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Case report

A case of congenital retinal macrovessel in an otherwise normal eye

Margaret R. Strampe [a, b], William J. Wirostko [a], Joseph Carroll [a, c, *]

[a] Ophthalmology & Visual Sciences, Medical College of Wisconsin, Milwaukee, WI, United States
[b] University of Minnesota Medical School, Minneapolis, MN, United States
[c] Cell Biology, Neurobiology, & Anatomy, Medical College of Wisconsin, Milwaukee, WI, United States

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ABSTRACT

Purpose: To present the case of a 37-year-old female with a foveal macrovessel.
Observations: The patient had an incidental finding of congenital retinal macrovessel (CRM) in the left eye on optical coherence tomography (OCT). Visual acuity was normal, and slit lamp and dilated fundus examinations were otherwise unremarkable. OCT angiography (OCTA) imaging allowed for visualization of the depth profile of the vessel as well as the foveal avascular zone (FAZ). The FAZ and foveal pit were both smaller in the affected eye compared to the fellow eye.
Conclusions and importance: We describe findings of OCTA imaging in a patient with CRM. Previous reports have relied on examination using fluorescein angiography, which does not provide sufficient axial resolution to discern the different vascular plexuses. This report further characterizes how this rare condition can affect foveal morphology and retinal vasculature.

1. Introduction

Congenital retinal macrovessels (CRMs) are aberrant vessels, typically retinal veins, crossing the horizontal raphe in the region of the fovea.[1] This phenomenon was first described by Mauthner[2] in 1869, and the condition is most often an incidental finding. CRMs are rare,[3] but they have been observed in conjunction with several other ocular pathologies including branch retinal artery occlusion,[4] cavernous hemangioma,[5] macroaneurysm,[6] retinal detachment,[7] telangiectasias,[8] vitreous hemorrhage,[9] and reduced visual acuity due to obscuration of the fovea.[10] CRMs have previously been examined using fluorescein angiography,[3,11] but optical coherence tomography angiography (OCTA) now allows for noninvasive imaging of retinal vasculature and segmentation of the superficial and deep vascular layers. OCTA has previously been used to examine vasculature in patients with CRM, but imaging has focused solely on the superficial plexus at the fovea.[10] Here we present a case of congenital retinal macrovessel examined using OCTA.

2. Case report

A 37-year-old female with no past ocular history had apparent interocular asymmetry of the foveal pit while participating in a research study of normal eyes. Subsequent OCT imaging and color fundus photos showed a congenital retinal macrovessel in the left eye (Figs. 1–2). The study protocol was approved by the Institutional Review Board at the Medical College of Wisconsin, and the subject provided informed written consent after the nature and possible consequences of the study were explained. On examination, visual acuity was 20/16 + 1 OD and 20/16 -2 OS. Slit lamp examination was normal. Fundus exam OD revealed normal macula with no vascular abnormality. Fundus exam OS showed a congenital retinal macrovessel in the region of the macula and was otherwise normal.

Multiple 3 × 3 mm OCTA scans (RTVue XR 100 Avanti, Optovue, Inc., Fremont, CA) were obtained in both eyes to allow for image registration and averaging. OCTA imaging of the affected eye

* Corresponding author. 925 N 87th Street, Milwaukee, WI 53226-0509, United States.
E-mail address: jcarroll@mcw.edu (J. Carroll).

