

# Paracentral acute middle maculopathy secondary to high intraocular pressure: a case report

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**Dear Editor,**

Paracentral acute middle maculopathy (PAMM), defined as an optical coherence tomography (OCT) finding characterized by a band-like, hyperreflective lesion at the level of the inner nuclear layer (INL), is the result of impaired flow in the deep and intermediate retinal capillary plexuses<sup>[1]</sup>. It has been demonstrated that PAMM was related to various retinal vascular disorders, including central retinal vein occlusion, retinal artery occlusion, diabetic retinopathy, and hypertensive retinopathy<sup>[2]</sup>. We are writing to report a case of PAMM with acute primary angle-closure glaucoma, which suggested that PAMM can also be caused by acute intraocular pressure (IOP) elevation-induced ischemia in the retina. Our findings may provide new insight into the pathogenesis of PAMM.

**Ethical Approval** This study was approved by the Review Board of the Second Hospital of Jilin University. Written informed consent to participate and allow publication was obtained from the patient.

## CASE REPORT

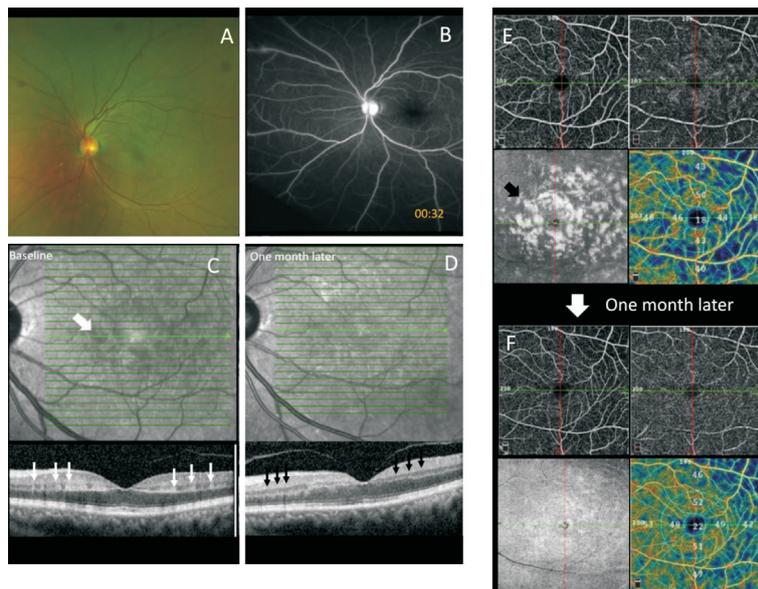
A 64-year-old female came to our hospital with vision deterioration in the left eye that persisted for one week. One week prior, she was diagnosed with an acute angle-closure glaucoma attack and treated with carteolol, brinzolamide and brimonidine tartrate to lower the IOP. She denied a history of hypertension or diabetes.

On presentation, her best-corrected visual acuity was 0.5 in the right eye, and finger counting was 15 cm in the left eye. The IOP of the right eye was 17 mm Hg, and that of the left eye was 21 mm Hg. Fundus examination revealed multiple localized roundish microhemorrhages within the posterior pole of the left eye (Figure 1A). Ultrasound biomicroscopy (UBM) of the left eye revealed complete anatomic obliteration of the trabeculum by the base of the iris (Figure 2D). A complete multimodal imaging study, including fundus fluorescein angiography (FFA), near-infrared reflectance imaging, OCT and optical coherence tomography angiography (OCTA), was carried out. FFA showed a small amount of fluorescein leakage in the superior nasal vein branch of the left eye (Figure 1B). Paracentral hyporeflective macular lesions were visible on near-infrared reflectance images of the left eye and the corresponding OCT B-scans illustrated multiple hyperreflective plaque-like lesions involving the INL in a skip pattern (Figure 1C). OCTA demonstrated perifoveal loss of capillary density of both the superficial and deep capillary plexuses in the left eye. En face OCT images illustrated a remarkable spectrum of fern-like perivenular (Figure 1E). The diagnosis of PAMM was confirmed with the above examination.

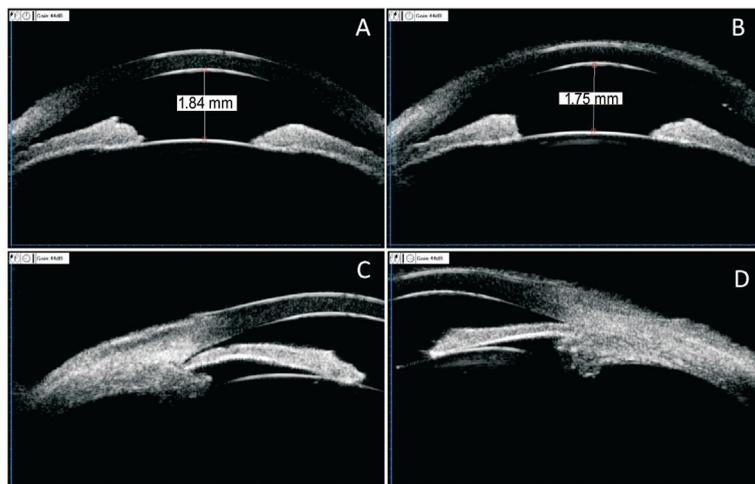
The patient underwent phacoemulsification combined with trabeculectomy surgery and received drug treatment (hypodermic injection of compound anisodine around the superficial temporal artery; oral ginkgo biloba hevert) to improve microcirculation. A month later, her best-corrected visual acuity of the left eye was 0.05, and the IOP was 14 mm Hg. Fundus examination showed roundish microhemorrhages in the posterior pole of the left eye that had been absorbed. OCT showed resolution of the hyperreflective lesions and sequelae of INL thinning and an irregular, attenuated outer plexiform layer (Figure 1D). OCTA showed partial recovery of the density of the capillary network in the left eye, and the perivenular fern-like pattern in the en face OCT disappeared (Figure 1F).

