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LETTER TO THE EDITOR

Paracentral Acute Middle Maculopathy Revealing a Giant Aortic Aneurysm

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ABSTRACT

Background: This report describes a case with paracentral acute middle maculopathy revealing a giant aortic aneurysm.

Case Description: A 41-year-old man presented with unilaterally unexplained sudden vision loss in the left eye. Fundoscopy revealed a slightly pale r -26etinal reflex in the superior retinal hemifield. SDOCT revealed blurred boundaries between retinal segments and prominent edema in the middle retinal layers. OCTA revealed decreased vascular density in the deep capillary plexus. FA showed low arterial circulation mainly in the superior retinal hemifield. Humphrey VFT revealed an inferior altitudinal defect in the left eye, implying a related circulatory disturbance in the corresponding region.

A thorough systemic work-up, including a cardiovascular etiological investigation, revealed giant aortic aneurysm in the ascending aorta, necessitating emergency surgery.

Conclusion: This report highlights the importance of a systemic etiological investigation in patients with PAMM to rule out any potential cardiovascular issues.

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Aortic aneurysm; optical coherence tomography angiography; paracentral acute middle maculopathy; spectral domain optical coherence tomography; visual field test

Intoduction

Paracentral acute middle maculopathy (PAMM) is characterized by a hyperreflective band that initially spans the inner nuclear layer (INL). At more advanced stages of the disease, INL atrophy is observed. Although PAMM pathophysiology is uncertain, ischemia has been shown to play a significant role at the intermediate capillary plexus (ICP) and deep capillary plexus (DCP) levels.[1,2] The current report describes a patient with PAMM revealing a giant aortic aneurysm.

Case description

A 41-year-old man complained of unilateral unexplained sudden onset of vision loss in the left eye (LE) that began two days earlier. In addition to an unremarkable ocular history, no associated systemic conditions were revealed in the patient's general medical history. Visual acuity in decimal was 1.0 in the right eye (RE) and counting fingers from two meters in the LE. Normal ocular movements were observed. Anterior segment evaluation was unremarkable. The Ishihara test showed 24/24 and 0/24 color vision in the RE and LE, respectively. Intraocular pressure was 15 mmHg in the RE and 13 mmHg in the LE. A dilated fundoscopy revealed a slightly pale retinal reflex in the LE, prominently located in the superior retinal hemifield. However, the RE fundoscopy and spectral domain optical coherence tomography (SDOCT) analyses were unremarkable.

The boundaries between retinal segments in the LE fundus were indistinct during SDOCT scanning, and the middle retinal layers had significant edema due to ischemia (Figure 1).

The RE had normal superficial capillary plexus (SCP) and DCP on OCT angiography (OCTA). The DCP was associated with a significant decrease in vascular density in the LE, despite the existence of a normal SCP (Figure 2a,b). Superior retinal hemifield had poor arterial circulation on fluorescein angiography (FA) (Figure 3). A Humphrey visual field test (VFT) (Central 30–2 threshold test) revealed an inferior altitudinal defect in the LE, which correlated with the circulatory disturbance depicted in FA (Figure 4).

A thorough systemic investigation was done, including, but was not limited to, potential cerebrovascular events and other central nervous system associated disorders. No abnormalities in the nervous system were discovered. The results of a systematic investigation, which included hemogram and biochemical tests, thrombophilia screening, carotid and vertebral artery Doppler ultrasound, and cranioorbital magnetic resonance imaging, are summarized in Table 1.

An echocardiogram conducted by a cardiologist to rule out any possible sources of thromboembolism or other cardiovascular disorders revealed a giant 97 × 89 mm aortic aneurysm in the ascending aorta, requiring emergency surgery (Figure 5a,b).

