

Paracentral Acute Middle Maculopathy Associated With Peripheral Retinal Capillary Hemangioma

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Case Report

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Abstract

Background

Paracentral acute middle maculopathy (PAMM) has not previously been reported in association with peripheral retinal capillary hemangioma (RCH). We report a novel case of PAMM associated with a peripheral RCH, illustrating a venous congestion–driven ischemic mechanism.

Case presentation:

A 36-year-old woman presented with a superior paracentral visual field defect in the right eye with 20/20 vision. Fundus examination revealed multifocal parafoveal whitish lesions and an orange-red capillary hemangioma with surrounding hemorrhages in the inferotemporal peripheral retina, accompanied by venous dilation and tortuosity. Spectral-domain optical coherence tomography (OCT) demonstrated a hyperreflective band involving the inner plexiform, inner nuclear, and outer plexiform layers, consistent with PAMM. OCT angiography showed focal hypoperfusion at the deep capillary plexus with preserved superficial plexus. Ultra-widefield fluorescein angiography revealed early arterial filling, delayed venous drainage, and late peritumoral leakage, suggesting localized venous outflow impairment.

Conclusions

This case broadens the clinical spectrum of RCH-related complications by demonstrating an association between peripheral RCH, localized venous congestion, and PAMM, highlighting the importance of careful vascular assessment and longitudinal surveillance.

Background

Retinal capillary hemangioma (RCH) is a benign vascular hamartoma typically arising in the peripheral retina, occurring sporadically or with von Hippel-Lindau disease [1, 2]. Although histologically benign, RCH threatens vision through exudation, macular edema, and tractional detachment [2, 3]. RCH may also impair vision through secondary vascular complications. Retinal vein occlusion has been reported with juxtapapillary RCH, but venous complications from peripheral RCH remain poorly characterized [4].

Paracentral acute middle maculopathy (PAMM) is an optical coherence tomography (OCT)-defined entity characterized by hyperreflectivity predominantly involving the inner nuclear layer, reflecting deep capillary plexus (DCP) ischemia [5]. PAMM is commonly associated with retinal vein occlusion [6–11]. Because the DCP lies within the venous outflow territory, it is vulnerable to hypoperfusion with venous congestion [12, 13].

PAMM has not been reported with peripheral RCH. We describe a unique case of peripheral RCH complicated by localized venous outflow impairment and PAMM. Using multimodal imaging, we

illustrate a plausible venous congestion-to-deep capillary ischemia cascade, broadening the recognized spectrum of RCH-related complications.

Case presentation

A 36-year-old woman presented with a 1-week history of superior paracentral visual field defect in the right eye. She had no systemic disease, recent vaccination, medication use, or prior ocular surgery. Best-corrected visual acuity was 20/20 bilaterally.

Ultra-widefield (UWF) color fundus photography (Optos California, Optos plc, Dunfermline, UK) revealed subtle multifocal parafoveal whitish lesions. Dilated tortuous veins were observed along the inferotemporal arcade, and a well-demarcated orange-red RCH with surrounding intraretinal hemorrhages was identified in the inferotemporal peripheral retina. Spectral-domain optical coherence tomography (SD-OCT; Spectralis OCT, Heidelberg Engineering, Heidelberg, Germany) demonstrated a band-like hyperreflective lesion involving the inner plexiform layer, inner nuclear layer, and outer plexiform layer, consistent with PAMM.

Optical coherence tomography angiography (OCTA) was acquired using AngioVue (Optovue Inc., Fremont, CA, USA) for 6 × 6-mm macular scan. OCTA showed focal flow deficit at the DCP corresponding to the PAMM lesion, with relative preservation of the superficial plexus. UWF fluorescein angiography (UWF-FA; Optos California, Optos plc, Dunfermline, UK) revealed early filling of a feeder arteriole, delayed filling and late leakage of the draining vein, consistent with localized venous outflow impairment. Three coexisting conditions were identified: peripheral RCH, localized venous outflow impairment, and PAMM.

Systemic evaluation for von Hippel-Lindau disease, including brain/spinal MRI, abdominal imaging, and genetic testing, detected no abnormalities except a small hepatic hemangioma requiring observation.

