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EC OPHTHALMOLOGY
Case Report

Choroidal Neovascularization and Polypoidal Choroidal Vasculopathy Associated with Choroidal Nevus Detected by Optical Coherence Tomography Angiography

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Received: August 18, 2021; **Published:** September 29, 2021

DOI: 10.31080/ecop.2021.12.00812

Abstract

Background: A choroidal neovascularization (CNV) is a rare complication of a choroidal nevus. The purpose of this report is to present our findings in a case with a CNV that was detected in an eye with a choroidal nevus.

Case Presentation: A 50-year-old woman, who complain of a distortion of her vision in the left eye, developed a CNV secondary to a choroidal nevus that was detected by optical coherence tomography (OCT) and OCT angiography (OCTA). The CNV was accompanied by a steep retinal pigment epithelium elevation, and the eye was clinically diagnosed with PCV but without performing fluorescein and indocyanine green angiography. The patient was treated with intravitreal aflibercept injections, and a dry macula was obtained after 6 months of therapy.

Conclusion: We conclude that a CNV can be a complication of a choroidal nevus. Our findings showed that OCT and OCTA are helpful methods for detecting PCV and should be performed soon after a choroidal nevus is detected.

Keywords: *Choroidal Neovascularization; Polypoidal Choroidal Vasculopathy; Choroidal Nevus; Optical Coherence Tomography Angiography; Intravitreal Aflibercept Injection*

Abbreviations

RPE: Retinal Pigment Epithelium; CNV: Choroidal Neovascularization; PCV: Polypoidal Choroidal Vasculopathy; FA: Fluorescein Angiography; IA: Indocyanine Green Angiography; OCT: Optical Coherence Tomography; OCTA: Optical Coherence Tomography Angiography; SRF: Subretinal Fluid; PED: Pigment Epithelium Detachment; Anti-VEGF: Anti-Vascular Endothelial Growth Factor

Introduction

A choroidal nevus is the most common intraocular tumor [1]. Although eyes with a choroidal nevus have a good prognosis, it rarely causes vision loss. However, it may cause damage to the retinal pigment epithelium (RPE) and a choroidal neovascularization (CNV). The incidence of CNV is estimated to be 1 - 2% of all cases of choroidal nevus [2]. In addition, the incidence of polypoidal choroidal vasculopathy (PCV) is believed to be lower in eyes with a choroidal nevus [3-8]. The findings of fluorescein angiography (FA) and indocyanine green angiography (IA) are generally needed to diagnose a CNV and a PCV. However, FA and IA cannot be performed in some cases because of potential complications.

We report a case with a CNV that developed secondary to a choroidal nevus, and the CNV was detected by optical coherence tomography (OCT) and OCT angiography (OCTA) without FA and IA. The eye was believed to have PCV as a subtype of the CNV.

Case Report

A 50-year-old woman was referred from a local eye clinic because of a complaint of distorted vision in her left eye. The referring doctor suspected she had age-related macular degeneration. Her best-corrected visual acuity (BCVA) in the left eye at our initial examination was 24/20. A grayish white lesion with a pigmented choroidal nevus was seen beneath the macula of the left eye by ophthalmoscopy. The horizontal OCT images (DRI-OCT, Topcon) showed subretinal fluid (SRF) and an irregularity of the RPE. The vertical OCT images showed hyperreflective zones in the choroid. FA and IA were not performed because the patient refused the examination due to a history of anaphylactic shock caused by the contrast media used for the computed tomography. However, a CNV was detected mainly in the choroid capillary layer and only partially in the outer retinal layer in the OCTA (RTVue XR Avanti, Optovue Inc.) en face images. Cross sectional OCT images with angio overlay at the area where the blood flow was detected in the OCTA images showed a steep RPE elevation indicating a pigment epithelial detachment (PED). En face OCTA images showed multiple PEDs in the region of the CNV. According to the Consensus Nomenclature and Non-Indocyanine Green Angiograph Diagnostic Criteria PCV Workgroup as part of the Asia-Pacific Ocular Imaging Society [9] our case had a sub-RPE ring-like lesion, sharp-peaked PED, double-layer sign, multiple PEDs in the *en face* OCT images, and SRF. These findings suggested that this area was likely to have a polypoidal lesion, and a diagnosis of secondary PCV was made (Figure 1).

