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Polypoidal choroidal vasculopathy with an exceptionally elevated pigment epithelial detachment

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ABSTRACT

Purpose: To present a distinctive case of polypoidal choroidal vasculopathy (PCV) with an exceptionally elevated pigment epithelial detachment (PED).

Observations: We describe the case of a 48-year-old African-American woman who presented with a substantial lesion in the right eye. Fundus examination revealed an exceptionally elevated lesion extending in the inter-papilla-macular region with multiple dark pigmented spots. Indocyanine Green Angiography (ICGA) in the early phase displayed focal hyperfluorescent spots and a blockage of fluorescence within the lesion, particularly overlying the papillomacular bundle. In the late phase, hyperfluorescent spots within the lesion became evident, with a hyperfluorescent outline of the lesion indicating vascularization. Optical coherence tomography in the right eye disclosed an exceptionally elevated PED temporal to the optic nerve with an elevation of more than 2500 μm, along with subretinal fluid and trace intraretinal fluid.

Conclusions and importance: Multimodal imaging unveiled an atypical case of PCV featuring an exceptionally extensive polypoidal lesion overlying the papillomacular bundle with choroidal neovascularization. Given the presence of a highly conspicuous, elevated PED, it was felt that the risk of retinal pigment epithelium tear was high either with anti-VEGF therapy or even due to natural history. In this scenario, the initial treatment choice was photodynamic therapy rather than intravitreal anti-VEGF injection, which led to complete regression with excellent visual acuity.

1. Introduction

Idiopathic polypoidal choroidal vasculopathy (PCV) is a member of the "Pachychoroid spectrum of diseases" of the retina. This condition is characterized by the presence of terminal reddish-orange, aneurysmal polyp-like structures, along with a Branching Vascular Network (BVN). Clinically, PCV often presents with multiple recurrent serosanguineous pigment epithelial detachment (PED). While the majority of PCV cases are diagnosed in Asia, particularly in the submacular region, PCV is prevalent among black females in the US,[1] primarily occurring in the peripapillary region. In this report, we present a unique case of PCV with a PED of exceptional elevation and discuss the diagnosis and management in view of the significant risk of retinal pigment epithelium (RPE) tear.

2. Case report

We present the case of a 48-year-old African-American woman who was referred to our clinic by another retina specialist due to concern regarding a possible ocular mass tumor lesion in her right eye. She had no prior ocular history but had a medical history that included Sjogren syndrome, managed with daily hydroxychloroquine at a dosage of 400 mg daily for 1.5 years (4.86mg/kg). She also had a history of hypertension, anemia, asthma, HPV, anxiety, and depression.

Visual acuity was 20/60 OD, with no improvement using a pinhole occluder, and 20/20 OS. Intraocular pressures were 18 OD and 19 OS. Visual fields were full. Extraocular movements were intact. Anterior segment examination was normal in both eyes. Fundus examination was remarkable for a 2–3 disc diameter substantially elevated lesion overlying the papillomacular bundle with multiple dark pigmented spots (Fig. 1).

SD-OCT in the right eye revealed an extensive PED (thickness >2500 μm) temporal to the optic nerve with subretinal fluid (SRF) and trace intraretinal fluid (IRF). The choroid was not visualized posterior to the PED due to shadowing (Fig. 2). ICGA findings in the early phase displayed focal hyperfluorescent spots and blockage of fluorescence within

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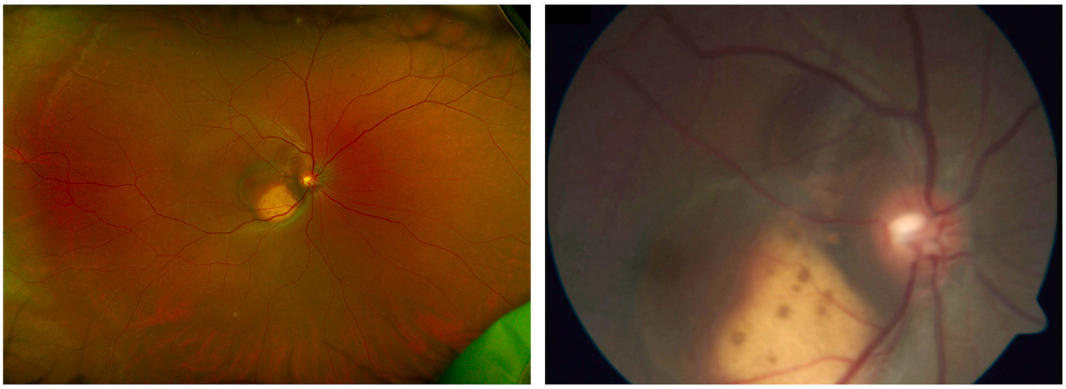