The peripheral RCH was treated with focal argon laser photocoagulation in four sessions over two months. At one-month follow-up, UWF fundus photography showed photocoagulation scars with reduced hemorrhages and mild improvement in venous congestion. SD-OCT showed PAMM evolution with resolution of the hyperreflective band and subsequent focal inner nuclear layer thinning without macular edema. Visual acuity remained 20/20. At six-month follow-up, vision remained stable, hemorrhages resolved, and no additional vascular complications occurred.

Discussion and Conclusions

RCH is a benign vascular hamartoma that most commonly arises in the peripheral retina but can lead to significant visual morbidity through both direct and indirect mechanisms [1]. Direct effects include exudation, secondary macular edema, and tractional changes, whereas indirect effects are mediated by tumor-related alterations in retinal hemodynamics [2, 3, 14]. Although retinal vein occlusion has been described in association with juxtapapillary RCH, vascular complications related to peripheral RCH remain incompletely characterized [4].

In the present case, multimodal imaging demonstrated localized venous outflow impairment downstream from a peripheral RCH, accompanied by PAMM. PAMM is an OCT-defined manifestation of ischemia involving the middle retinal layers, most prominently the inner nuclear layer, and is increasingly recognized as a marker of DCP hypoperfusion [5, 12]. Because the DCP lies predominantly within the venous outflow territory, it is particularly vulnerable to ischemia in conditions associated with venous congestion or elevated intraluminal venous pressure, even when superficial capillary perfusion is relatively preserved [13].

The angiographic findings in this case—early filling of the feeder arteriole combined with delayed venous drainage and late leakage—suggest a tumor-related arteriovenous shunt–like hemodynamic configuration. Such a high-flow, low-resistance vascular circuit may impose chronic pressure overload on the draining vein, leading to venous congestion and impaired capillary transit. Reduced capillary transit time can, in turn, compromise oxygen delivery to the distal capillary beds, disproportionately affecting the DCP and the inner nuclear layer–outer plexiform layer watershed zone [13, 15]. This hemodynamic cascade provides a plausible explanation for the development of PAMM observed in our patient.

The structural evolution of PAMM observed on follow-up OCT in this case—resolution of the acute hyperreflective band followed by focal thinning and atrophy of the inner nuclear layer—is consistent with the established natural history of PAMM reported in association with retinal venous disorders [7, 12]. Despite preserved central visual acuity, such structural changes may correspond to persistent paracentral scotomas and underscore the clinical relevance of recognizing PAMM as a marker of retinal ischemia rather than a transient imaging finding.

Management of RCH aims to control tumor activity while minimizing secondary vascular and ischemic complications [2, 3]. In the present case, focal argon laser photocoagulation was applied to reduce tumor-related vascular flow, resulting in regression of intraretinal hemorrhages and partial improvement in venous congestion. Although causality cannot be definitively established from a single case, the temporal and anatomic correlations observed here support a mechanistic link between RCH-related venous congestion and secondary DCP ischemia.

This report has limitations inherent to a single observational case without direct hemodynamic measurements. Nevertheless, it broadens the clinical spectrum of RCH-related complications by highlighting PAMM as a potential downstream manifestation of tumor-associated venous outflow impairment. Multimodal imaging plays a central role in elucidating such pathophysiologic relationships and may facilitate earlier recognition of ischemic sequelae in patients with RCH, even in the absence of systemic risk factors.

In summary, this case documents a peripheral RCH complicated by localized venous outflow impairment and PAMM. Multimodal imaging findings support a plausible venous congestion–to–DCP ischemia cascade, broadening the recognized spectrum of RCH-related complications. Recognition of this mechanism underscores the importance of careful vascular assessment and longitudinal surveillance in patients with RCH, even in the absence of systemic risk factors.

Abbreviations

DCP

Deep capillary plexus

IRB

Institutional Review Board

MRI

Magnetic resonance imaging

OCT

Optical coherence tomography

OCTA

Optical coherence tomography angiography

PAMM

Paracentral acute middle maculopathy

RCH

Retinal capillary hemangioma

SD-OCT

Spectral-domain optical coherence tomography

UWF

Ultra-widefield

UWF-FA

Ultra-widefield fluorescein angiography

Declarations

Ethics approval and consent to participate

This study was conducted in accordance with the tenets of the Declaration of Helsinki. The study protocol was approved by the Institutional Review Board of Yeungnam University Hospital (IRB No. 2024-03-039). Written informed consent was obtained from the patient.

