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En-face OCT and OCT angiography analysis of macular choroidal macrovessel☆

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ABSTRACT

Purpose: To analyze en-face optical coherence tomography (OCT) and decorrelation signals on OCT angiography (OCTA) in two cases of macular choroidal macrovessel (MCM).

Observations: Case report. Both the 64-year-old and 71-year-old females presented for a routine evaluation, and multimodal imaging analysis, including color fundus photography, indocyanine green angiography (ICG), spectral-domain optical coherence tomography (SD-OCT) and OCTA, was performed to diagnose a MCM. En-face OCT, en-face OCTA and decorrelation signals were analyzed through the MCM. In both reported cases, color fundus photograph revealed a serpiginoid lesion in the temporal macula. Red-free imaging enhanced the appearance of this lesion resembling a dilated choroidal vessel. Cross-sectional OCT showed an enlarged choroidal vessel causing elevation of the retinal pigment epithelium (RPE) within the fovea. En-face OCTA with segmentation below the choriocapillaris enhanced the MCM delineation. En-face OCT with segmentation below the choriocapillaris showed MCM with a greater distinctness than the en-face OCTA imaging. Decorrelation signals were not observed within MCM on cross-sectional OCTA.

Conclusion and importance: En-face OCT and decorrelation signals on OCTA may have diagnostic value in distinguishing macular choroidal macrovessel from other choroidal vascular diseases.

Macular choroidal macrovessel (MCM) corresponds to a rare vascular anomaly described as a tortuous and extensive choroidal vessel, and usually appears in middle and older-aged women. Commonly, MCM persists ignored due to its subtle clinical features. Although MCM could be considered as a confined pachychoroid condition, the MCM pathogenesis remains doubtful, and there is no certainty whether it is congenital or acquired lesion and whether it is associated with systemic diseases.[1,2]

The analysis of multimodal imaging is very important in cases of MCM since it may be mistaken for other conditions such as subretinal nematode track, choroidal tumor or anomalous posterior ciliary vessel.[3,4] On fluorescein angiography (FA), the filling and laminar flow of MCM is usually normal and occurs before the perfusion of the retinal vasculature. Indocyanine green angiography (ICG) typically shows normal fluorescence in the early phase and hypofluorescence in the late phase of the exam. Spectral-domain optical coherence tomography (SD-OCT) through the MCM reveals a hollow and elevated choroidal lesion in the macula associated with posterior shadowing.[2,3]

Although there are some reports showing MCM on en-face and OCT angiography (OCTA),[5–7] they did not analyze both imaging tools in conjunction or the decorrelation signals inside the MCM. In the present report, we demonstrate the analysis of en-face OCT and decorrelation signals on OCTA in two cases of MCM which to our knowledge has not been previously reported.

1. Cases report

1.1. Case #1

A 64-year-old white female presented for a routine evaluation. There

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was no history of trauma or travelling through developing countries. Best-corrected visual acuity (BCVA) was 20/25 in the right eye and 20/30 in the left eye. Slit-lamp examination revealed mild cataract in both eyes. Fundus examination demonstrated posterior vitreous detachment in the right eye, and the silhouette of an enlarged and tortuous choroidal vessel extending from the fovea to the temporal retina in the left eye. Red-free of the left eye enhanced the silhouette of the dilated choroidal vessel. ICG of the left eye depicted a hyperfluorescent and enlarged vascular structure within the macular area corresponding to a dilated choroidal vessel, suggesting the diagnosis of MCM. Cross-sectional OCT showed the presence of an enlarged choroidal vessel causing elevation of the retinal pigment epithelium (RPE) within the fovea. En-face OCTA (6 × 6 mm scanning pattern) with segmentation below the choriocapillaris showed a hyporreflective and tortuous tract enhancing the delineation of the MCM seen on ICG. En-face OCT (6 × 6 mm scanning pattern) with segmentation below the choriocapillaris showed the MCM with a greater distinctness than the en-face OCTA imaging. Decorrelation signals that represents vascular flow were not observed inside the MCM on cross-sectional OCTA (Fig. 1).