Fig. 1. Color fundus widefield photography (Optos) of the right eye at presentation. The hemorrhagic retinal PED overlying the papillomacular bundle is demonstrated in the left photo. The fundus photo on the right shows the substantially elevated lesion with multiple dark-pigmented spots. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

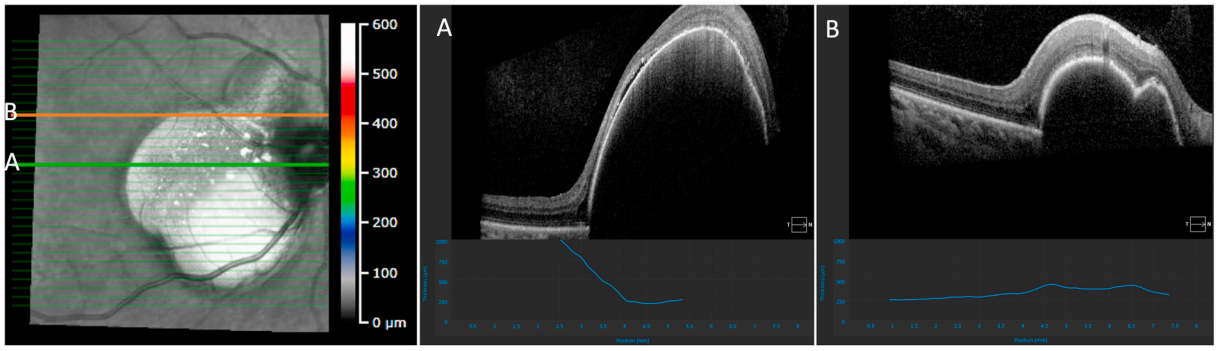

Fig. 2. SD-OCT image with en-face of the right eye macula. A, A >2500-µm-diameter PDE with a subretinal fluid overlying the pigment epithelial detachment (PED). B, Multiple PEDs showing thumb-like protrusions, narrow peaked with inverted V-like complex.

the lesion, particularly overlying the papillomacular bundle (Fig. 3). In the late phase, hyperfluorescent spots within the lesion were noted, and a hyperfluorescent outline indicated vascularization within the lesion (Fig. 3).

A diagnosis of PCV with associated choroidal neovascularization (CNV) was made. Given the remarkably elevated nature of the polypoidal lesion and the potential risk of an RPE tear, the treatment approach began with verteporfin photodynamic therapy (PDT) with 11.4 mg visudyne (5.7 mL in D5W 24.3 mL) using half-fluence and 3500 µm spot size. Three months later, another PDT treatment was performed with visudyne 9.6mg (4.8 mL in D5W 25.2 mL) using the same fluence and spot size as the first time. (treatment located as indicated in the lower right image in Fig. 3). After two courses of PDT treatment, the thickness of the PED decreased dramatically. Six aflibercept injections followed in a treat and extend fashion, and the patient's visual acuity in the right eye improved to 20/20. Notably, the massive PED lesion exhibited significant improvement in terms of flattening of the PED and resolution of the accompanying SRF (Fig. 4), demonstrating complete polypoidal lesion regression. Due to the possibility of recurrent disease activity, the patient will be closely monitored indefinitely.

3. Discussion

Distinguishing PCV from Neovascular AMD (nAMD) is crucial due to PCV's suboptimal response to anti-vascular endothelial growth factor (VEGF) therapies, often requiring additional PDT and/or laser treatment. Clinical signs of PCV differ from those of nAMD, notably in the projection of polyps beneath the RPE without breaching it, in contrast to nAMD, where neovascular tufts extend from the choroid through Bruch's membrane into the sub-RPE or subretinal space.[2]

The diagnosis of PCV relies on a combination of fundus findings, ICGA, and OCT. ICGA has traditionally been considered the gold standard for definitive PCV diagnosis, enabling the visualization of choroidal veins under the RPE and choroidal blood circulation. It reveals two key vascular features of PCV: a BVN beneath Bruch's membrane in the early phase (within the first 30 seconds) and vascular aneurysmal dilations at the network's border in the mid-phase (within the first 5 minutes).[3] However, ICGA has limitations due to its invasive nature, time-consuming procedure, and limited availability.[4]

Recent research suggests that OCT, alone or in conjunction with color fundus photography, can accurately differentiate PCV from nAMD, even without ICGA,[5] offering high sensitivity and specificity. A combination of three major OCT-based criteria for PCV diagnosis, including the presence of an RPE ring-like lesion, en-face OCT complex RPE elevation, and sharp-peaked PED, achieved an area under the receiver operating characteristic curve of 0.90.[6] Additionally, OCT is widely employed to assess treatment response in PCV, evaluating parameters such as SRF, intraretinal cysts (IRC), PED height, and PED volume.