Consent for publication

Written informed consent for publication of the clinical details and accompanying images was obtained from the patient.

Availability of data and materials

All the data supporting our findings is contained within the manuscript.

Competing interests

SJ and CL declare that they have no competing interests. MS reports honoraria from Allergan, Bayer, Curacle, Novartis, Samsung Bioepis, and Roche for lecture fees, consultancy, and board membership, and reports research grants from Allergan, Bayer, and Novartis.

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Authors' contributions

MS contributed to the design and concept of the work. SJ and CL contributed to data acquisition, analysis, and interpretation. SJ drafted the manuscript. MS supervised the study and revised the manuscript. All authors read and approved the final manuscript.

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References

1. Singh AD, Shields CL, Shields JA. von Hippel-Lindau disease. *Surv Ophthalmol*. 2001;46(2):117–42.
2. Webster AR, Maher ER, Moore AT. Clinical characteristics of ocular angiomas in von Hippel-Lindau disease and correlation with germline mutation. *Arch Ophthalmol*. 1999;117(3):371–8.
3. Singh AD, Nouri M, Shields CL, Shields JA, Perez N. Treatment of retinal capillary hemangioma. *Ophthalmology*. 2002;109(10):1799–806.
4. AlBloushi AF, Taskintuna I, Nowilaty SR. Retinal capillary hemangioblastoma and hemiretinal vein occlusion in a patient with primary congenital glaucoma: A case report. *Saudi J Ophthalmol*. 2019;33(4):401–4.
5. Sarraf D, Rahimy E, Fawzi AA, Sohn E, Barbazetto I, Zacks DN, Mittra RA, Klancnik JM Jr., Mrejen S, Goldberg NR, et al. Paracentral acute middle maculopathy: a new variant of acute macular neuroretinopathy associated with retinal capillary ischemia. *JAMA Ophthalmol*. 2013;131(10):1275–87.
6. Casalino G, Williams M, McAvoy C, Bandello F, Chakravarthy U. Optical coherence tomography angiography in paracentral acute middle maculopathy secondary to central retinal vein occlusion. *Eye (Lond)*. 2016;30(6):888–93.
7. Rahimy E, Sarraf D, Dollin ML, Pitcher JD, Ho AC. Paracentral acute middle maculopathy in nonischemic central retinal vein occlusion. *Am J Ophthalmol*. 2014;158(2):372–e380371.
8. Zhang Z, Jiang Y, Huang X, Wu Z, Ke B. Clinical Characteristics of Paracentral Acute Middle Maculopathy in Eyes with Retinal Vascular Occlusion Diseases in Chinese Patients. *J Ophthalmol*. 2021;2021:8867570.

9. Abrishami M, Hosseini SM, Shoeibi N, Heidarzadeh HR. Paracentral Acute Middle Maculopathy and Nonischemic Central Retinal Vein Obstruction in a Young Patient with Protein C Deficiency. *Case Rep Ophthalmol Med.* 2022;2022:1237148.
10. Parodi MB, Arrigo A, Rajabian F, Tecilazich F, Giustina A, Bandello F, Jampol LM, PARACENTRAL ACUTE MIDDLE MACULOPATHY IN CENTRAL RETINAL VEIN OCCLUSION COMPLICATING AMYLOID LIGHT-CHAIN AMYLOIDOSIS. *Retin Cases Brief Rep.* 2022;16(5):543–6.
11. Takai Y, Sr., Inoue K. A Case of Impending Central Retinal Vein Occlusion Associated With Antiphospholipid Antibody Syndrome Complicated by Paracentral Acute Intermediate Layer Maculopathy. *Cureus.* 2023;15(4):e37256.
12. Rahimi E, Kuehlewein L, Sadda SR, Sarraf D. Paracentral Acute Middle Maculopathy: What We Knew Then and What We Know Now. *Retina.* 2015;35(10):1921–30.
13. Scharf J, Freund KB, Sadda S, Sarraf D. Paracentral acute middle maculopathy and the organization of the retinal capillary plexuses. *Prog Retin Eye Res.* 2021;81:100884.
14. Qin XJ, Huang C, Lai K. Retinal vein occlusion in retinal racemose hemangioma: a case report and literature review of ocular complications in this rare retinal vascular disorder. *BMC Ophthalmol.* 2014;14:101.
15. Yu DY, Cringle SJ. Oxygen distribution and consumption within the retina in vascularised and avascular retinas and in animal models of retinal disease. *Prog Retin Eye Res.* 2001;20(2):175–208.