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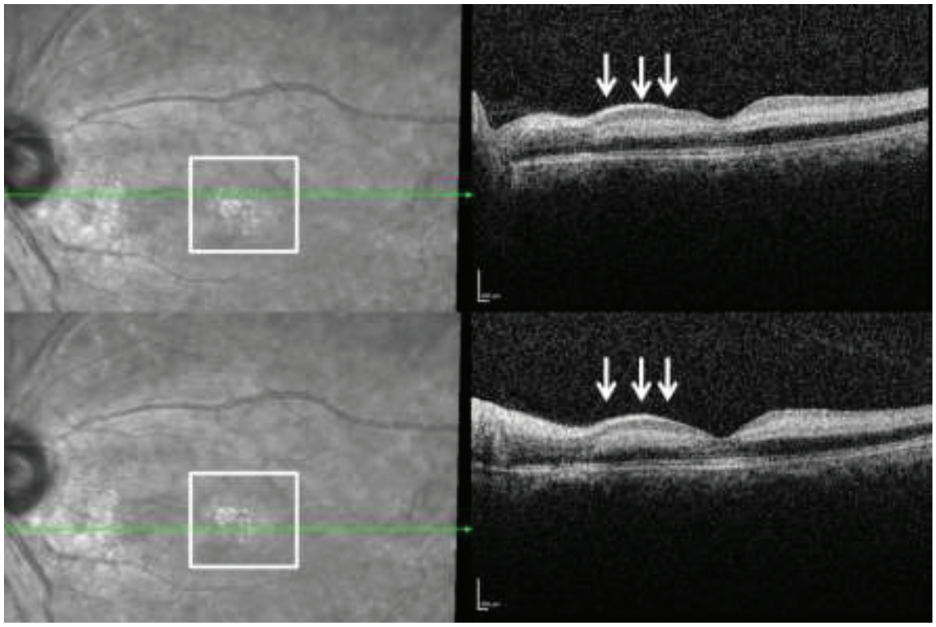

Figure 1. An SDOCT scan of a PAMM patient's left eye reveals a placoid, hyperreflective band at the level of the inner nuclear layer (white arrows), with the outer retina relatively spared. There are noticeable indistinct boundary patterns between retinal segments, as well as severe edema in the middle retinal layers due to ischemia. There is also a broad hyporeflective area over the affected region with PAMM (white box).

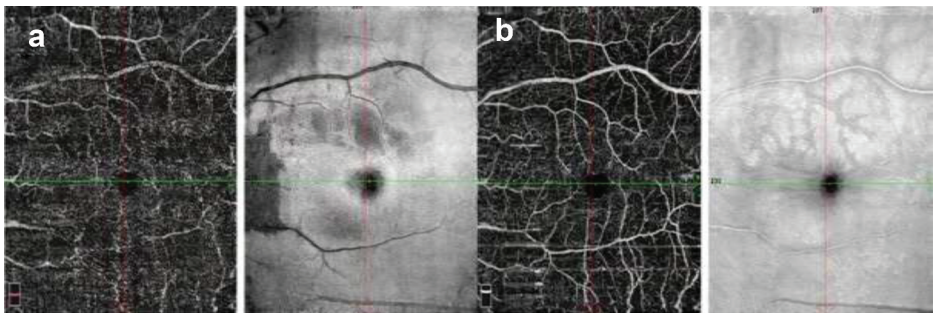

Figure 2. An OCTA of a PAMM patient's left eye. The SCP is largely unaffected (b), but en face projection at the DCP level reveals a relatively reduced vascular density. Also, superficial retinal vessels are seen as projection artifacts (a).

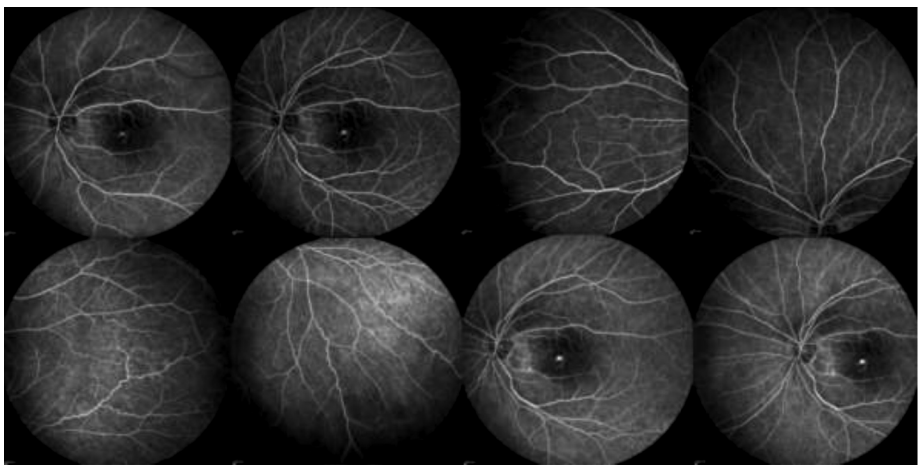

Figure 3. Low arterial circulation in the superior retinal hemifield during the early and late FA phases.