## DISCUSSION

PAMM is an infarct lesion in the INL of the retina caused by insufficient perfusion in the intermediate and deep retinal capillary plexuses. The typical PAMM lesions can be detected by OCT. Initially, the level of the INL in the paracentral area



**Figure 1 The imaging examination of the patient** A: Fundus photograph demonstrated multiple localized roundish microhemorrhages within the posterior pole. B: FFA showed a small amount of fluorescein leakage in the superior nasal vein. C, D: Near-infrared reflectance images showed paracentral hyporeflective macular lesions (white arrow); OCT B-scans illustrated multiple hyperreflective plaque-like lesions in the INL (white arrows). One month later, OCT showed resolution of the hyperreflective lesions and sequelae of INL thinning and an irregular, attenuated outer plexiform layer (black arrows). E, F: OCTA demonstrated perifoveal loss of capillary density of both the superficial and deep capillary plexuses. En face OCT images illustrated a remarkable spectrum of fern-like perivenular (black arrow). One month later, OCTA showed partial recovery of the density of the capillary network in the left eye, and the perivenular fern-like pattern in the en face OCT disappeared.



**Figure 2 UBM images showing the anterior chamber structures** A, B: Shallow anterior chamber in both eyes; C: Narrow anterior chamber angle and iris bombe in the right eye; D: Complete angle closure with straight iris conformation in the left eye.

manifests as hyper-reflective band lesions during the acute phase. Finally, a sequela of INL thinning develops due to the atrophy of tissue infarction as the PAMM lesions resolve<sup>[2-3]</sup>. At present, there is no clear and standardized treatment plan for PAMM patients. Because the disease is usually considered to be associated with a variety of retinal vascular diseases and systemic diseases, the treatment of the primary vascular disease and systemic high-risk factors is the most important intervention<sup>[2,4]</sup>. Ocular perfusion pressure is determined by subtracting the IOP from the mean blood pressure in the ophthalmic artery.

Therefore, ocular perfusion pressure can be decreased by an increase in IOP. In healthy humans, the autoregulation of retinal blood flow is entirely efficient only if the perfusion pressure is lowered by less than 50%, which means IOP cannot exceed 27-30 mm Hg<sup>[5]</sup>. If the IOP is elevated more than 20 mm Hg for a duration of 5min, retinal blood flow is decreased<sup>[6]</sup>. In the case presented here, the elevation in IOP in the acute attack of glaucoma exceeded the retinal vascular autoregulation ability, which caused global ocular perfusion impairment. On the other hand, the deep capillary plexus usually maintains a lower perfusion pressure since

it only receives arteriolar inflow from the intermediate capillary plexus and has no other significant arterial supply<sup>[7]</sup>. In addition, due to the higher metabolic activity, the middle retina may consume more oxygen<sup>[8]</sup>. As a result, a selective infarction occurs in the INL or the middle retina because of the global insufficiency of blood flow across the retinal capillary plexuses<sup>[2]</sup>. In the case presented here, elevated IOP resulted in impaired global perfusion, which eventually led to infarction in the INL. To the best of our knowledge, this is not the first reported case of PAMM secondary to elevated IOP. Aribas *et al*<sup>[9]</sup> also reported PAMM in a patient with primary congenital glaucoma who had an increase in IOP before being diagnosed with PAMM. The authors believed that PAMM was caused by elevated IOP, which led to ischemia in the deep retinal plexus, and they suggested that PAMM was a factor contributing to permanent vision loss in the patient with acute IOP elevation secondary to primary congenital glaucoma.

The pathogenesis of PAMM is generally believed to be related to retinal vascular diseases, especially retinal vein occlusion. In the case presented here, typical central retinal vein occlusion (CRVO) findings were not observed. The localized multiple roundish microhemorrhages were considered to be caused by ocular decompression retinopathy. Ocular decompression retinopathy may be caused by a loss of autoregulation, which allows retinal capillaries to maintain a constant perfusion pressure. When autoregulation ability is lost, a transient decrease in IOP can result in an increased flow because of the reduced retinal arterial resistance and lead to leakage through the already fragile capillaries<sup>[10]</sup>. In the case presented here, PAMM was caused by a decrease in retinal vascular autoregulation that led to retinal blood flow perfusion damage under high IOP conditions. When IOP decreased, ocular decompression retinopathy occurred because of retinal vascular reperfusion.

In conclusion, ischemia-reperfusion injury caused by acute elevated IOP may be an important risk factor for PAMM. More studies are necessary to elucidate the etiology of PAMM to prevent permanent vision loss in these patients.

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