Case report

Novel insights into retinal neovascularization secondary to central serous chorioretinopathy using 3D optical coherence tomography angiography

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A B S T R A C T

Purpose: To describe the clinical presentation and novel anatomical features of a patient with chronic central serous chorioretinopathy (CSCR) complicated by retinal neovascularization (RNV).

Observations: A 48 year-old patient with a long-standing history of bilateral CSCR presented to our clinic complaining about a sudden onset of tiny floaters. Multimodal imaging including fundus autofluorescence (FAF), fundus fluorescein (FA) and ICG angiography (ICG) and spectral domain optical coherence tomography (SD-OCT) confirmed the diagnosis of CSCR and revealed a pre-retinal neovascularization and concurring vitreous hemorrhage. Swept source OCT angiography (OCTA) and 3D reconstruction virtual reality determined the retinal origin of the neovascularization. Follow-up examination revealed clearing of the vitreous hemorrhage and spontaneous obliteration of the RNV without any treatment three months following the initial presentation.

Conclusion and importance: To the best of our knowledge, this is the first report of a RNV associated with CSCR which was determined by three-dimensional (3D) OCTA reconstruction.

1. Introduction

Central serous chorioretinopathy (CSCR) is one of the most-common vision-threatening maculopathies, affecting mainly male individuals between 39 and 51 years.1,2 Classically, two different entities of the disease can be distinguished: an acute self-limiting form, exhibiting subretinal fluid, focal alterations of the retinal pigmented epithelium (RPE) and fluorescein leakage. The chronic form is characterized by extensive photoreceptor-, RPE- and choroidal tissue degeneration. Patients with acute CSCR typically report blurred vision, a relative central scotoma and metamorphopsia,3 while patients with chronic CSCR develop absolute scotoma and irreversible loss of visual acuity depending on the extent of RPE and photoreceptor damage.4

Chronic CSCR can be complicated by choroidal neovascularization (CNV) in about two to nine percent of patients mostly affecting patients older than 50 years.5,6 Retinal neovascularizations, in contrast, are a common hallmark of inner retinal vascular disease such as retinopathy of prematurity, retinal vein occlusion or diabetic retinopathy.

We report for the first time on a patient with chronic CSCR complicated by a retinal neovascularization which was diagnosed by the 3D-OCTA reconstruction and interactive virtual reality image display.7

1.1. Case report

A 48-year-old man with a longstanding history of CSCR presented to our clinic complaining about a sudden onset of tiny floaters drifting through the field of view of his left eye. Previously, episodic symptoms of blurred vision had been noticed in both eyes. Past medical history included ethyl toxic liver cirrhosis with liver decompensation and hepatorenal syndrome. Apart from this, no other systemic disease, in particular no diabetes mellitus or cardiovascular disease, were reported. Blood examination excluded other known causes of RNV such as diabetes or infections. The initial ophthalmologic examination revealed a best-corrected visual acuity of 20/20 in both eyes and an intraocular pressure within normal limits. Slit-lamp examination showed a regular anterior segment with clear lenses and without any evidence for iris neovascularization or anterior segment inflammation in both eyes.
Funduscopy revealed normal optic nerve discs, but extensive retinal pigment epithelium (RPE) and retinal alterations in the macula of both eyes. In the right eye, a pigmented epithelial detachment (PED) surrounded by subretinal fluid (SRF) was suspected near the temporal upper vessel suggesting an active choroidal leakage site in CSCR. In the left eye, a vitreous hemorrhage and a branched neovascularization projecting into the vitreous with concurring bleeding was found on the inferior temporal arcade which developed in an area of RPE atrophy (Fig. 1A). The remaining retinal vasculature and peripheral retina was normal in both eyes. Optical coherence tomography (OCT) confirmed extensive areas of photoreceptor and RPE degeneration in both eyes and the presence of a PED and SRF at the temporal superior arcade in the right eye. Fundus autofluorescence (FAF) imaging revealed large areas of reduced FAF surrounded by increased FAF in the macula of both eyes which were consistent with gravitational tracks and emphasized the diagnosis of chronic CSCR. Fundus fluorescein angiography (FFA) demonstrated widespread RPE window defects and an active vascular leakage point at the temporal superior arcade in the right eye. In the left eye, FFA revealed an active neovascularization on the inferior temporal arcade (Fig. 1E). No signs of diabetic retinopathy, vascular occlusive disease or peripheral capillary non-perfusion were observed during funduscopy or FFA. Indocyanine green (ICG) angiography demonstrated multifocal, partially confluent choroidal atrophy with marked reduction in vessel density underneath the vascular proliferation in the left eye. En-face OCT angiography (OCTA) confirmed the presence of a vitreal neovascularization with underlying vessel voids at the level of the choriocapillaris and choroid (Fig. 1G–I) while the retina showed atrophy around the aforementioned epicenter. Three-dimensional (3D) reconstruction of OCTA (3D-OCTA) images confirmed the neovascular origin from retinal vessels that was previously suspected in FFA. Virtual reality OCTA (VR-OCTA) and 3D-OCTA combined with a novel interactive virtual reality rendering method allowed for a detailed investigation of the neovascular complex and its relationship to its surroundings (Video 1). One relatively orthogonal vessel budding out from a larger, superficial retinal vein was found to expand into a cauliflower-like convolute of vessels below and into the detached and condensed posterior vitreous (Fig. 2E, F).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.ajoc.2020.100609.