Discussion

Paracentral acute middle maculopathy has been linked to changes in the ICP and DCP vasculature.[3] The DCP in PAMM eyes has been found to have lower vessel density, suggesting that PAMM could be caused by ischemia of the deep retinal circulation.[2] In the current report, the patient's

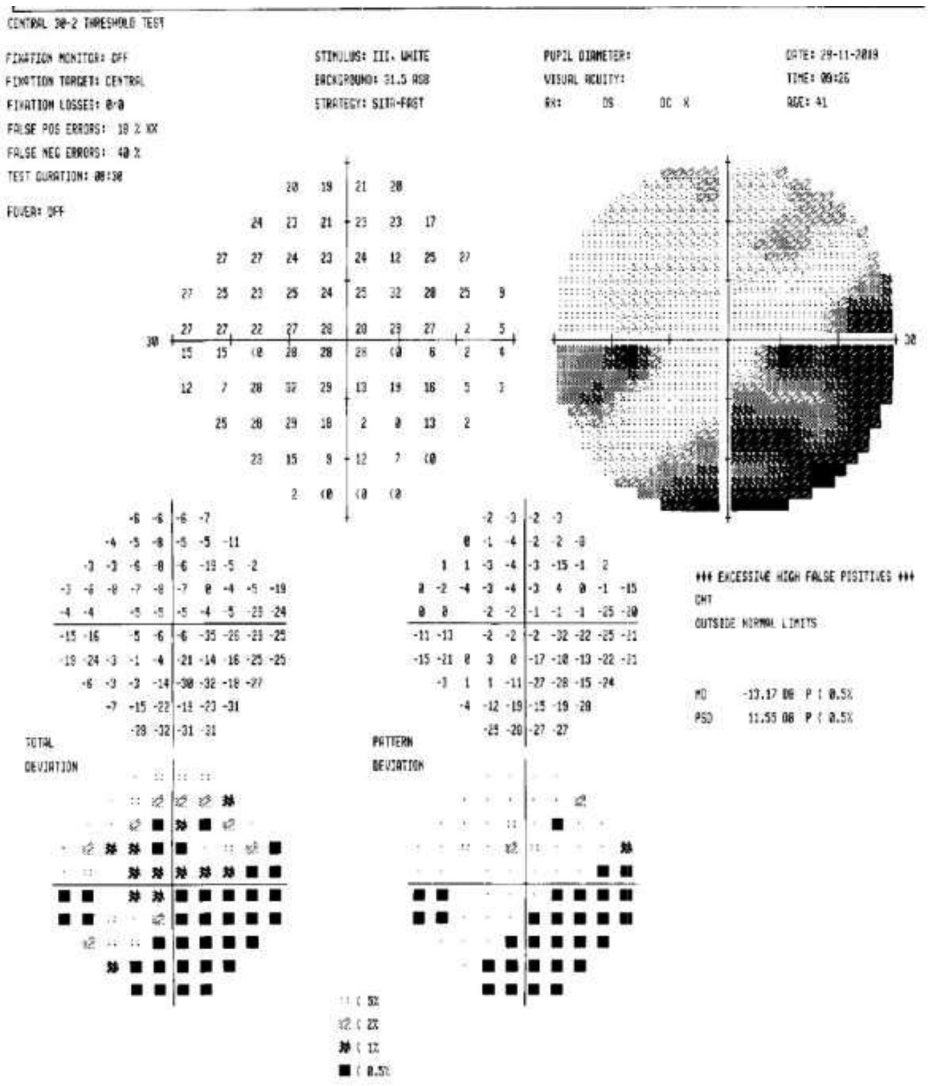

Figure 4. A Humphrey VFT of a patient's LE revealed an inferior altitudinal visual field defect that was correlated with circulatory disturbance in the superior retinal hemifield depicted in FA.