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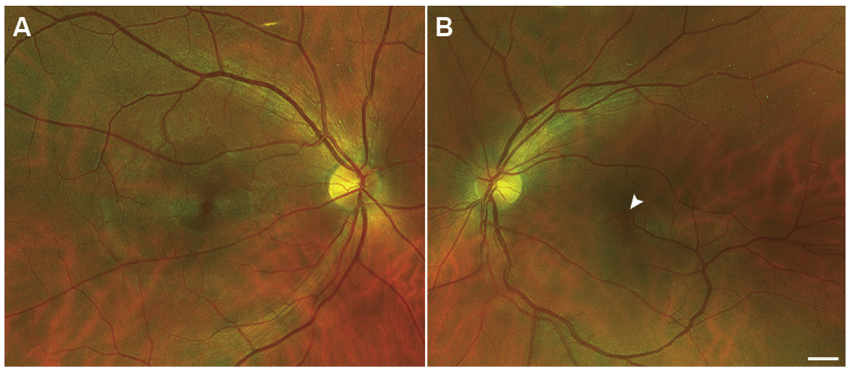

Fig. 1. Color fundus photos of the (A) right and (B) left eyes. White arrow indicates the congenital retinal macrovessel. Scale bar = 1 mm. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

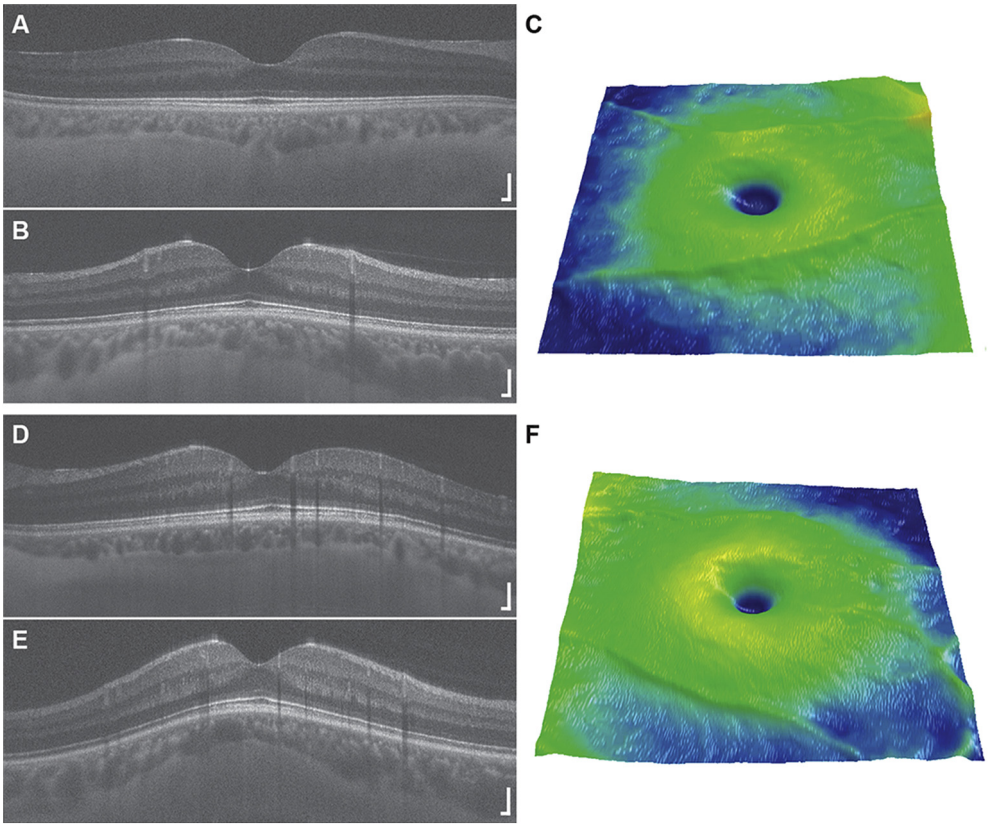

Fig. 2. (A) Horizontal and (B) vertical optical coherence tomography (OCT) scans and a (C) retinal thickness map of the right eye. (D) Horizontal and (E) vertical OCT scans and a (F) retinal thickness map of the left eye demonstrating a large, abberant retinal vein in the region of the fovea. Scale bar = 200 μm.

showed a large retinal vein circumscribing the superior half of the FAZ (Fig. 3). OCTA scans were segmented at the level of the superior and deep vascular plexuses, and the resulting images were color merged. The large retinal vessels in the left eye span both the superficial and deep vascular layers, while the vessels in the right eye are confined to the superficial plexus (Fig. 4).

