with OCTA. There was a diffuse network of superficial and deep capillaries associated with this vessel in the central macula. Consequently, there was an anatomical reduction of the foveal avascular zone (FAZ) in the right eye. No flow signal was observed in the area of the ruptured macroaneurysm (Fig. 2).

Case 2

A 56-year-old woman presented for evaluation of a cataract in her left eye. She had an ocular history of longstanding poor visual acuity in the right eye since childhood secondary to anisometropic amblyopia. She had a past medical history of rheumatoid arthritis and ulcerative colitis.

On initial presentation, visual acuity was 20/50 and 20/60 in the right and left eye, respectively. The anterior segment examination showed cataracts in both eyes. Intraocular pressure was normal in both eyes. Ophthalmoscopic examination of the right eye revealed a large aberrant retinal vein and artery along the inferior arcade that extended into the central macula and crossed the horizontal raphe. These findings were consistent with a venous and arterial CRM. A macroaneurysm was clearly visible inferior to the fovea along the wall of the aberrant artery. Ophthalmoscopic examination of the left eye was unremarkable. FAF in both eyes was unremarkable (Fig 3).

Fluorescein angiography of the right eye showed a saccular dilatation along the arterial vessel wall inferior to the fovea. There was no dye leakage associated with the macroaneurysm (Figure 3). Spectral domain optical coherence tomography (SD-OCT) showed a normal foveal contour of the right eye with inner retinal hyperreflectivity and posterior shadowing corresponding to the macroaneurysm. SD-OCT of the left eye showed no abnormalities (Fig 4).

Optical coherence tomography angiography showed the flow within the macrovessels to be associated with a diffuse network of capillaries in the superficial and deep retina (Fig 4).

Presumed arteriovenous communications were detected in the central macula that reduced the size of the foveal avascular zone. The macroaneurysm exhibited a flow signal.

Discussion

Congenital retinal macrovessel has been reported in eyes with retinal cavernous hemangioma^{8,9}, arterial macroaneurysms, and branch retinal vein occlusion.^{2,6,7} When a CRM is not associated with other vascular abnormalities, it typically does not affect visual function. Previous reports have described vision loss from macroaneurysms either branching from or adjacent to CRMs. Goel et al. reported a leaking macroaneurysm in an arterial CRM and Koizumi et al. reported a macroaneurysm in a branch retinal artery with an incidental and non-communicating venous CRM noted on retinal imaging^{6,7}. To our knowledge, no reports to date have described the multimodal imaging findings of CRMs, including OCTA, in patients with these combined findings.

Our first patient's vision loss resulted from rupture of the venous macroaneurysm and resultant foveal-involving hemorrhage. No flow was detected within the macroaneurysm on follow-up OCTA several months after the initial presentation, consistent with spontaneous thrombosis after rupture. Several microaneurysms were noted along the course of this patient's macrovessel. While this patient presented with macular edema, the majority of her reduced vision seemed attributable to the intra- and subretinal hemorrhage associated with outer retinal disruption. The patient's concurrent diagnosis of AML may have played a role in the dilation of the vein and retinal hemorrhage. We elected to observe the patient closely, which led to improvement in her structural abnormalities and visual acuity as the blood slowly resorbed.

Our second patient's CRM and associated arterial macroaneurysm were found incidentally during an evaluation for a cataract in her fellow eye. In contrast to the first case where the ruptured macroaneurysm showed no flow on OCTA, our second patient's macroaneurysm exhibited a clear flow signal due to a presumably intact aneurysmal wall.

OCTA in both patients also showed a reduction in size of the FAZ related to a network of capillaries within the central macula, a finding previously noted by Chawla et al.¹⁰ In both of our cases, the details of this anomalous central vascular network were better visualized on OCTA compared to FA and presumed arteriovenous communications within the capillary network were visualized on the imaging of our second patient.

In conclusion, our two cases demonstrate that the spectrum of vascular changes associated with CRMs, including micro- and macroaneurysms, can be effectively imaged using OCTA. Vision loss attributable to macroaneurysm rupture may improve with observation. OCTA appears useful for monitoring these lesions and provides a more precise delineation of the fine arteriovenous vascular networks when compared to FA.

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Summary Statement

Macroaneurysms can occur in the setting of congenital retinal macrovessels and these vascular anomalies, along with an extensive central capillary network can be successfully visualized using OCT angiography. Macrovessels can also harbor a series of microaneurysms in their vicinity.

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Figure 1.

Color fundus photograph of the right eye (A) of Case #1 demonstrating a ruptured macroaneurysm with surrounding hemorrhage. Some of the hemorrhage is dehemoglobinized. There is a large aberrant retinal vein passing through the area of hemorrhage (white arrow). Corresponding fundus autofluorescence (B) shows central hypoautofluorescence due to hemorrhage with hyperautofluorescence in areas of dehemoglobinized blood. Fluorescein angiogram (FA) shows a large saccular dilatation along the vessel wall corresponding to the aneurysm (yellow arrow) as well as several smaller isolated saccular dilatations along the distal course of the anomalous vessel (asterisks). There is no dye leakage. SD-OCT (D, E) shows retinal thickening overlying subretinal hyperreflective material producing posterior shadowing. The adjacent retina shows disruption of the ellipsoid zone and a small amount of subretinal fluid (red arrows).

Figure 2.

Color fundus photograph (A) of the right eye of Case #1 6 months after initial presentation demonstrating resolution of the retinal hemorrhage and involution of the aneurysm. SD-OCT (B) shows persistent SHRM with surrounding ellipsoid zone disruption (arrow). OCTA (C,D) shows the flow within the macrovessel to be associated with a central network of capillaries in the superficial and deep retina (asterisks). No flow signal was observed in the ruptured macroaneurysm.

Figure 3.

Color fundus photograph of the right eye (A) of Case #2 shows a large aberrant retinal vein (yellow arrow) and adjacent artery (white arrow) along the inferior arcade, extending into the central macula and crossing the horizontal raphe. A macroaneurysm was clearly visible inferior to the fovea along the wall of the aberrant artery (white asterisk). Other than the presence of the macroaneurysm, the corresponding fundus autofluorescence (B) was unremarkable. FA (C) showed no significant dye leakage associated with the hyperfluorescent macroaneurysm.

Figure 4.

SD-OCT of the right eye of Case #2 (A) shows a normal foveal contour, with a small hyperreflective lesion corresponding to the cross-sectional location of the macroaneurysm (B, yellow arrow). OCTA shows the flow within the macrovessels (C, red arrows), to be associated with a central network of capillaries in the superficial and deep retina with presumed communication between arterial and venous circulations (yellow asterisks; C,D). A flow signal is present in the area of the macroaneurysm.