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.

1. Introduction

Congenital retinal macrovessels (CRMs) are aberrant vessels, typically retinal veins, crossing the horizontal raphe in the region of the fovea. This phenomenon was first described by Mauthner in 1869, and the condition is most often an incidental finding. CRMs are rare, but they have been observed in conjunction with several other ocular pathologies including branch retinal artery occlusion, cavernous hemangioma, macroaneurysm, retinal detachment, telangiectasias, vitreous hemorrhage, and reduced visual acuity due to obscuration of the fovea. CRMs have previously been examined using fluorescein angiography, 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. Here we present a case of congenital retinal macrovessel examined using OCTA.
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 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...
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, and foveal pit volume was decreased in the affected eye (0.0966 mm³ OD, 0.0680 mm³ 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, 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. 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. 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, and foveal pit metrics. In this patient, the presence of the CRM causes distortion of the FAZ in the affected eye, resulting in interocular asymmetry in...
FAZ metrics and foveal pit volume. Previous reports of CRM have described patients with small, distorted FAZs\textsuperscript{10} or normal FAZs\textsuperscript{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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