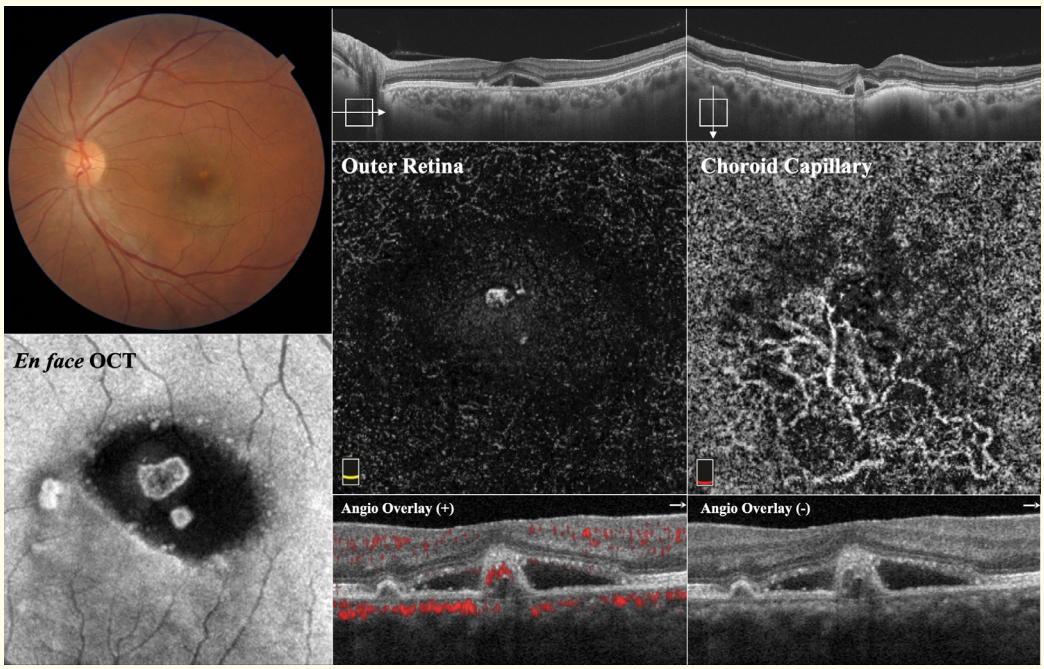

Figure 1: *Multimodal images at initial visit of an eye with a choroidal nevus.*
Color fundus photograph showing a grayish white lesion with a pigmented choroidal nevus.
Cross sectional OCT in the horizontal scan shows the subretinal fluid and retinal pigment epithelium irregularities
Cross sectional OCT image from a vertical scan shows the sharp-peaked PED with the double-layer sign and hyperreflective zones in the choroid. OCTA at the outer retina shows small area of blood flow information corresponding to the sharp-peaked PED. The enface OCTA image at the choroid capillary shows choroidal neovascularization.
En face OCT shows the multiple PEDs.
Cross sectional OCT with angio overlay shows the blood flow information within the sharp-peaked PED. Cross sectional OCT without angio overlay shows the sub-RPE ring-like lesion.
OCT = Optical Coherence Tomography; PED = Pigment Epithelium Detachment; OCTA = Optical Coherence Tomography Angiography; RPE = Retinal Pigment Epithelium.

Intravitreal aflibercept injections (Eylea; Regeneron, Tarrytown, NY, and Bayer HealthCare, Berlin, Germany) were performed seven times in one year for the exudation from the CNV including three consecutive doses in the loading phase. The eye was dry at six months after the start of treatment and has maintained a dry macula for at least one year. The final visual acuity was maintained at 24/20. However, the RPE elevation remained even after one year, and OCTA of the outer retina showed blood flow information in the CNV in the same area suggesting that the CNV remained even after the exudation was suppressed (Figure 2).

