Visual impairment associated with choroidal macroaneurysm in a patient with presumed anomalous short posterior ciliary artery

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ARTICLE INFO

Keywords:
Choroidal macroaneurysm
Ciliary artery
Polypoidal choroidal vasculopathy

ABSTRACT

Purpose: To report the clinical findings and treatment outcomes of a patient with a choroidal macroaneurysm associated with a presumed anomalous short posterior ciliary artery.

Observations: A 74-year-old woman with anomalous choroidal vessels had mild visual impairment. Best-corrected visual acuity (BCVA) was 20/25 in the left eye. Funduscopic examination showed a protruded orange lesion temporal to the fovea with exudative changes and retinal hemorrhage. An extensive, thick choroidal vessel network was observed in the superficial choroid. Optical coherence tomography showed a steep protruded lesion beyond the retinal pigment epithelium (RPE) and RPE elevations corresponding to an anomalous choroidal vessel network located at the RPE/Bruch’s membrane complex. Optical coherence tomography angiography showed decorrelation signals corresponding to the steep protruding lesion and anomalous choroidal vessel network. Fluorescein angiography and indocyanine green angiography revealed an aneurysmal lesion and anomalous choroidal vessel network in the choroidal arterial phase. A choroidal macroaneurysm associated with a presumed anomalous short posterior ciliary artery was diagnosed. Regardless of repeated treatment with intravitreal injections of aflibercept/broluzumab and photodynamic therapy, the patient’s BCVA decreased to 20/50. Finally, direct photocoagulation for choroidal macroaneurysm was performed, which resolved the exudative change, and the patient’s BCVA improved to 20/25.

Conclusions and importance: A choroidal macroaneurysm associated with an anomalous short posterior ciliary artery could be a differential diagnosis of polypoidal choroidal vasculopathy. When visual impairment develops due to exudative changes, direct photocoagulation may be the most appropriate treatment option.

1. Introduction

Polypoidal choroidal vasculopathy (PCV) is a common type of age-related macular degeneration, especially in Asian populations. The typical characteristics of PCV include the presence of a central branching vascular network associated with polypoidal lesions at the periphery of the network. We report here a rare case of a choroidal macroaneurysm associated with an anomalous choroidal vascular network, presumably an anomalous short posterior ciliary artery network. The patient developed visual impairment associated with exudative changes in the choroidal macroaneurysm, which resembled, but had different characteristics from typical PCV.

1.1. Case report

A 62-year-old asymptomatic woman with a suspected diagnosis of age-related macular degeneration was referred to our hospital. The patient’s medical history was unremarkable, including ocular trauma, systemic neoplastic diseases, and travel through developing countries. The patient had well-controlled hypertension. Best-corrected visual acuity (BCVA) was 20/20 in both eyes. The intraocular pressure was normal in both eyes. Slit-lamp examination revealed that the anterior segment was unremarkable in both eyes. Funduscopic examination revealed the anterior segment was unremarkable in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes. Spectral-domain optical coherence tomography (SD-OCT; 3D OCT 1000, Topcon, Tokyo, Japan) revealed RPE elevation in both eyes.

https://doi.org/10.1016/j.ajo.2022.101755

Received 2 August 2022; Received in revised form 17 October 2022; Accepted 11 November 2022

Available online 20 November 2022

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Fig. 1. Images in the left eye at the initial visit. A: Color fundus photograph showing thick choroidal vessels at the level of the retinal pigment epithelium (RPE) from the fovea to the temporal macula (dashed circle). There was an area of pigmentary alteration inferiortemporal to the macula. B: Horizontal spectral-domain optical coherence tomography (SD-OCT) showed RPE elevation. C: Indocyanine angiography in the early phase showing anomalous choroidal vessels (arrows) before dye filling in the retinal veins. Arrows indicate the same site as arrows in D. D: Fluorescein angiography in the late phase showed anomalous choroidal vessels from the fovea to the temporal macula (arrows) and an area of background hyperfluorescence inferio-temporal to the macula (dashed circle). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

