Case report

Optical coherence tomographic angiography and ultra-widefield indocyanine green angiography of a choroidal macrovessel

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A B S T R A C T

Purpose: We evaluated a choroidal macrovessel using optical coherence tomography angiography (OCTA) and indocyanine green angiography (ICGA). Observations: A 79-year-old female presented with blurred vision in both eyes and metamorphopsia of the left eye. Mild cataract was noted in both eyes. Color fundus photography of the left eye revealed a red-orange tortuous vessel originating from the fovea and running in an inferior-temporal direction. Enhanced-depth imaging OCT revealed a large caliber choroidal vascular shadow and ambiguous line of the photoreceptor and retinal pigment epithelium layers. OCTA demonstrated a serpentine-shaped choroidal vessel. This anomalous vessel was seen by early phase ICGA as a rapidly perfused vessel connected to a vortex vein. We diagnosed this anomalous vessel as a choroidal macrovessel. We identified that cataract induced blurred vision in both eyes and choroidal macrovessel induced metamorphopsia in left eye. She was received cataract surgery for both eyes. The degree of metamorphopsia and the choroidal macrovessel of the left eye remains unchanged after a year of follow-up.

Conclusions and importance: OCTA and ICGA are useful techniques to diagnose choroidal macrovessels.

1. Introduction

Choroidal macrovessel was first reported in 2011 by Lima LH et al. as a dilated and serpentine-shaped choroidal vessel extending from the macula to the temporal periphery without fluorescent leakage.1 Afterwards, a few reports1-4 concerning choroidal macrovessels showed a characteristic appearance using color fundus photography (CFP), near infrared reflectance (NIR), enhanced depth imaging optical coherence tomography (EDI-OCT), and indocyanine green angiography (ICGA). However, no reports have been published that describe choroidal macrovessel appearance and diagnosis by OCT angiography (OCTA). Herein, we report a case of a choroidal macrovessel imaged by OCTA and describe novel choroidal vascular features by ultra-widefield ICGA.

1.1. Case report

A 79-year-old woman was referred to Kansai Medical University Hospital in September 2017 due to metamorphopsia in her left eye and blurred vision in both eyes. She had no previous systematic or ocular disorders in her history. The family medical history was unremarkable for any major disorders. To investigate metamorphopsia of her left eye and blurred vision of both eyes, we performed standard examinations. The best corrected visual acuity (BCVA) at initial examination was 20/50 in each eye. M-CHARTS (KDM-3*, Inami Co, Tokyo, Japan) score of the left eye at initial examination was 0.5. Mild cataract was noted in both eyes.

CFP (TRC50DX®, Topcon, Tokyo, Japan and California®; Optos, Dunfermline, United Kingdom) of the left eye revealed retinal pigment epithelium (RPE) mottling in the fovea and a red-orange dilated tortuous vessel in the choroid extending from the fovea to the temporal area (Fig. 1). To investigate abnormal choroidal vessels of the left eye, further examinations were performed. EDI-OCT (Heidelberg Spectralis OCT®; Heidelberg Engineering GmbH, Heidelberg, Germany) was performed by scanning the macular area including the choroidal abnormal vessel. EDI-OCT of the left eye revealed multiple characteristic findings: (1) thickened choroid due to enlarged choroidal vascular shadow, (2) obscurity of the choroidal-scleral junction at the fovea, (3) steep elevations of the RPE, (4) unable to distinguish line of RPE, ellipsoid zone (EZ), and interdigitation zone (IZ) in the juxtapfoveal area (Fig. 2). En face OCTA (PLEX Elite 9000; Carl Zeiss, Meditec, Inc, Dublin, CA, USA) of the left eye revealed an enhanced serpentine-shaped and dilated choroidal vessel from the fovea to the temporal area, aligned with CFP and ICGA findings (Fig. 3). Fluorescein angiography (FA) (TRC50DX®; Topcon, Tokyo, Japan) of the left eye demonstrated a transmission
Fig. 1. Standard and ultra-widefield color fundus photograph of the left eye demonstrates a reddish-orange serpentine-like choroidal vessel which extended from the fovea towards the temporal area (yellow arrow) and mottling of retinal pigment epithelium around the fovea.