Various treatment modalities are available for managing PCV, including anti-VEGF therapy, PDT, a combination of these modalities, and focal laser photocoagulation.[3] The EVEREST-II and PLANET studies demonstrated that both anti-VEGF monotherapy and combination therapy give excellent anatomical and functional visual outcomes in PCV patients.

Of note, the mean diameter of the largest polyp within a single eye of the EVEREST cohort was 0.35mm (range, 0.15–1.45).[7] In cases where

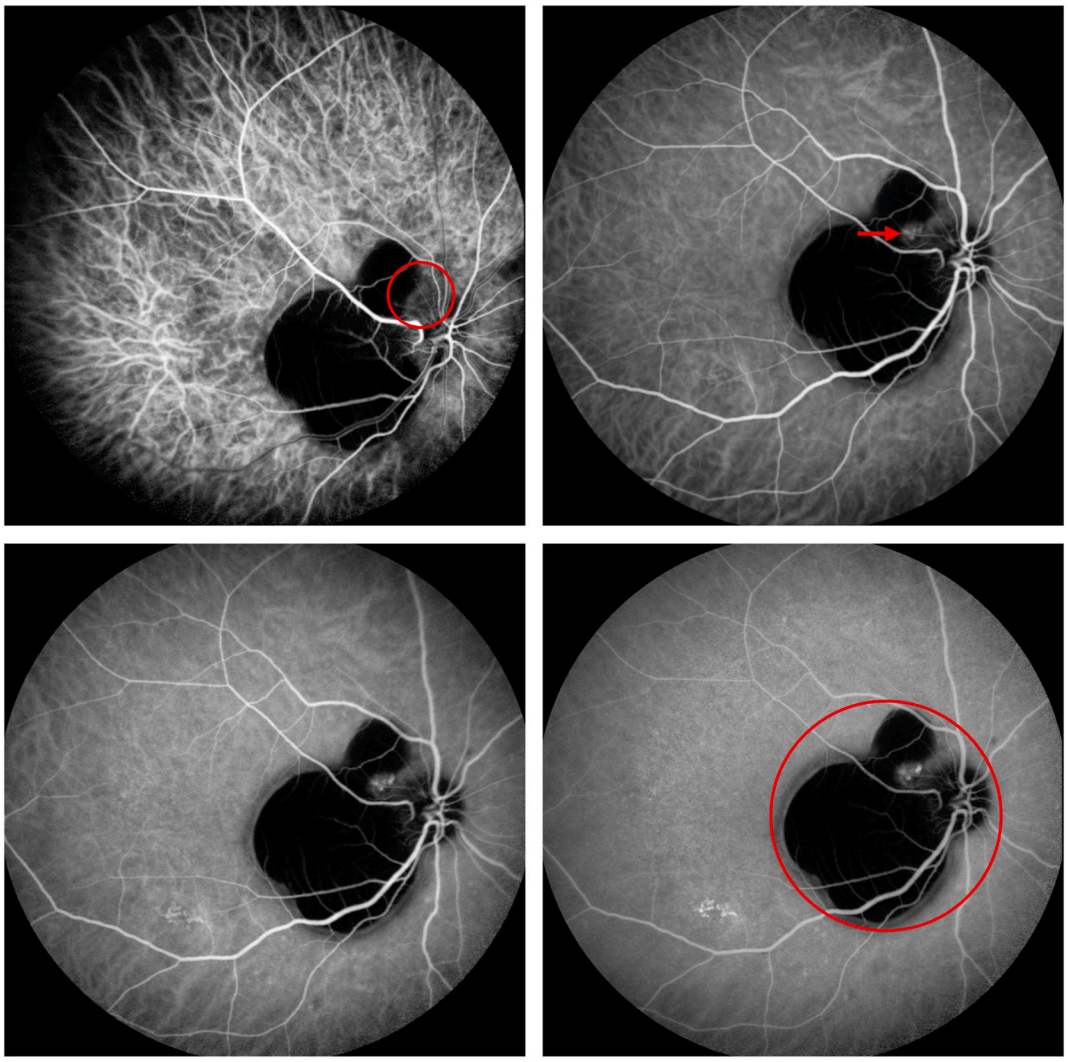

Fig. 3. ICGA of the right eye. Upper left at 12s ICGA showing abnormal vascular channels. Upper right at 3 min ICGA showing nodular areas of hyperfluorescence (arrow), with a halo of hypofluorescence around the nodule. Lower left at 5:44 min and lower right at 10 min, ICGA showing hyperfluorescent spots within the lesion and a hyperfluorescent outline of the lesion, indicating that it is vascularized. PDT treatment location was indicated with red circle. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