LE had a significantly reduced vascular density in the DCP, which was consistent with previous findings. Reduced oxygen content raises oxygen stress near the superficial retinal layers and choroid, rendering this area more susceptible to ischemia. A high-oxygen intake of horizontal cells also makes the middle retina highly vulnerable to ischemic damage. Paracentral acute middle maculopathy pathogenesis may thus start at the ICP and progress to the DCP via secondary downstream modifications.[1]

A number of ocular and retinal vascular diseases have been associated with PAMM.[4,5] Although this supports the hypothesis that vascular dysfunction plays a role in PAMM, no related ocular diseases were found in our case. Small chronic PAMM lesions have been observed in up to 88% of patients with moderate systemic arterial hypertension and low cardiovascular risk. This may reflect early changes in retinal microcirculation that cause quantifiable changes in retinal vascular density.[6] Furthermore, a PAMM case presenting with sudden unilateral vision loss due to cholesterol embolization syndrome has been reported after endovascular aortic aneurysm repair.[7] The current report, on the other hand, describes how an otherwise healthy individual with PAMM features was diagnosed with a giant aortic aneurysm in the ascending aorta after an extensive systemic etiological analysis. It is believed that thromboembolic events and subsequent LE retinal hypoperfusion could have been triggered by this aneurysm. In addition, vasculopathic risk factors may be associated with PAMM, a disorder that has been reported as a source of visual symptoms in otherwise healthy patients.[5,8,9]

Regardless of the etiology, PAMM lesions often progress to INL atrophy,[5,10] which explains why patients may complain of persistent scotoma.[8] Also, there may be unilateral or bilateral involvement, which is typically accompanied by a rapid onset of single or multiple paracentral scotomas, distorted central vision, or focusing difficulties.[5] Visual acuity may be normal or slightly reduced.[5,10] The average age at presentation is usually between 49 and 53 years old. There is no gender predisposition. An average of eight days of symptoms prior to presentation has been reported.[11] Fundus assessment may be grossly normal. However, in certain cases deeper, smoother, and grayer lesions have been identified.[5] As far as the current report is concerned,

Table 1. Systemic work-up involving radiological and laboratory investigation.

Parameters	Value (Unit)	Normal Limit
Laboratory tests		
Rheumatoid Factor	11.9 (IU/ml)	0–14
Anti-Nuclear Antibody	Negative	0–1000
Activated partial thromboplastin time	28.4 (Sec)	23.6–36.1
Erythrocyte sedimentation Rate	7 (mm/sec)	1–15
Prothrombin time	13.3 (Sec)	12–15.5
Leucocytes	8.59 (10^3/uL)	4–10
Neutrophils	5.5 (10^3/uL)	1.4–6
Lymphocytes	2.9 (10^3/uL)	1.2–4
Monocytes	0.58 (10^3/uL)	0.2–0.8
Erythrocyte	5.5 (10^6/uL)	3.9–6
Hemoglobin	15.8 (g/dL)	12–17
Hematocrit (%)	48.3	34–50
Mean corpuscular volume	87.80 (fL)	80.00–97.00
Mean corpuscular hemoglobin	28.7 (pg)	25–33
Mean corpuscular hemoglobin concentration	32.7 (g/dL)	31.5–36.7
Platelets	182 (10^3/uL)	160–370
Platelet distribution width	11.9 (fL)	10–16
Alanine aminotransferase	9.5 (U/L)	0–41
Aspartate aminotransferase	13.3 (U/L)	0–40
Lactate dehydrogenase	220 (U/L)	135–225
Cholesterol (Total)	186.7 (mg/dL)	60–240
Cholesterol (low-density lipoprotein)	142.3 (mg/dL)	50–150
Radiological analyses		
Diffusion magnetic resonance imaging		Normal
Bilateral contrast orbit magnetic resonance imaging		Normal
Cranial magnetic resonance imaging		Normal
Bilateral carotid Doppler ultrasonography	Vascular lumen width was within normal limits in both common carotid artery and extracranial internal carotid artery and external carotid artery segments that can be tracked. Increased thickness of the intima media in the bilateral common carotid artery. Presence of plaque at bilateral carotid bifurcation leading to <25% stenosis.	
Bilateral vertebral artery Doppler ultrasonography	Right vertebral artery: 58 cm/sec velocity, 95 ml/min flow volume; left vertebral artery: 107 cm/sec velocity, 53 ml/min flow volume.	

