Choroidal macrovessel: a multimodal imaging analysis

Satoru Kase, Kousuke Noda, Susumu Ishida
Department of Ophthalmology, Faculty of Medicine and Graduate School of Medicine, Hokkaido University; N-15, W-7, Kita-ku, Sapporo 060-8638, Japan

The authors declare no conflict of interest on this study.

•**Purpose**: Choroidal macrovessel （CMV） is a vascular anomaly characterized by a large serpiginous choroidal vessel with early filling on indocyanine green angiography (ICGA). According to the several reports on CMV, the etiology of CMV seems to be stagnations of choroidal veins due to blood flow obstruction or local vasodilation; however, little is known about the choroidal circulation and hemodynamics. The aim of this study is to analyze the choroidal circulation and hemodynamics in a case with CMV using laser speckle flowgraphy (LSFG).

•**Methods**: A 39-year-old woman with CMV was examined by funduscopic examination, swept-source optical coherence tomography (SS-OCT), fluorescein angiography (FA), ICGA, and LSFG.

•disease spectrum.

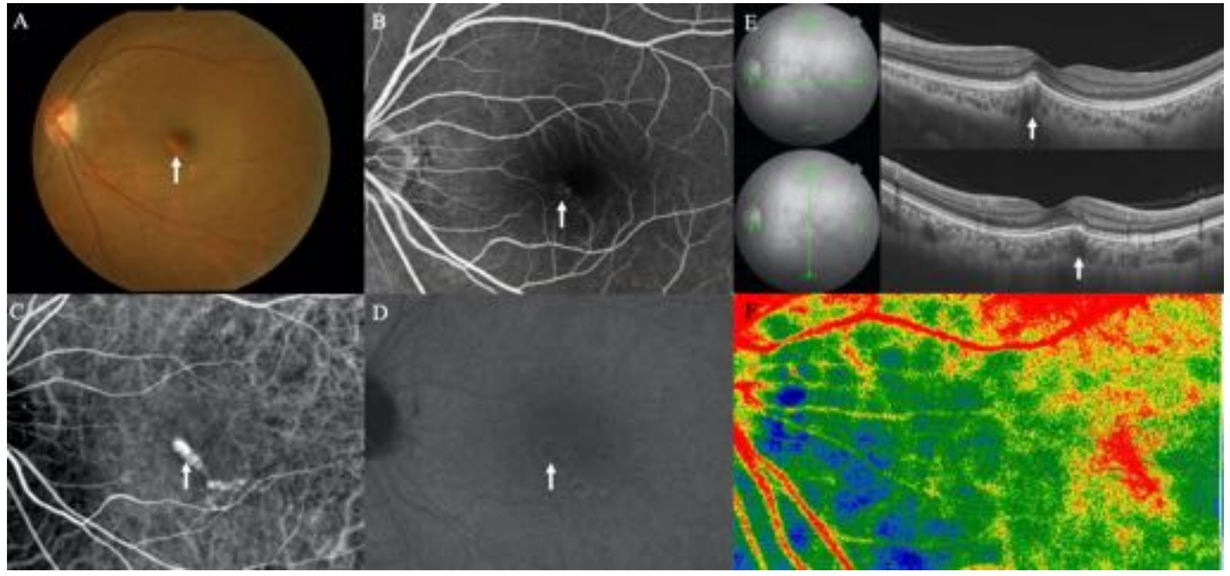

Figure 1. Photographs of the left eye in a patient with CM at the initial visit (A-F).
Fundus shows an elevated choroidal reddish lesion (A). FA shows hyperfluorescence as a window defect (B). ICGA reveals hyperfluorescence and hypofluorescence in the lesion during the initial and late phases, respectively (C, D). SS-OCT shows a dilated luminal structure with low reflectivity localized in Haller's layer. The luminal structure elevates the RPE together with Bruch's membrane (E, white arrows). LSFG displays a focal warm-color pattern, consistent with the abnormal choroidal lesion (F).

Results: SS-OCT showed a dilated hollow configuration in the Haller's layer, and retinal pigment epithelium (RPE) elevation together with Bruch's membrane to the sensory retina. FA demonstrated hyperfluorescence as a window defect corresponding to the choroidal lesion. ICGA detected hyperfluorescence and hypofluorescence in the mass during the initial and late phases, respectively. LSFG showed a warm color reflection, consistent with the CMV site.

Discussion

In the present case, ICGA showed initial hyperfluorescence and late hypofluorescence corresponding to the CM site in synchronization with the dynamics of choroidal circulation. LSFG showed a warm-color pattern indicating hyper-perfusion, consistent with the lesion's hollow structure. These imaging modalities indicated that the CM lesion is a choroidal vascular dilatation with ample blood flow. Therefore, this study demonstrated for the first time that CM is characterized by the hemodynamics of focal hyper-perfusion.

Inflammation causes vasodilation as found in the present case; however, choroidal blood flow on LSFG commonly decreases (*i.e.*, cool-color pattern) in inflammatory diseases such as Vogt-Koyanagi-Harada disease [5] because of leukocyte adhesion to the inner wall of vessels and subsequent infiltration into the choroidal stroma, the latter of which would in turn exert compression from outside of vessels. Moreover, in case inflammation persists in the choroid, other additional pathological events such as choroidal neovascularization (CNV) may occur with time. However, there have so far been no cases of CNV arising from CM even after long-term observation, which is also true of the present case.

In contrast to inflammatory diseases exhibiting choroidal hypo-perfusion, we have shown that the opposite state of choroidal hyper-perfusion is the characteristic hemodynamics of central serous chorioretinopathy (CSC) [6], a typical non-inflammatory disease. As shown in the present case, CM and pachychoroid spectrum diseases (*i.e.*, CSC, pachychoroid neovasculopathy, and polypoidal choroidal vasculopathy) manifest similar fundus findings such as choroidal reddish lesions with RPE elevation, Haller's layer vein dilation, and choroidal thickening. However, pachychoroid spectrum diseases are complicated by RPE detachment and CNV, both of which involve separation between Bruch's membrane and RPE [7], unlike the present case of CM. In addition, ICGA findings in CNV-associated pachychoroid diseases include branching vascular networks and choroidal vascular hyperpermeability [7], whereas the present case of CM showed neither of them on ICGA. Therefore, there are features which distinguish CM from pachychoroid spectrum diseases.

Conclusions: CMV is a morphological vascular change showing hyper-perfusion, but not stagnations of choroidal veins. Although there are some common morphological features between CMV and pachychoroid spectrum diseases, our multimodal imaging analysis indicated that the two disorders should be a different

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