Paracentral acute middle maculopathy secondary to retinal artery macroaneurysm

Paracentral acute middle maculopathy (PAMM) is an optical coherence tomography (OCT) finding defined by the presence of a hyperreflective band at the level of the inner nuclear layer (INL), indicating INL infarct secondary to hypoperfusion of the deep vascular complex.[1,2] Patients present with acute paracentral scotoma that can be permanent.[1,3] Common etiologies include primary retinal vascular diseases such as retinal artery or vein occlusions, diabetic and hypertensive retinopathies, medications (e.g., vasopressors), and others.[4] We report a novel observation of a ruptured retinal artery macroaneurysm (RAM) with secondary incomplete branch retinal artery occlusion (BRAO) that resulted in PAMM.

A 72-year-old female presented with a 3-month history of sudden-onset left paracentral scotoma (**Fig. 1A**). Her medical history included hypertension and hyperlipidemia. Visual acuity was 20/25 and 20/150 OD and OS, respectively. Anterior-segment examination and intraocular pressures were unremarkable.

A ruptured RAM was seen along a second-order artery at the inferotemporal arcade. A small area of deep retinal whitening along the distal branches of this artery also was observed (**Fig. 1B**).

Corresponding OCT showed a round superficial retinal hyperreflective lesion consistent with RAM (**Fig. 2A**) and a hyperreflective band involving the outer plexiform layer (OPL), INL, and inner plexiform layer (IPL) consistent with PAMM (**Fig. 2B**).

Confocal scanning laser ophthalmoscopy dynamic fluorescein angiogram demonstrated a ruptured and hemorrhagic RAM with significantly delayed distal filling of the artery, which represented an incomplete BRAO. This was accompanied by resultant PAMM in an area supplied by the tributary arterioles of the affected artery (**Fig. 2C**).

Oxygen consumption is highest at the metabolically active middle retinal layer. The inner and middle retinal layers are supplied collectively by the superficial capillary plexus in the ganglion cell layer, intermediate capillary plexus (ICP) in the INL-IPL boundary, and deep capillary plexus (DCP) in the INL-OPL boundary. Schraf et al.[1] suggested that the middle retina (OPL-INL-IPL) is particularly prone to ischemic insult for 2 reasons.[3] First, it serves as a site for major venous outflow from the DCP (major) and ICP (versus the superficial capillary plexus, where major arterial inflow occurs), resulting in lower oxygen saturations.[2] Second, it receives arteriolar inflow solely from the ICP (the superficial and deep retinal layers), lacking major arterial supply. As such, it maintains a lower perfusion pressure and is susceptible to states of global insufficiency.[4]

Infarction of the middle retinal layer, resulting in PAMM, may occur selectively because of the combined effects of decreased perfusion pressure, decreased oxygen saturation, and increased metabolic oxygen demands.

Our patient had hypertension, which predisposed her to developing RAM.[4] Rupture and subsequent thrombosis of the RAM[5] resulted in mechanical narrowing of the vessel lumen and severe flow limitation distally, leading to an incomplete BRAO. This was consistent with the fluorescein angiogram findings of delayed filling in the distal portion of the affected artery. Flow limitation resulted in decreased blood supply to the retinal capillary plexuses and further decreased perfusion pressure, thereby disrupting the delicately balanced ICP and DCP blood supply of the middle retinal layer for reasons discussed earlier.[1,4] Middle retinal layer infarction occurs most readily and presented as PAMM in our patient.

Although the association of BRAO with PAMM has been described previously, this case merits discussion because the association of RAM with PAMM is novel and