1.2. Case # 2

A 71-year-old diabetic female was referred for a routine fundus examination. BCVA was 20/40 in the right eye and 20/30 in the left eye. Slit-lamp examination revealed pseudophakia in both eyes. Fundus examination was normal in the right eye and revealed a serpiginoid lesion in the temporal macula of the left eye. Red-free imaging enhanced the appearance of this lesion resembling a dilated choroidal vessel. ICG was not performed due to the patient's past medical history of allergy. Cross-sectional OCT showed a juxtafoveal intraretinal cyst (related to diabetic macular edema) and an enlarged choroidal vessel causing elevation of the RPE within the fovea. En-face OCTA (6 × 6 mm scanning pattern) with segmentation below the choriocapillaris enhanced the MCM delineation. En-face OCT (6 × 6 mm scanning pattern) with segmentation below the choriocapillaris showed the MCM with a greater distinctness than the en-face OCTA imaging. Decorrelation signals were not observed inside the MCM on cross-sectional OCTA (Fig. 2).

2. Discussion

The first MCM was reported in 2011,[1] and there are only very few reported cases of MCM in the literature. These abnormal choroidal vessels, usually with a tortuous configuration, are located within the inner choroid, temporally to the fovea.[8,9] MCM is an unusual vascular anomaly that can simulate other fundus conditions such as a parasitic track and choroidal neoplasm. Even though MCM can be recognized as a localized pachychoroidopathy or pachyvessel, the primary pathophysiology might be dissociated from disorders that involve the entire choroid. A precise explanation regarding the vascular nature of the lesion and a clinico-histopathological correlation is still lacking.[3]

In the present reported cases, multimodal imaging showed similar findings of those described in the literature as MCM. A serpentine trackline was observed in the macular area extending from the fovea to the temporal periphery. ICG demonstrated an early arterial phase filling of a dilated and tortuous choroidal vessel, and SD-OCT showed an enlarged and hollow choroidal vessel causing a tubular elevation of the RPE. En-face OCTA with segmentation below the choriocapillaris showed a tortuous tract, with a low OCTA signal, enhancing the delineation of the MCM seen on ICG. En-face OCT with segmentation below the choriocapillaris showed the MCM with a greater distinctness than the en-face OCTA imaging. It could be explained by the faster capture speed and less motion artifacts of a structural image (en-face OCT) in comparison with an angiography image (en-face OCTA). In addition, because of the low OCT signal within the macrovessels, OCTA imaging of macrovessels will