Fig. 2. Green arrow represents choroidal macrovessel with B-scan images. Enhanced depth imaging optical coherence tomography shows a thickened choroid due to a large caliber choroidal vascular shadow (yellow asterisk) and an unclear choroidal-scleral junction (yellow arrow). An enlarged image reveals the degeneration of the ellipsoid zone (EZ), interdigitation zone (IZ), and retinal pigment epithelium (RPE). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

Fig. 3. An en-face Optical coherence tomography angiography (OCTA) image demonstrates a serpentine-shaped and dilated choroidal vessel extending from the fovea to the temporal macula (red arrow). Cross-sectional OCTA shows hyporeflective space in the choroid corresponding to a macrovessel. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)
defect around the fovea (Fig. 4). Ultra-wide-field ICGA (California®; Optos, Dunferline, United Kingdom) revealed characteristic findings at the early phase: (1) tortuous vessels were filled rapidly during the arterial phase, and (2) the anomalous vessel was connected with vortex veins without the interposition of capillary vessels (Fig. 5).

From these findings, we diagnosed that cataract induced blurred vision in both eyes and a choroidal macrovessel induced metamorphopsia in the left eye. After the diagnosis of choroidal macrovessel in the left eye, the patient received cataract surgery for both eyes in November 2017. Both cataract operations were successful without complications. Post-operative BCVA was improved to 20/20 in both eyes. The degree of metamorphopsia in her left eye was unchanged for over a year of follow-up.

Fig. 4. Fluorescein angiography shows a transmission defect around the fovea (yellow arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

Fig. 5. Ultra-widefield indocyanine green angiography shows that the observed choroidal macrovessel filled rapidly and is connected with vortex veins at the early phase without via capillary plexus. Yellow arrow indicates connection between choroidal artery and vortex vein. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)
2. Discussion

There have been a few reports on choroidal macrovessels.1–4 We described a rare clinical case of a choroidal macrovessel that was characterized using multimodal imaging, particularly CFP, EDI-OCT and ICGA.

Most previous reports showed asymptomatic cases. One prior case report by Choudhry N. et al.2 reported that a choroidal macrovessel induced metamorphopsia through increased choroidal thickness which led to the degeneration of the RPE and EZ. In our case study, the deformation of both the RPE and photoreceptor layers was caused by compression due to the dilated choroidal vessel. This resulted in metamorphopsia as Choudhry N et al.2 described. A thickened choroid often leads to disruption of photoreceptor and RPE layers and the occurrence of choroidal neovascularization and/or serous retinal detachment.5,6 This thickened choroid has been commonly called pachyvessel.5,6 Generally, pachyvessel is a status of thickened choroid due to a dilated choroidal “vein” in Haller’s layer. In our case, the patient’s thickened choroid should not be described as pachyvessel because the diagnosed choroidal macrovessel was due to dilated choroidal artery.

Using ultra-widefield ICGA and OCT, blood supply of the choroidal macrovessel originated from the scleral side around the fovea and connected directly to vortex veins without interposition of capillary vessels. From these findings, choroidal macrovessels might be defined as choroidal arteriovenous malformation (AVM). There are several reports about retinal AVMs. Retinal AVMs are congenital and unilateral disorders that usually remain asymptomatic. It is also known as Wyburn-Mason syndrome7,8 if it is accompanied by intracranial AVMs. On the other hand, there have been no reports about choroidal AVMs. This patient had no clear history of intracranial AVM. The overall pathogenesis of choroidal macrovessels remains unclear: whether it is congenital or acquired, and whether there are systemic associations or not.

In this report, the patient’s left eye was stable for over a year. Hereafter, we need to observe whether choroidal macrovessels may induce other severe ocular events.

3. Conclusions

To our knowledge, this is the first report to diagnose and describe a choroidal macrovessel using OCTA and detect choroidal AVM. OCTA is a non-invasive and useful examination to diagnose choroidal macrovessels clinically. Using further ultra-widefield fundus imaging can help address the pathogenesis of choroidal macrovessels.

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Authorship

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

Patient consent

Consent to publish this case report has been obtained from the patient.

Declaration of competing interest

All authors declare that there are no financial or other conflicts of interest.

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