Figures

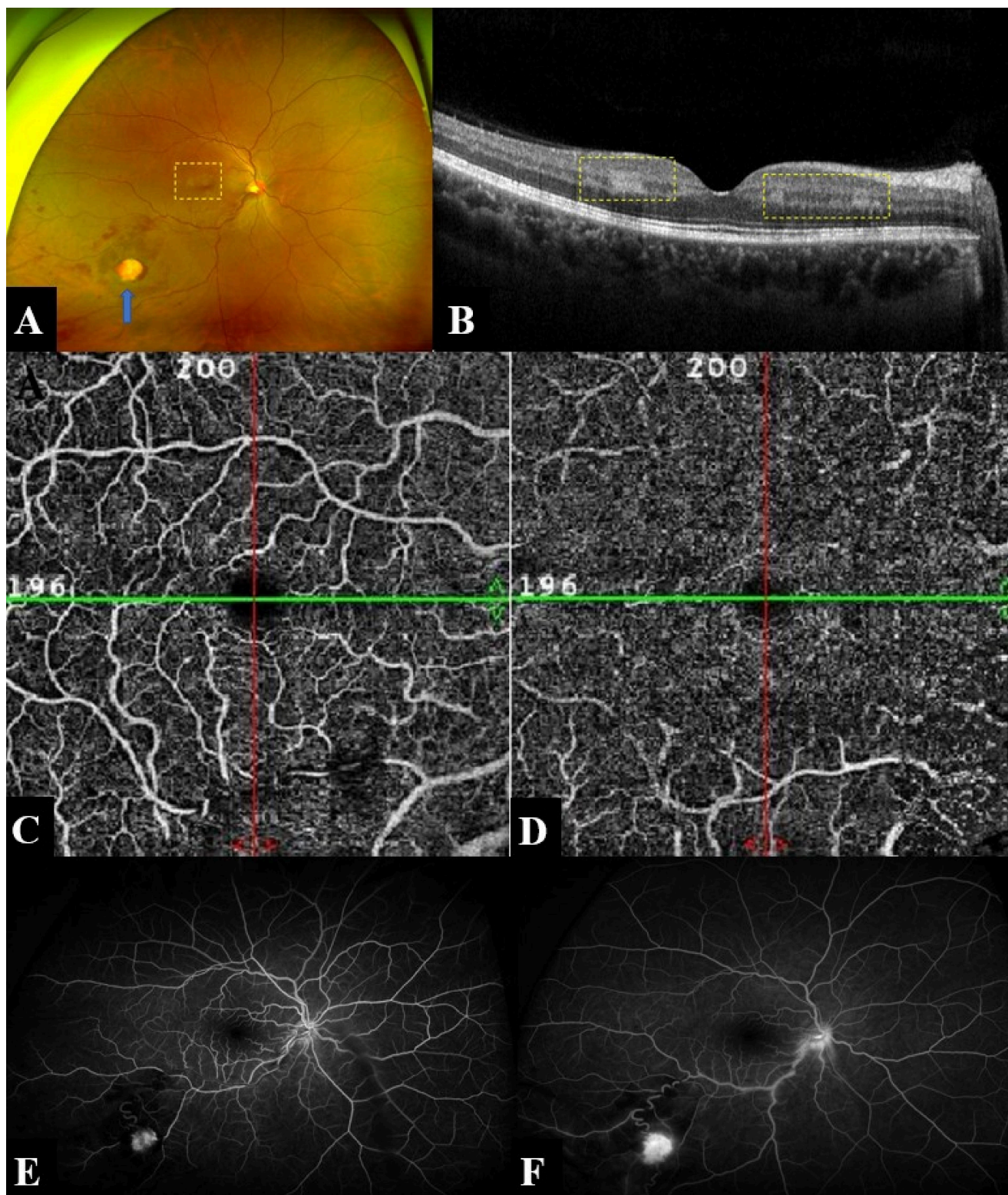


Figure 1

Multimodal imaging findings at presentation.