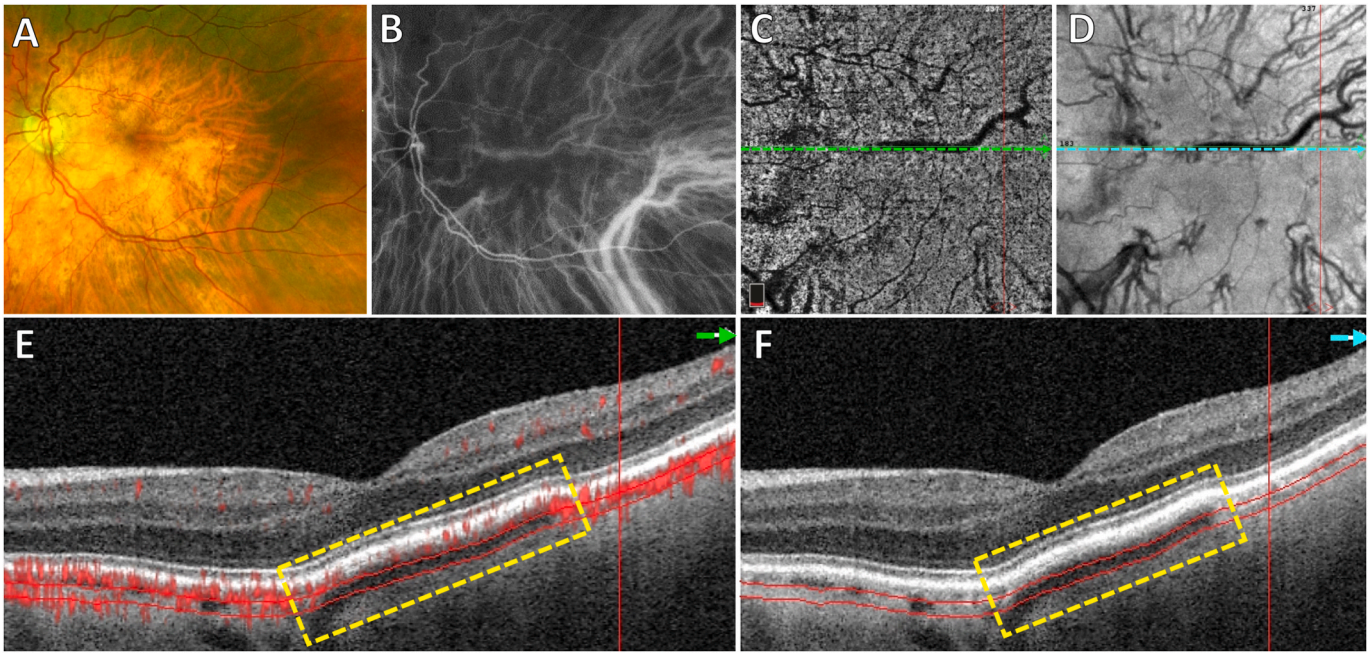

Fig. 1. A. Color fundus photograph of the left eye shows the presence of an enlarged and tortuous choroidal vessel extending from the fovea to the temporal retina. **B.** Indocyanine green angiography (ICG) depicts an enlarged hyperfluorescent vascular structure within the macular area. **C.** 6 × 6 mm en-face optical coherence tomography angiography (OCTA) with segmentation below the choriocapillaris demonstrates a hyporreflective and tortuous tract enhancing the delineation of the macular choroidal macrovessel (MCM) seen on ICG. **D.** 6 × 6 mm en-face OCT with segmentation below the choriocapillaris depicts the MCM with a greater distinctness than the en-face OCTA imaging. **E.** Manual segmentation of the total thickness of the MCM represented on the cross-sectional OCTA by the two red lines within the yellow frame demonstrates hyporreflectivity in the path corresponding to the section of the MCM on the en-face OCTA **(C)** and en-face OCT **(D)**. Note that the decorrelation signals were not observed inside the MCM on cross-sectional OCTA. **F.** Manual segmentation of the total thickness of the MCM represented on the cross-sectional OCT. This is exactly the same segmentation observed on **D**, and clearly shows the hyporreflectivity within the MCM, on a coronal visualization of the MCM. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

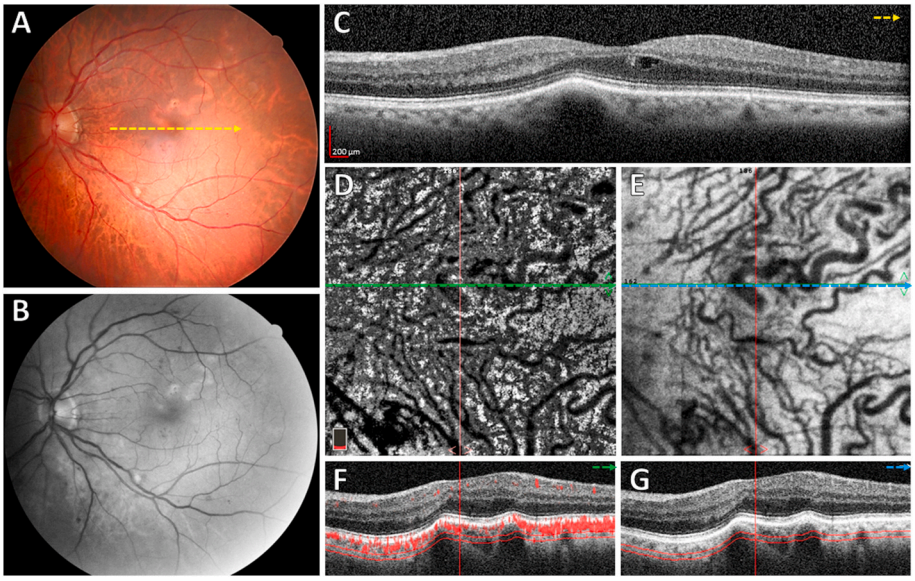

Fig. 2. A. Color fundus photograph of the left eye shows a serpiginoid lesion in the temporal macula. **B.** Red-free of the left eye enhances the appearance of this lesion resembling a dilated choroidal vessel. **C.** Cross-sectional OCT represented with the yellow line in **A**, shows a small juxtafoveal intraretinal cyst and an enlarged choroidal vessel causing elevation of the retinal pigment epithelium (RPE). **D.** 6 × 6 mm en-face OCTA with segmentation below the choriocapillaris demonstrates a hyporreflective and tortuous tract enhancing the delineation of the MCM. **E.** 6 × 6 mm en-face OCT with segmentation below the choriocapillaris depicts the MCM with a greater distinctness. **F.** Note that the decorrelation signals were not observed inside the MCM on cross-sectional OCTA. **G.** This is exactly the same segmentation observed on **(E)**, and clearly shows the hyporreflectivity within the MCM, on a transversal visualization of the MCM. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