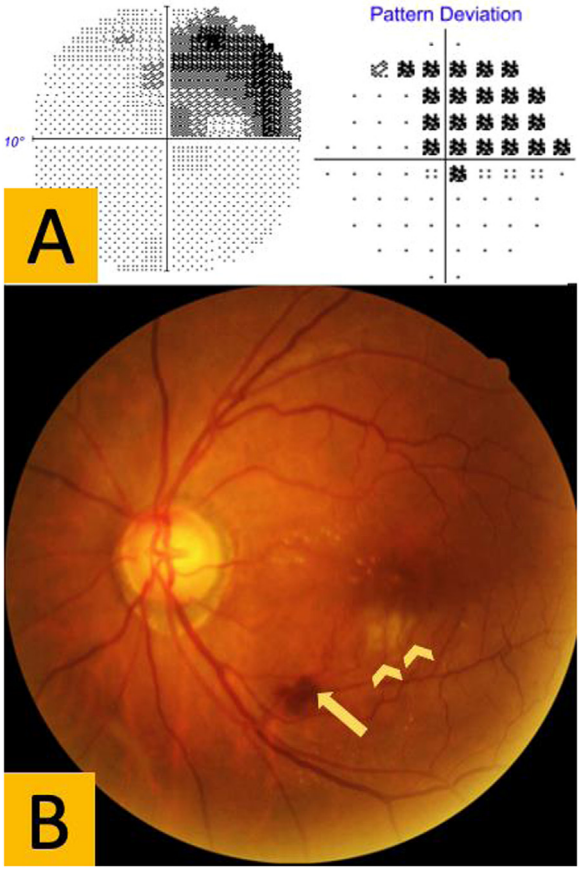

Fig. 1—(A) OS 10-2 automated static visual field assessment reveals a paracentral scotoma in the superonasal quadrant. (B) OS fundus photograph reveals the presence of a ruptured retinal artery macroaneurysm with surrounding intraretinal hemorrhage (yellow arrow) and small area of deep retinal whitening (yellow arrowheads) along the distal arteriolar branches.

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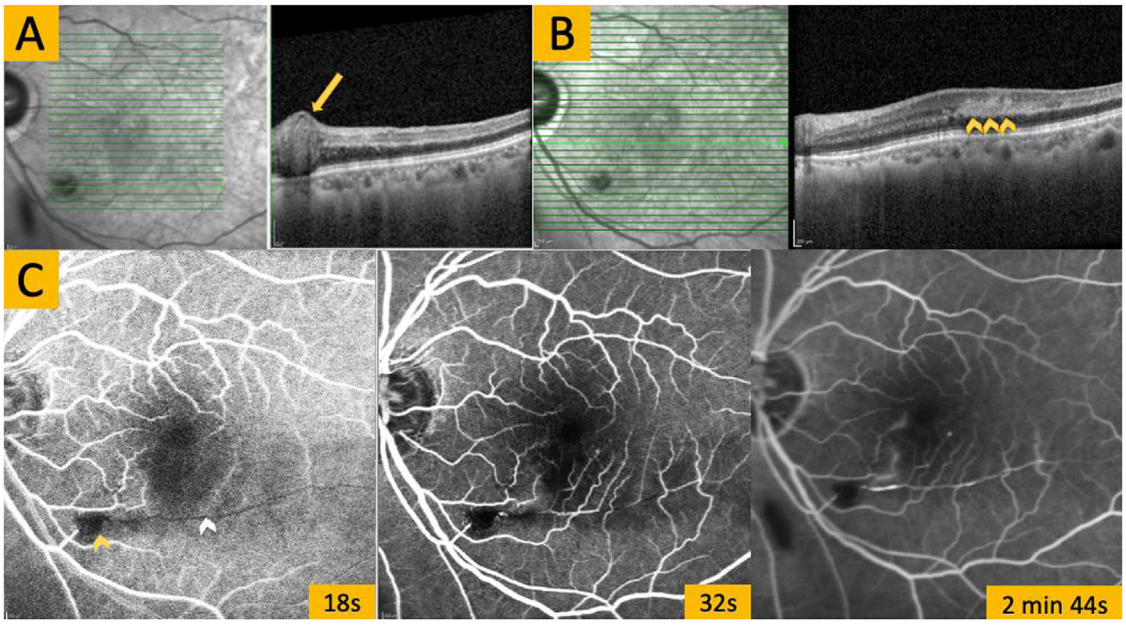

Fig. 2—(A) OS retinal artery macroaneurysm appears a round superficial retinal hyperreflectivity (yellow arrow). (B) OS band of hyperreflectivity (yellow arrowheads) in the outer plexiform layer, inner nuclear layer, and inner plexiform layer consistent with middle retinal ischemia and paracentral acute middle maculopathy. (C) Confocal scanning laser ophthalmoscopy video fluorescein angiogram shows delayed filling of artery (white arrowhead) and its branches supplying the area just inferior to the fovea at 18 seconds distal to the area of blocked hypofluoresence (yellow arrowhead) from the ruptured retinal artery macroaneurysm and hemorrhage. At 32 seconds, there is partial filling of the artery, but patchy vascular filling defects are seen in the area just inferior to the fovea. Finally, at 2 minutes and 44 seconds, the artery filling is completed. The area of paracentral acute middle maculopathy (just inferior to the fovea) shows mild hypofluorescence and is clearly seen to be supplied by the tributary arterioles of the affected artery.

has not yet been reported. Additionally, the clinical and imaging findings were consistent with and clearly elucidated the anatomy and physiology of the retinal capillary plexuses as well as the pathophysiology of PAMM.

Eunice Jin Hui Goh, Wei Kiong Ngo
National Healthcare Group Eye Institute, Singapore.

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Correspondence to Wei Kiong Ngo, MD; wkngo@hotmail.com.

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Footnotes and Disclosure

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