Since the patient was only mildly symptomatic with a visual acuity of 20/20 and presented with a stable retinal situation without any fibrovascular reaction around the RNV, we opted for close clinical monitoring without intervention. During the follow-up period of six months the vitreous hemorrhage resolved and the RNV obliterated spontaneously.

Fig. 1. Multimodal fundus imaging of a retinal neovascularization secondary to chronic central serous chorioretinopathy. (A) Widefield fundus imaging shows extensive retinal and RPE atrophy (white arrow) with concomitant vitreous hemorrhage (double arrow) and with horizontal mirror line formation. The white box highlights the localization of the retinal neovascularization (RNV). (B) Inset with a magnified display of the RNV (arrow head). (C) Swept source OCTA with flow visualization of the retina (red) and choroid (green) in the area of a retinal neovascularization (RNV) protruding into the vitreous cavity (arrowhead). (D) FAF disclosed extensive RPE degeneration (arrow) and gravitational tracks which are pathognomonic for CSCR. (E) Early fluorescein angiography revealed a vascular proliferation at the lower temporal vessel that developed in the area of marked RPE atrophy (arrow head). (F) Late FA displayed RNV (arrow head) and hyperfluorescent areas originated from RPE atrophies. (G–I) OCTA en-face imaging shows the RNV (arrow head) (G) and the underlying choroidal (H) and choriocapillaris atrophy (I). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)
2. Discussion

Retinal neovascularization is a common hallmark and final path of various inner retinal diseases such as retinopathy of prematurity, retinal vein occlusion and diabetic retinopathy. Outer retinal vascular disease, such as age-related macular degeneration and central serous chorioretinopathy, in contrast, can be complicated by the formation of choroidal neovascularization which occur in two to nine percent in patients with chronic CSCR. To the best of our knowledge, we present for the first time a patient with chronic CSCR complicated by a retinal neovascularization, which has been suspected by FFA and confirmed by virtual reality OCTA imaging.

The diagnosis of chronic CSCR is based on clinical examination and multimodal imaging. The presented case demonstrated active choroidal leakage with concomitant subretinal fluid in the right eye and widespread photoreceptor and RPE degeneration in both eyes which imposed as gravitational tracks and emphasized the diagnosis of bilateral chronic CSCR. Yannuzzi reference Based on the described clinical and imaging findings, other potential differential diagnosis such as choroidal hemangioma, dome-shaped maculopathy, infectious maculopathy or age-related macular degeneration were excluded.

At first sight, a choroidal neovascularization was suspected which is a common complication in chronic CSCR which could have penetrated the retina and into the vitreous. Surprisingly, however, the origin of the neovascularization was found to be retinal and its location preretinal instead of subretinal. Virtual reality OCTA and 3D-OCTA imaging displayed that the proliferative vessel emerged from a retinal vein and extended into the detached posterior vitreous causing mild vitreous bleeding and consecutive densification of the outer vitreous. While Yannuzzi et al. have already described subretinal neovascularization and retinal capillary dilatation (telangiectasia) in CSCR patients, the occurrence of retinal neovascularization in CSCR has not yet been described to our knowledge.

Interestingly, the RNV developed in an area of extensive RPE and retina degeneration which was located just above a large area of confluent choriocapillaris and choroidal atrophy which can be observed in longstanding and severe forms of CSCR. We speculate that the reduced oxygen supply from the choroid and the concomitant retinal degeneration with compromised blood-retina barrier may have favoured the formation of an ischemia-driven retinal neovascularization. Interestingly, the RNV obliterated spontaneously within the following six months.

3. Conclusion

In conclusion, this case demonstrates a novel finding in CSCR which
can be complicated not only by a CNV, but also by RNV. This unique finding poses interesting questions regarding the pathogenesis of retinal neovascularization and illustrates the added value of novel OCTA imaging tools such as a 3D rendering and virtual reality OCT display for the better discrimination between retinal and choroidal neovascularization.

Patient consent

The patient consented in writing to the publication.

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Authorship

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

Declaration of competing interest

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