there are prominent, elevated, vascularized PEDs, there is a risk of rips or tears occurring in the RPE. Predictors and risk factors for RPE tear development include an increased surface area and a large linear diameter of the subfoveal PED, particularly when the PED basal diameter and PED height exceed 400 μm, a smaller ratio of CNV to PED size, fibrovascular PED, and more recent PED.[8] In our specific case, the patient exhibited a notably high risk of experiencing an RPE tear with a height exceeding 2500 μm. Given that 15–20 % of vascularized PEDs may develop RPE tears after anti-VEGF therapy due to the progressive contraction of the type 1 choroidal neovascular membrane in a vulnerable PED,[9] as well as the transient IOP spikes after injection,[10] we opted for a different treatment approach. While we acknowledge that RPE tears can also occur after PDT,[11] they are much less commonly reported than with anti-VEGF treatments. Additionally, although a paradoxical increase in VEGF levels has been observed following PDT,[12] this was not the primary consideration for our treatment choice. Instead of initiating treatment with an anti-VEGF injection, we commenced with PDT and repeated it once to achieve an improvement in the PED. This approach yielded a favorable outcome for the patient, yet perhaps this could have similarly been achieved with anti-VEGF therapy alone as well. Vigilant observation and monitoring for recurrence or involvement of the second eye are advisable in PCV management.

Patient consent

The patient(s)/patient's legal guardian consented to publication of the case orally.

CRediT authorship contribution statement

Yanliang Li: Writing – original draft, Visualization, Investigation, Data curation. **Hesham Gabr:** Resources. **William F. Mieler:** Writing – review & editing, Validation, Supervision, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

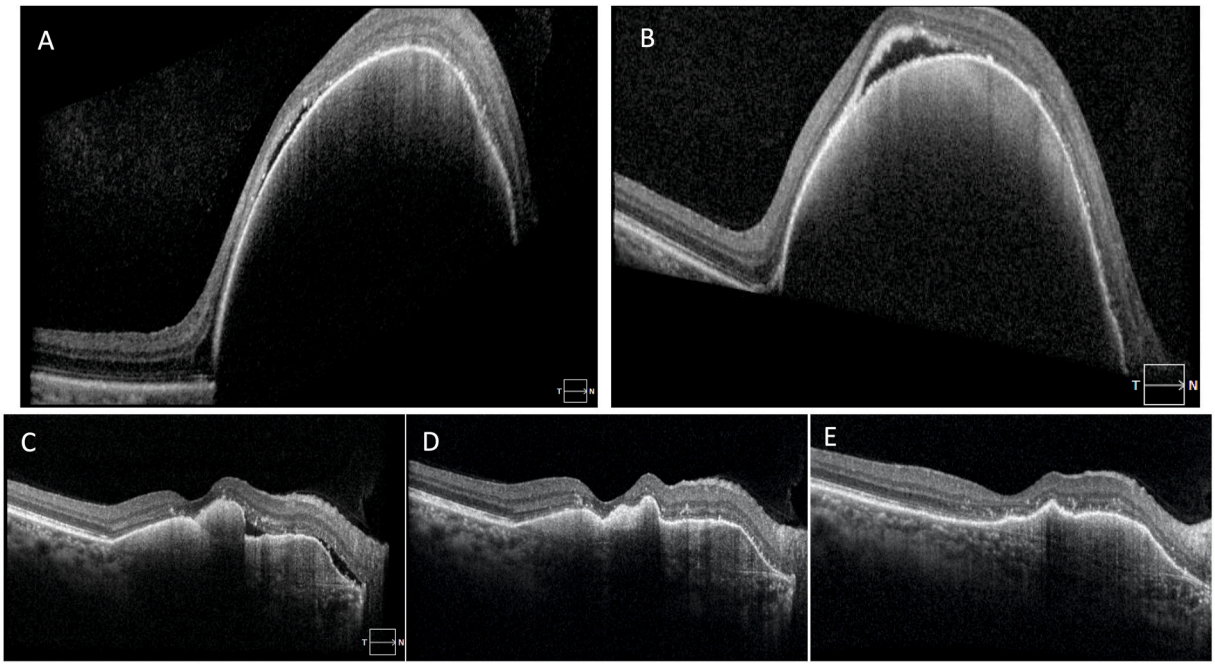

Fig. 4. Sequential SD-OCTs of the right eye macula. A. OCT at baseline. VA was 20/60. B. After the first PDT treatment VA was 20/400. PED was more extensive in width but less elevated with more prominent SRF. Repeated PDT was recommended due to the concern that anti-VEGF could cause RPE tear, given the elevation of PED. C. After the second PDT treatment. VA was 20/40. The appearance of PEDs was significantly improved. D. Six months after the first PDT treatment, Marked improvement in PED and SRF. VA 20/200. The patient was proceeding with aflibercept. E. One year after the first PDT treatment. VA 20/20 with complete regression of all polypoidal lesions.

Acknowledgments and Disclosures

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