Fig. 2. Images acquired 4 years after the patient’s epiretinal membrane surgery. A: Color fundus photograph showing an extended area with anomalous choroidal vessels; polypoidal lesions were observed inferio-temporally to the macula. B: A horizontal SD-OCT showing disappearance of the epiretinal membrane with restored foveal pit. RPE elevation with a double-layer sign of the RPE/Bruch’s membrane complex was persistent. C: Indocyanine angiography delineated anomalous choroidal vessels in the arterial phase. D: Indocyanine angiography in the mid-phase delineated anomalous choroidal vessels and polypoidal lesions inferio-temporally to the macula. E: The cross-sectional optical coherence tomography angiography (OCTA) scan along the green arrow in Fig. 2F shows decorrelation signals (white arrows) corresponding to the anomalous choroidal vessels seen in the en-face image at the level of the RPE/Bruch’s membrane complex. F: An en-face image of optical coherence tomography angiography (OCTA) of a 3 × 3 mm square centered at the foveal center showed an anomalous choroidal vessel network in the RPE/Bruch’s membrane complex slab. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)
in the sensory retina with faint subretinal fluid. Because the patient was asymptomatic at this time, she was observed without intervention. It gradually enlarged and accompanied exudative changes, including retinal hemorrhage, hard exudates, and subretinal fluid. The BCVA had worsened to 20/25. SD-OCT showed a protruding lesion that broke into the sensory retina through the rupture of the RPE. A diagnosis of choroidal macroaneurysm with exudative changes was suspected. Seven monthly intravitreal aflibercept injections and three subsequent monthly intravitreal brolucizumab injections were administered; however, the exudative changes worsened, demonstrating circinate retinitis pigmentosa. The patient’s BCVA had decreased to 20/63. FA and IA delineated the aneurysmal lesion in the arterial phase, and dye leakage and pooling at the macroaneurysm in the late phase of FA and IA, respectively. OCTA (Triton, TOPCON) showed an aneurysmal lesion in the photoreceptor slab and an anomalous choroidal vessel network in the RPE/Bruch’s membrane complex slab. Diagnosis of choroidal macroaneurysm was confirmed, and we assumed that an anomalous choroidal vessel network was derived from an anomalous posterior ciliary artery.

Conventional photodynamic therapy with emission diameter of 1000 μm centered on the protruded portion in combination with intravitreal aflibercept injection was performed; however, this combined therapy was ineffective. Finally, direct laser photocoagulation of the choroidal macroaneurysm was performed. Four sessions of laser photocoagulation using a yellow laser with spot size of 200 μm, power of 100–200 mW, and duration of 0.15–0.2 seconds were performed, which led to the remission of exudative changes (Fig. 5). The patient’s BCVA improved to 20/25.

2. Discussion

Our patient had a parafoveal protruding aneurysmal lesion in the anomalous choroidal vascular network. In SD-OCT, the vascular network was located just under the RPE, showing a double-layer sign of the RPE/Bruch’s membrane complex, suggesting that they are located between the basement membrane of the RPE and the rest of the Bruch’s membrane. The foveal choriocapillaris layer was preserved. IA delineated the protruded lesion and anomalous choroidal vessel network in the arterial phase before the dye filling of choroidal veins, which confirmed that the vessels originated from the artery. OCT and OCTA confirmed that the elevated RPE lesion was identical to the course of anomalous choroidal vessels.

One of the differential diagnoses of this condition may be a branching vascular network and polypoidal lesion of PCV. Interestingly, our patient had thinner vessels and polypoidal lesions at the inferotemporal edge of the lesion. We believe that this temporarily altered pigment lesion had similar characteristics to those of PCV. However, in most PCV cases, the branching vascular network shows flat RPE elevation in the form of dense, thinner, and abnormal vessels. Commonly, a single thin vessel does not show a steeply elevated RPE, which was
observed in the central macular lesion in our case. The parafoveal aneurysmal lesion might resemble the polypoidal lesion of PCV; however, the lesion was located centrally, broke into subretinal space through the rupture of RPE, and was clearly delineated in the arterial phase of IA. These were inconsistent with the characteristics of typical polypoidal lesions of PCV, which are commonly located under the RPE at periphery of the lesion, and are delineated in early but not clearly in arterial phase of IA. Indeed, PCV has also been described as aneurysmal type I neovascularization, and previous papers described the association of polypoidal lesion of PCV with aneurysmal change with hyalinized vessels. However, the precise origins and composition of polypoidal lesions of PCV are yet to be determined.

In this sense, we could not completely differentiate the choroidal macroaneurysm in our case from polypoidal lesion of PCV.

Although intravitreal injection of anti-vascular endothelial growth factor with and without combination of photodynamic therapy has been reported to be effective in most PCV cases, the repeated intravitreal injections of aflibercept/brolucizumab and photodynamic therapy showed no effect on exudative changes associated with the patient’s foveal aneurysmal lesions. The ineffectiveness of anti-vascular endothelial growth factor therapy suggests that elevated levels of vascular endothelial growth factor contribute less to this condition.

