Secondary Macular Neovascularization Arising from Choroidal Hemangioma

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Abstract

A 75-year-old Asian man presented with blurred vision and metamorphopsia in the left eye. Fluorescein angiography and indocyanine green angiography revealed round mottled hyperfluorescence corresponding to the choroidal hemangioma at the temporal macula and a focal hyperfluorescent portion corresponding to a macular neovascular lesion at the subfovea. Swept-source optical coherence tomography showed shallow subretinal fluid at the macula and a dome-shape choroidal lesion occupying the choroidal hemangioma. En face optical coherence tomography angiography showed abnormal type 1 macular neovascularization comparable to a skein of thread at the level of the outer retina slab.

Secondary macular neovascularization may develop at the border of a choroidal hemangioma. This report presents a case of type 1 macular neovascularization arising from the border of a choroidal hemangioma detected using optical coherence tomography angiography.

Keywords: Choroidal hemangioma; Macular neovascularization; Optical coherence tomography angiography

Introduction

An isolated circumscribed choroidal hemangioma (CCH) is a benign vascular tumor characterized by “congested blood vessels separated by thin inter-vascular septa” in the choroid space [1]. It comprises an oval, orange or orange-red colored elevated mass, usually located near the macula or between the macula and the equator, and has a slow growth rate [1,2]. In 200 cases studied by Shields et al. [2] most CCHs contained subretinal fluid at the site of the tumor, and eventually reduced visual acuity despite complete resolution of subretinal fluid. Among untreated CCHs, only one case involved a choroidal neovascular membrane.

Most CCH cases have been diagnosed by indirect ophthalmoscopy or slit-lamp, A/B-scan ultrasonography, fluorescein angiography (FAG), and indocyanine green angiography (ICGA). In recent years, spectral domain optical coherence tomography (SD-OCT) and optical coherence tomography angiography (OCTA) have been utilized to evaluate CCHs [3,4]. The new diagnostic tool, OCTA, can reveal the choroidal vascular structure and new vessels arising from the inner choroidal vessels. To the best of our knowledge, there is no case report of a macular neovascularization (MNV) associated with CCH captured by OCTA image. So, we report a case of type 1 MVN arising from the border of a CCH detected using OCTA.

Case Report

A 75-year-old Asian man visited our ophthalmology department complaining of decreased visual acuity and metamorphopsia in the left eye. His best-corrected visual acuity was 20/20, and 20/100 in the right and left eye, respectively. Horizontal and vertical metamorphopsia scores using M-charts (Inami Co., Tokyo, Japan) were 0.4° and 0.2° in the left eye, respectively.

Fundus examination showed sensory retinal detachment, yellowish deposits involving the fovea, and an approximately four disc-diameter, orange-red oval choroidal mass on the temporal side of the macula (Figure 1A). FAG revealed prominent hyperfluorescence in the nasal macula and patchy hyperfluorescence in the temporal macula. ICGA showed a focal area of hyperfluorescence corresponding to a net of MNV at the nasal area and a large round area of hyperfluorescence on a surrounding hypofluorescent halo corresponding to an elevated orange-colored mass lesion at the temporal macular area (Figure 1B).

Swept-source optical coherence tomography (OCT) (DRI OCT Triton, Topcon Medical System Inc., Tokyo, Japan) revealed irregular neovascular vessels similar to a skein of thread at the level of the outer retina slab. Swfit-source optical coherence tomography angiography showed abnormal type 1 macular neovascularization comparable to a skein of thread at the level of the outer retina slab.

The patient was administered three monthly intravitreal anti-vascular endothelial growth factor injection (bevacizumab 1.25 mg per 0.05 mL, Avastin; Genentech, San Francisco, CA, USA) for subretinal fluid absorption and suppression of macular neovascularization activity. However, the amount of subretinal detachment and best corrected visual acuity did not change during the follow-up period.
“club-like” irregular choroidal vessels on a slab in the deeper choriocapillary layer. In our case, CCH was not clearly detected due to its large size; however, we detected irregular subretinal neovascular vessels similar to a skein of thread at the level of the outer retina on OCTA. The presence of subretinal fluid and MNV were directly correlated with poor visual acuity.

Secondary MNV may rarely develop from a CCH. Choroidal neovascularization associated with an untreated CCH may be due to suble inflammation and chronic ischemia, which may stimulate the release of angiogenic factors [6]. Recently, Akkaya [7] reviewed the pachychoroid disease spectrum, these diseases have three common characteristics: choroidal thickness exceeding 390 µm, dilated Haller’s vein, and thinning in Satter’s vein and the choriocapillaris. In this case, ICGA showed a prominent hypofluorescent area surrounding the choroidal hemangioma, while OCT presented bulky subfoveal choroidal thickness and dilated Haller’s vein below the neovascular networks. So we hypothesize that type 1 MNV may develop at the margin of a CCH, on the pachy-vein of Haller’s layer, secondary to a chronic choroidal capillary ischemia.

Conclusion

MNV secondary to CCH is an uncommon complication. Our report highlights that a MNV arising from the border of a CCH was detected by OCTA. While OCT images show the subretinal fluid accompanied by a subretinal lesion and choroidal space occupying mass, OCTA and ICGA can help detect choroidal neovascularization of the macula. So, Retina specialists should be aware of this complication, as timely diagnosis and treatment are required to prevent visual deterioration.

References

1. Choroidal thickness and dilated Haller’s vein, and thinning in Satter’s vein and the choroidal capillaris. In this case, ICGA showed a prominent hypofluorescent area surrounding the choroidal hemangioma, while OCT presented bulky subfoveal choroidal thickness and dilated Haller’s vein below the neovascular networks. So we hypothesize that type 1 MNV may develop at the margin of a CCH, on the pachy-vein of Haller’s layer, secondary to a chronic choroidal capillary ischemia.

Discussion

While previous reports have investigated the characteristics of CCH using OCTA [3,4], our report describe a secondary choroidal neovascularization associated with a CCH in macular lesion examined by OCTA.

ICGA has been considered a good choroidal hemangioma diagnostic modality due to indocyanine dye leakage and the hypofluorescent and hyperfluorescent patterns. According to Arevalo et al. [5], the fast filling of intratumoral vessels in the early phases, the teneable hyperfluorescence with ICG dyes for up to 120 seconds, and a slight “wash out” of the dye in the late phases of the ICGA are characteristic CCH findings. In our case, we found two hyperfluorescent lesions in the early phase of ICGA, a large round area of hyperfluorescence surrounding a hypofluorescent halo comprising a solitary CCH lesion, and a small area of hyperfluorescence adjusted to the most hypofluorescent area corresponding to FAG dye leakage that indicated a MNV lesion.

OCT images can show the structural changes in the macula associated with choroidal neovascularization and a choroidal space occupying mass, OCTA can capture MNV lesions and the choroidal vascular tumor not visualized with SD-OCT using motion contrast image, detecting flow through retinal and choroidal blood vessels. According to Konana et al. [4], CCH exhibited irregularly arranged vessels on an OCTA slab at the superficial choriocapillary layer, and

Figure 1: A 75-year-old Asian man case of type 1 macular neovascularization arising from the border of a choroidal hemangioma on the left eye. (A) Fundus photography (B) Fluorescein and indocyanine green angiographic image (C) The 12 mm horizontal line image of swept-source optical coherence tomography (D) The 3 mm x 3 mm scans of optical coherence tomographic angiography. Asterisk (*) indicate a dome-shaped choroidal hemangioma. Yellow arrow indicate a net of macular neovascularization, multiple blue triangles indicate a border of choroidal hemangioma on the (B) Image.