a pronounced visual loss in the LE of a 41-year-old male occurred within two days, suggesting a significant hypoperfusion which could have contributed to the ischemia-induced rapid cell death. While ocular movements and anterior segment analysis were normal, color vision was significantly impaired, especially in the LE, implying a potential occult optic nerve ischemia. A slightly pale retinal reflex was also observed in the LE, particularly in the superior hemifield in FA. Additionally, the visual field findings were correlated with circulatory disturbance in the superior retinal hemifield at the DCP level, which could have contributed to the development of PAMM lesions and a much larger INL atrophy.

To rule out other conditions including acute macular neuroretinopathy, giant cell arteritis, and re-perfused central retinal artery occlusion, a PAMM diagnosis should be accompanied by a detailed history and physical examination. Moreover, a detailed ophthalmic examination is often essential for evaluating visual function and identifying signs of other ocular diseases that are commonly associated with aorta aneurism, such as iris floccule,[12] cornea ectasia,[13] and pseudoexfoliation syndrome.[14] Appropriate diagnostic tests, such as a complete blood count, should also be considered.[10] In the current report, a slightly pale retinal reflex during fundoscopy, as well as analyses such as OCT, OCTA, FA, and Humphrey VFT, revealed the presence of PAMM in the LE, as previously stated. All hematological and medical biochemistry parameters were within normal ranges. Most importantly, the authors suggest an individualized diagnostic work-up that takes into account the patient's prior medical history, the presence of cardiovascular risk factors, as well as ocular and systemic

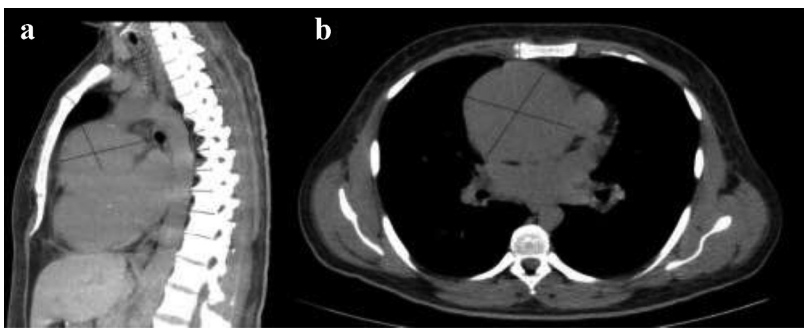

Figure 5. Sagittal (a) and axial (b) views of a thoracic computed tomography scan depicting a giant aortic aneurysm in a patient with PAMM.

findings. In certain patients, no etiology can be determined despite comprehensive diagnostic testing.[5,8–10]

Much remains uncertain about the natural course of PAMM, and no therapy to reverse PAMM has been discovered to date.[15] Long after the initial episode, several other patients develop INL atrophy and symptomatic paracentral scotoma, which may improve with time, but are usually irreversible.[10] Patients with PAMM should be checked for both local and systemic vascular diseases.[16] Identifying and reducing vascular risk factors should be the focus of management. It's also crucial to closely follow-up a PAMM patient since diffuse PAMM lesions may conceal occult retinal occlusions, which can put patient's vision in jeopardy.[5]

Conclusively, PAMM may be the root process of inner retinal ischemia. This report highlights the importance of a comprehensive etiological investigation in patients with PAMM to rule out any potential systemic problems, especially associated cardiovascular diseases. It's also worth noting that multimodal ocular imaging plays an important role in the diagnosis of PAMM.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Informed consent

Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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