(A) Ultra-widefield (UWF) color fundus photograph of the right eye shows subtle multifocal parafoveal whitish lesions (boxed), dilated and tortuous veins along the inferotemporal arcade, and a well-demarcated orange-red retinal capillary hemangioma (RCH) with surrounding intraretinal hemorrhages in the inferotemporal peripheral retina (arrows). (B) Spectral-domain optical coherence tomography (OCT)

shows a band-like hyperreflective lesion involving the inner plexiform layer, inner nuclear layer, and outer plexiform layer corresponding to the parafoveal lesion, consistent with paracentral acute middle maculopathy (PAMM). The foveal contour is preserved, with no intraretinal or subretinal fluid. (C, D) OCT angiography demonstrating selective deep capillary plexus (DCP) ischemia. The 6 × 6-mm macular images show relative preservation of the superficial capillary plexus (C) with a focal flow deficit at the level of the DCP corresponding to the PAMM lesion (D). (E, F) UWF fluorescein angiography demonstrating tumor-related venous outflow impairment. Early-phase fluorescein angiography (E) shows early filling of a feeder arteriole supplying the RCH. The late-phase image (F) demonstrates delayed filling and late leakage of the draining vein extending toward the inferotemporal arcade, consistent with localized venous outflow impairment.

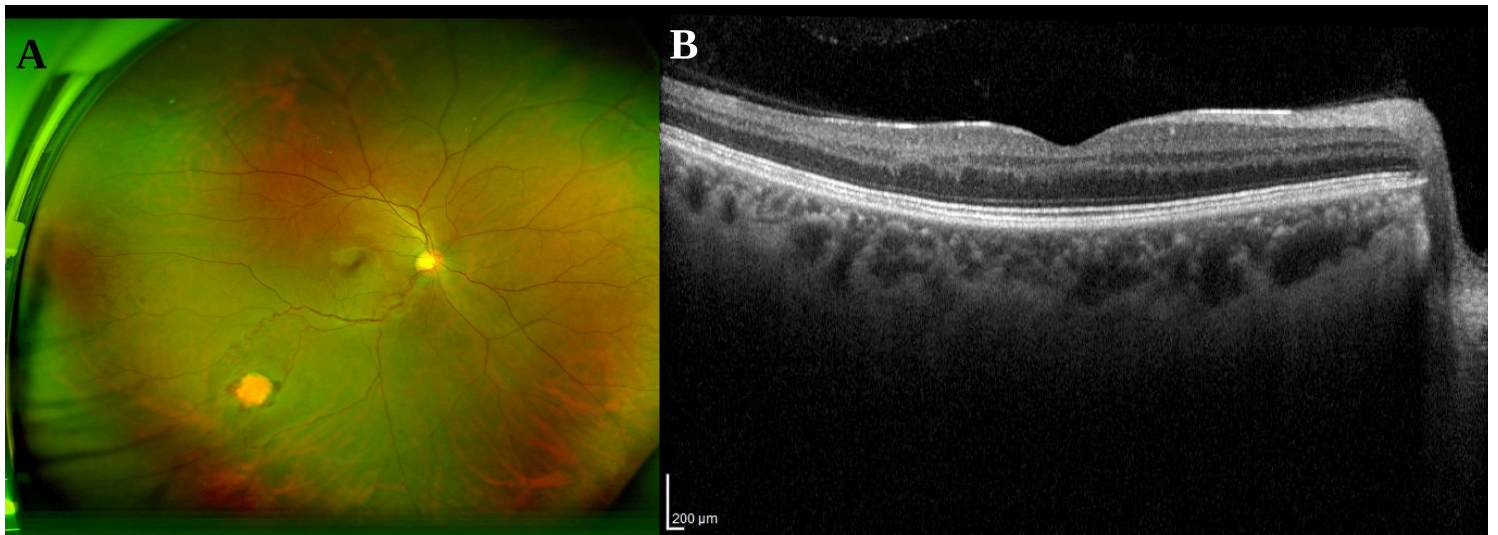


Figure 2

Follow-up findings after laser photocoagulation. (A) UWF color fundus photograph obtained at 1 month showing discrete photocoagulation scars surrounding the RCH with a reduction in adjacent intraretinal hemorrhages and mild improvement in venous tortuosity and congestion. (B) Spectral-domain OCT demonstrating interval evolution of PAMM, with resolution of the acute hyperreflective band and subsequent focal thinning and atrophy of the inner nuclear layer, without evidence of macular edema.