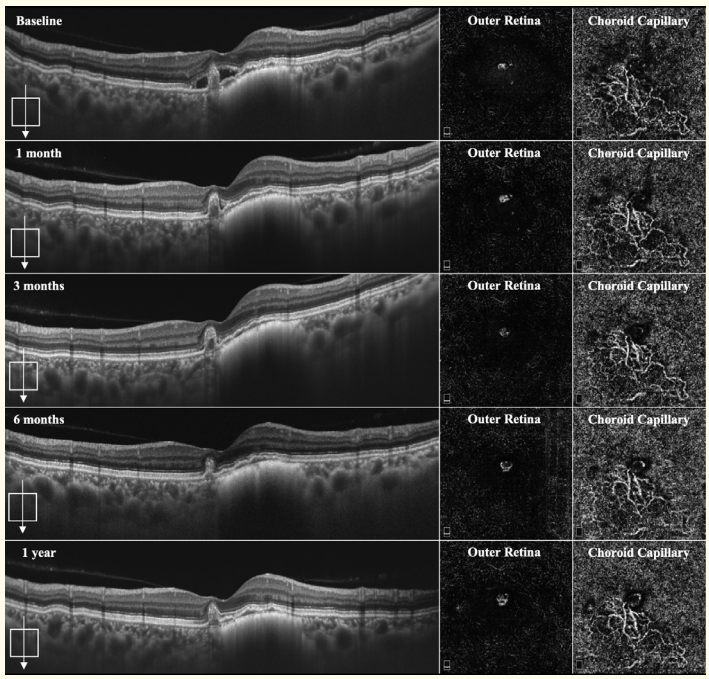

Figure 2: *OCT and OCTA images after anti-VEGF treatments.*
Cross sectional OCT image shows the eye with choroidal neovascularization and the dry polypoidal lesion at six months after the start of treatment and had maintained dry macula at one year.
OCTA shows the blood flow information of the choroidal neovascularization remains unchanged even during the follow-up with treatment. OCT = Optical Coherence Tomography; OCTA = Optical Coherence Tomography Angiography.

Discussion

We report our findings in a patient with a choroidal nevus and a CNV. This was a rare occurrence because the incidence of a CNV associated with a choroidal nevus has been reported to be 1 - 2% [2]. The ophthalmoscopic appearance of the eye resembled an eye with PCV but because the patient declined undergoing FA and IA the polyp-like aneurysmal dilatation of the vessels could not be confirmed.

There have not been many reports of eyes with PCV associated with a choroidal nevus and most of them are case reports [3-7]. Pellegrini., *et al*. [8] reported that among the 11 cases of CNV associated with choroidal nevus, 2 cases (18%) had polypoidal lesions. Although FA and IA angiography were not performed in this patient, the CNV was well-defined in the OCTA images, and the sub-RPE ring-like lesion, a sharp-peaked PED, double-layer sign, the multiple PEDs in the *en face* OCT images, and subretinal fluid indicated that the eye had PCV according to the Consensus Nomenclature and Non-Indocyanine Green Angiograph Diagnostic Criteria [9].

It is important to make an accurate diagnosis in cases of PCV without angiography. In our case, we were not able to diagnose PCV definitively, however we believe that it is acceptable to diagnose this eye as having PCV secondary to choroidal nevus from all of the findings.

The treatment for a PCV is the same as that for a CNV which is intravitreal injections of an anti-vascular endothelial growth factor (VEGF) agent. In our case, the PCV associated with the choroidal nevus was treated with intravitreal aflibercept injections with good results. Although it is necessary to continue with periodical treatments, it is possible to control the exudation with the same treatment as used for AMD. The size of the CNV was unchanged after the treatment. It is known that CNVs do not disappear even after anti-VEGF therapy, and an occlusion of polypoidal lesions might not be detected by OCTA [10]. Although OCTA was helpful for the diagnosis, it may have little clinical significance in the follow-up period.

There are some limitations in this study including examining only one case in a retrospective way. However, there have not been any reports of PCV secondary to the choroidal nevus that was diagnosed noninvasively by OCTA. We expect that the procedures used in this study will be more common as OCTA becomes more widely used.

Conclusion

We found that a PCV can be a rare complication of a choroidal nevus. OCT and OCTA are useful methods to diagnose the PCV and should be performed at least once when choroidal nevus is detected.

Acknowledgements

The authors thank Professor Emeritus Duco Hamasaki of the Bascom Palmer Eye Institute, University of Miami, for his critical discussion and editing of the manuscript.

Conflict of Interest

Dr. Shinozaki reports personal fees from Senju Pharmaceutical Co., Otuska Pharmaceutical Co., and Novartis, outside the submitted work.

Dr. Maruko reports grants from JSPS KAKENHI (Grant Number JP 20K09781), grants and personal fees from Novartis., personal fees from Bayer Yakuhin, Ltd., personal fees from Santen Pharmaceutical Inc., personal fees from Alcon Japan, Ltd., personal fees from Topcon Co., Ltd., personal fees from Senju Pharmaceutical Co., Ltd., personal fees from NIDEK Co., Ltd., outside the submitted work.

Dr. Kodama has nothing to disclose.

Dr. Maruko (R) has nothing to disclose.

Dr. Iida reports grants and personal fees from Novartis. (Japan), personal fees from Bayer Yakuhin, Ltd. (Japan), grants and personal fees from Santen Pharmaceutical Co., Ltd. (japan), grants from Nidek (Japan), grants from Senju Seiyaku (Japan), research support from Canon (Japan), research support from Kowa (Japan), research support from Topcon (Japan), outside the submitted work.

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Volume 12 Issue 10 October 2021