not provide reliable information. Thus, OCT imaging, which allows the macrovessels to be seen as regions of low OCT signal, is more useful given the current technology. Besides that, the angiography image could present projection artifacts, which the structural image does not have, especially in the situation the retinal structure to be imaging is deeper (closer to RPE and choriocapillaris).[10,11]

OCTA has emerged as a non-invasive technique to visualize imaging the microvasculature of the retina and the choroid. By overlaying high-resolution structural features, the en-face OCT has enhanced the interpretation of en-face OCTA imaging in several chorioretinal diseases.[12,13] The decorrelation signals on OCTA emerge predominantly from the movement of erythrocytes, and represent the blood flow and vascular structure. These signals are observed in the inner choroidal layers in healthy eyes, and they may change in the lesions from a variety of chorioretinal diseases.[14,15] In the two cases described herein, the decorrelation signals were not observed inside the MCM. We raise two conjectures for the absence of decorrelation signals inside the abnormal choroidal vessel. The MCM endothelium might be too thick to allow the OCTA laser penetration, making it impossible to detect vascular flow, the MCM blood flow speed might be below the minimum threshold of OCTA algorithm, or the disorganized movement of erythrocytes within these dilated vessels could affect the quality of the OCTA. In our opinion, the second hypothesis is less probable since ICG showed a rapid filling of the MCM during the early arterial phase of the exam, and, therefore, indicating a fast blood flow speed. On the other hand, looking at the OCT B-scans, the OCT signal is very low in the region of the macrovessel. This low OCT signal, according to Cole et al.,[16] means that there will be a low OCTA signal, irrespective of whether there is high or low blood flow in the vessel. Moreover, according to Yun et al.,[17] some likely possibilities for a low OCT signal would be absortion and multiple scattering by the blood in the vessel, as well as a phenomenon termed “fringed washout”, wherein fast moving blood flow causes rapid changes in the OCT signal, which, in turn, due to a finite integration time of the detector, cause a decrease in signal. SD-OCT instruments, which use a spectrometer, are particularly susceptible to fringe washout. Regardless of the ultimate cause, based on the images, it appears that low OCT signal is the likely cause of the low OCTA signal within the macrovessels.

Enhanced-depth imaging OCT (EDI-OCT) of the MCM has shown an enlarged lumen compared to the surrounding choroidal vessels.[18] The diameter of the MCM is reported to be larger (approximate mean diameter of 250 µm at the ampulla) than the diameter of the regular choroidal vessel that varies between 20 µm and 100 µm.[3]

En-face OCT was able to highlight the depth and course of a MCM at the level of the choroid and choriocapillaris, and demonstrated a reflectivity similar to these normal vessels in our two cases. This similar reflectivity between MCM and normal choroidal vessels was also observed in other three reports.[4,6,18] The absence of OCTA decorrelation signals shown in our 2 study cases may not help to differentiate MCM from a non-vascular structure such as a nematode track. Although choroidal neovascularization (CNV) lesion is distinct from macrovessels most obviously because of the vessel caliber, shape, and in some cases, its position relative to the RPE, OCTA may help to differentiate these neovascular lesions (increased decorrelation signals) from MCM that demonstrate decreased decorrelation signals on OCTA.[19,20] A previous report, using OCTA, described a low-flow MCM in the choriocapillaris, but it did not display or analyze decorrelation signals.[7]

We described two additional cases of this rare clinical entity named as MCM and the importance of both en-face OCT and OCTA imaging to simplify diagnosis. To the best of our knowledge, this is the first report of MCM showing absence of decorrelation signals on OCTA. En-face OCT and the absence of decorrelation signals inside MCM on OCTA imaging may have diagnostic value, and may differentiate this unusual choroidal vascular lesion from other choroidal vascular diseases.

Patient consent

Consent to publish this cases report has been obtained from patients in writing.

Authorship

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

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Declaration of competing interest

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