Another differential diagnosis may be a macular choroidal macrovessel. Macular choroidal macrovessels are usually isolated and course tortuously, but do not form a vascular network. They are located in the entire choroidal thickness and course from the fovea to the temporal periphery in a single direction. These characteristics are different from those observed in our case.

Okubo et al. reported a similar case with unusual dilated choroidal vessels located just beneath the RPE, as seen in our case. However, unlike in our case, the abnormal choroidal vessels were not arteries but veins, and they did not form the vascular network nor demonstrate exudative change.

We diagnosed this aneurysmal lesion as a choroidal macroaneurysm, and assumed that the anomalous choroidal vessel network was anomalous short posterior ciliary artery, because the choroidal arteries arise from the short posterior ciliary artery.

The etiology of this anomalous short ciliary artery and choroidal macroaneurysm remains unknown. This may be due to deterioration of congenital anomalies or acquired abnormalities. A possible explanation may be that the choroidal aneurysm developed at the highly curved portion of the congenital or acquired abnormal arterial networks. Hemodynamically, arterial aneurysms often develop at the curved or bifurcating portions of the artery, as observed in intracranial aneurysms.

The effect of surgical intervention for epiretinal membrane on these conditions is also unclear. Our patient underwent a non-eventful vitrectomy with ILM peeling 6 years before the development of a choroidal aneurysm, which was followed by an excellent visual outcome. It is possible that the loss of ILM above the anomalous choroidal vessels decreased retinal rigidity, causing saccular aneurysmal changes. However, we assume that because the anomalous choroidal vessel network

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**Fig. 4.** Images acquired 8 years after the patient’s epiretinal membrane surgery. **A:** Color fundus photograph shows choroidal macroaneurysm with retinal hemorrhage and hard exudates. **B:** Horizontal SD-OCT scan showing an enlarged protruding lesion with intra-/sub-retinal fluid. Hyperreflective spots corresponding to intra-retinal hard exudates were also observed. **C, D:** Indocyanine angiography delineating the macroaneurysm in the arterial phase (C) and dye pooling in the macroaneurysm in the late phase (D). **E:** Fluorescein angiography in late phase showed dye leakage from the macroaneurysm. **F:** OCTA of 6 × 6 mm square centered at the foveal center showed an aneurysmal lesion in the manually corrected photoreceptor slab (upper left) and an anomalous choroidal vessel network in the RPE/Bruch’s membrane complex slab (upper right). The B-scan image of the OCTA image shows a decorrelation signal at the aneurysmal lesion in the manually corrected photoreceptor slab (bottom). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)
from which a choroidal aneurysm developed was located under the RPE, the effect of ILM peeling on aneurysm formation may be less likely.

Direct photocoagulation of the aneurysm was effective for the remission of exudative changes after failed anti-VEGF and photodynamic therapies. We did not initially perform direct photocoagulation due to concerns regarding aneurysmal rupture or arterial occlusion. Fortunately, no complications were observed during the follow-up period of 18 months after the photocoagulations.

We could not find a similar report in the PubMed database using the keywords of "anomalous/abnormal ciliary artery," "abnormal/anomalous choroidal vessel," and "choroidal aneurysm," which points to the likelihood that ours is the first report of anomalous posterior ciliary artery and choroidal macroaneurysm, evaluated with multimodal imaging, including color fundus photography, FA, IA, SD-OCT, and OCTA, and then treated with anti-VEGF therapy, photodynamic therapy, and direct photocoagulation. The limitations of this paper include the use of a single case report and a relatively short post-treatment observational period. Long-term observations are necessary to elucidate the pathophysiology and confirm the treatment outcomes of this condition. Another limitation includes we did not measure the scleral thickness which may have a role in the pathogenesis of pachychoroid spectrum disease including PCV.14

In conclusion, we report the clinical features and course of a rare case of choroidal macroaneurysm in a patient with a presumed anomalous short posterior ciliary artery. Multimodal imaging was useful for the diagnosis and observation of this condition. This could be a differential diagnosis for polypoidal choroidal vasculopathy. When visual impairment develops due to exudative change in this condition, direct photocoagulation for choroidal macroaneurysm may be the most appropriate treatment option.

**Funding**

This study received no funding or grant support.

**Authorship**

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

**Declaration of competing interest**

None of the authors have any financial disclosures.

**Acknowledgements**

None.

**Appendix A. Supplementary data**

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2022.101755.
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