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## Case report

## A case of congenital retinal macrovessel in an otherwise normal eye

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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.© 2017 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

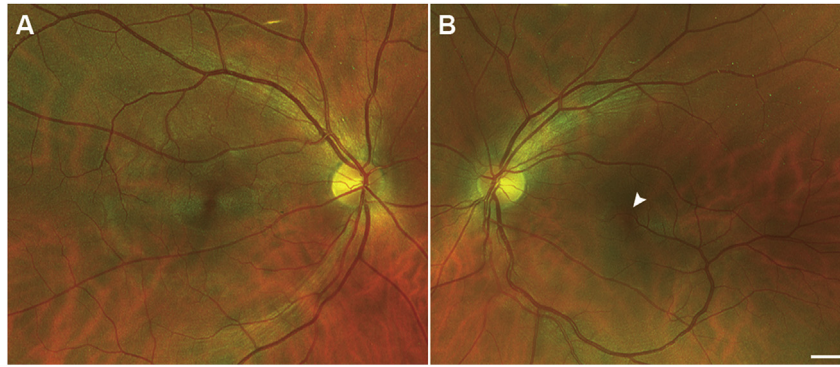
Congenital retinal macrovessels (CRMs) are aberrant vessels, typically retinal veins, crossing the horizontal raphe in the region of the fovea.<sup>1</sup> This phenomenon was first described by Mauthner<sup>2</sup> in 1869, and the condition is most often an incidental finding. CRMs are rare,<sup>3</sup> but they have been observed in conjunction with several other ocular pathologies including branch retinal artery occlusion,<sup>4</sup> cavernous hemangioma,<sup>5</sup> macroaneurysm,<sup>6</sup> retinal detachment,<sup>7</sup> telangiectasias,<sup>8</sup> vitreous hemorrhage,<sup>9</sup> and reduced visual acuity due to obscuration of the fovea.<sup>10</sup> CRMs have previously been examined using fluorescein angiography,<sup>3,11</sup> 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.<sup>10</sup> Here we present a case of congenital retinal macrovessel examined using OCTA.

## 2. Case report

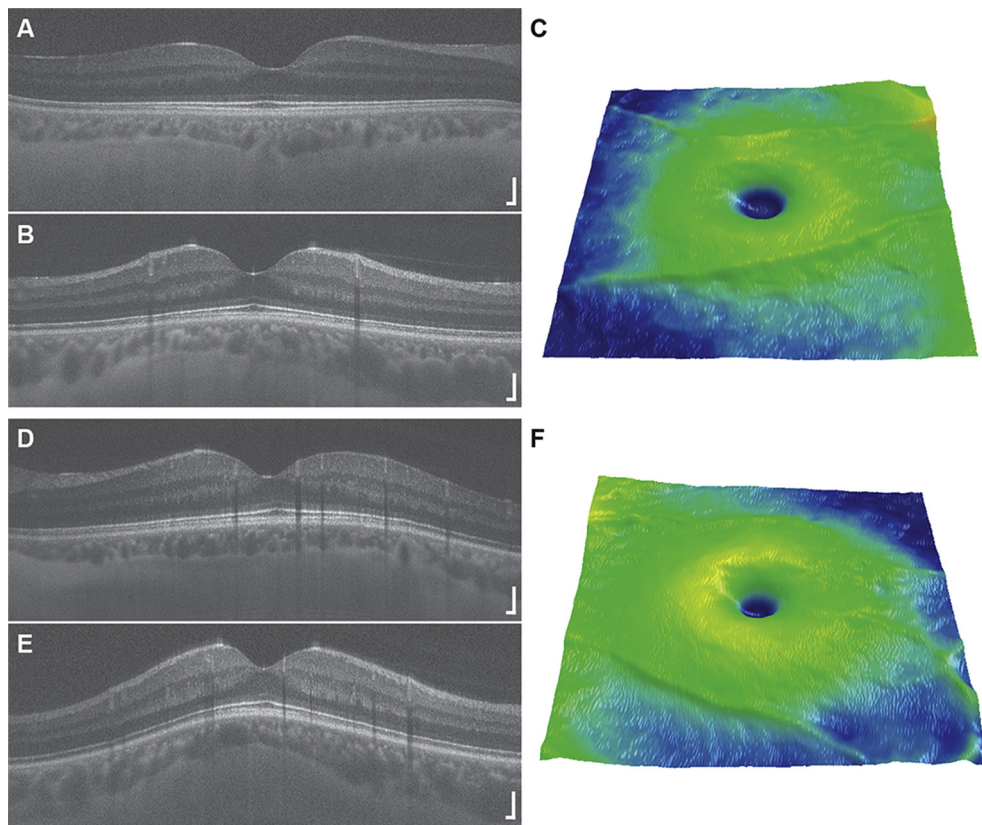
A 37-year-old female with no past ocular history had apparent interocular asymmetry of the foveal pit while participating in a research study of normal eyes. Subsequent OCT imaging and color fundus photos showed a congenital retinal macrovessel in the left eye (Figs. 1–2). The study protocol was approved 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

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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, aberrant retinal vein in the region of the fovea. Scale bar = 200  $\mu\text{m}$ .

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,<sup>12</sup> and area and acircularity were calculated using custom MATLAB software (Mathworks, Natick, MA). FAZ area (0.317 mm<sup>2</sup> OD, 0.179 mm<sup>2</sup> OS) and acircularity (1.140 OD, 1.026 OS) were

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