All measurements were scaled using the patient's axial length (24.36 mm OD, 24.11 mm OS), which was measured using an IOL Master (Zeiss, Dublin, CA). The FAZ was manually segmented using ImageJ,[12] and area and acircularity were calculated using custom MATLAB software (Mathworks, Natick, MA). FAZ area (0.317 mm^2 OD, 0.179 mm^2 OS) and acircularity (1.140 OD, 1.026 OS) were

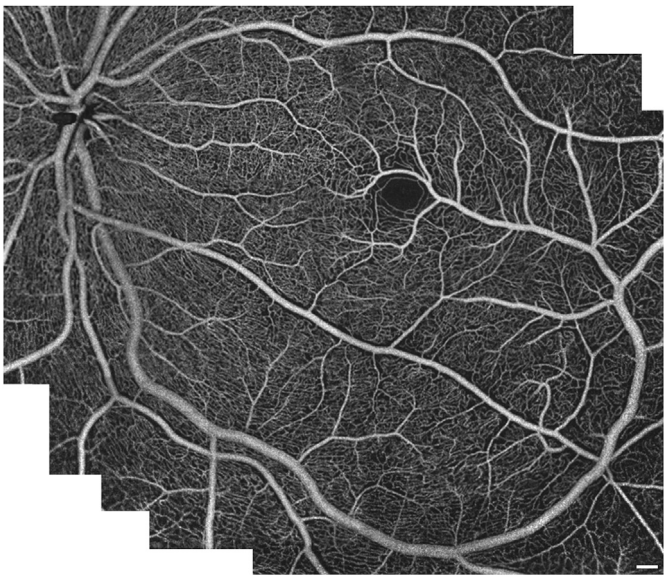

Fig. 3. Optical coherence tomography angiography montage of the superficial vasculature in the left eye, generated in Adobe Photoshop CS6 (Adobe, San Jose, CA). The congenital retinal macrovessel can be seen circumscribing the foveal avascular zone. Scale bar = 100 μm.

smaller in the affected eye. Volumetric images of the macula (6 × 6 mm, 128 B-scans, 512 A-scans/B-scan) were obtained using Cirrus HD-OCT (Zeiss, Dublin, CA). Foveal pit metrics were calculated from these scans using custom MATLAB software, as previously described,[13] and foveal pit volume was decreased in the affected eye (0.0966 mm^3 OD, 0.0680 mm^3 OS).

3. Discussion

OCTA has been established as a reliable tool for visualizing retinal vasculature that is fast, noninvasive, and does not require dilation or fluorescein dye,[14] which is useful for patients for whom these medications are contraindicated. OCTA has an additional advantage in that it allows separate visualization of the superficial and deep vascular layers.[15,16] However, the default OCTA segmentation settings provided by the manufacturer are often inaccurate, requiring manual adjustment of slab thickness to correctly demarcate the appropriate retinal layers.[17] In this case, the segmentation slab was manually adjusted to obtain images of the deep vascular plexus, and the CRM spanned both the superficial and deep vascular layers extending from the internal limiting membrane to the outer plexiform layer.

Analysis of the FAZ in normal eyes has shown a high degree of interocular symmetry of FAZ area, FAZ perimeter,[14] and foveal pit metrics.[18] In this patient, the presence of the CRM causes distortion of the FAZ in the affected eye, resulting in interocular asymmetry in

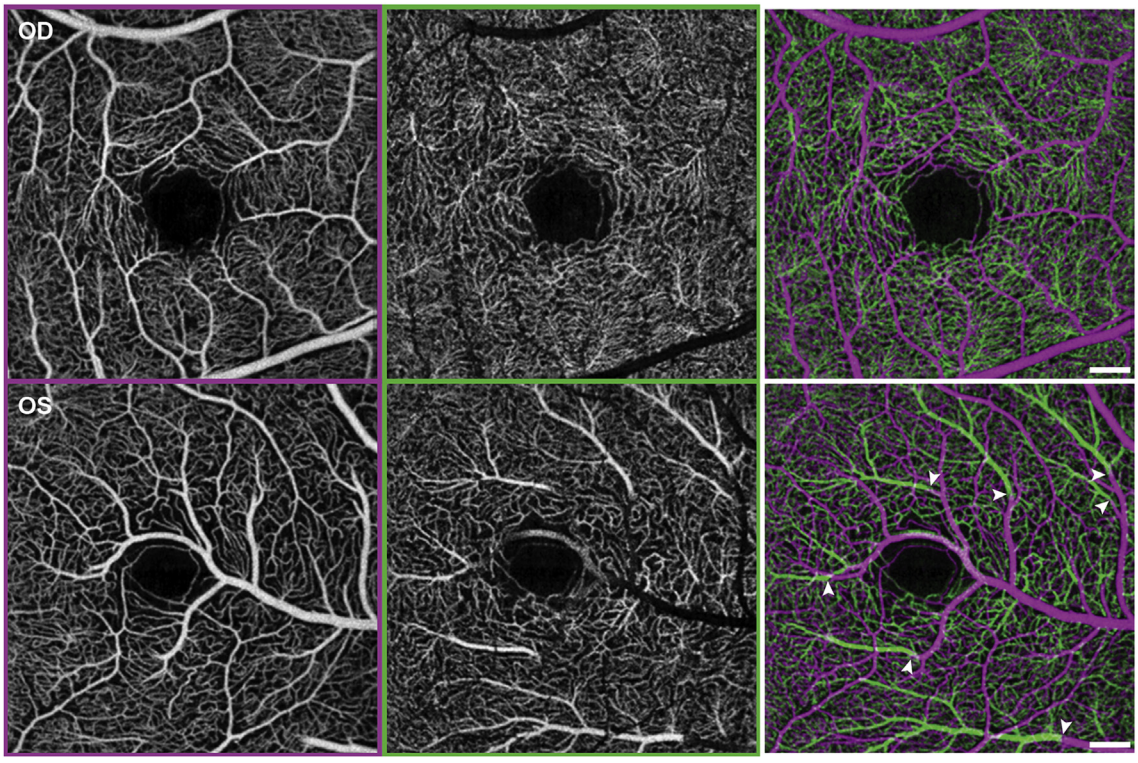

Fig. 4. Optical coherence tomography angiography images of the right (*top row*) and left (*bottom row*) eyes. Images were segmented into superficial (*left*; upper limit 3 μm anterior to the internal limiting membrane, lower limit 15 μm posterior to the inner plexiform layer (IPL)) and deep (*middle*; upper limit 25 μm posterior to the IPL, lower limit 55 μm posterior to the IPL) layers and were then color merged (*right*). White arrows indicate locations where retinal vessels cross from the superficial (pink) into the deep (green) vascular plexus. Scale bars = 100 μm. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

FAZ metrics and foveal pit volume. Previous reports of CRM have described patients with small, distorted FAZs[10] or normal FAZs,[8] but no FAZ measurements or fellow eye images were provided for interocular comparison of foveal morphology.

4. Conclusions

In conclusion, this report presents a case of CRM in an otherwise normal eye. OCTA allowed for noninvasive imaging of the superficial and deep vascular layers, although this did require manual adjustment of slab thickness. FAZ and foveal pit measurements were asymmetric between eyes due to the presence of the CRM.

Acknowledgements and disclosures

Patient consent

Research was conducted under an IRB-approved protocol, and written informed consent was obtained from the subject prior to the collection of any data. This report does not contain any patient identifiers.

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Conflicts of interest

None.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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