PARACENTRAL ACUTE MIDDLE MACULOPATHY: A CHAMELEON MISSED

Nidhi Panwar[1], Prachet Kulakarni[2], Bobby Jose[3]
[1]*Specialist Ophthalmologist, NMC Royal Hospital, Sharjah, United Arab Emirates.*
[2]*Consultant Neurologist, NMC Royal Hospital, Sharjah, United Arab Emirates.*
[3]*Specialist Neurosurgeon NMC Royal Hospital, Sharjah, United Arab Emirates.*
***Corresponding Author: Dr. Prachet Kulakarni,**
Consultant Neurologist, NMC Royal Hospital, Sharjah, United Arab Emirates.

KEYWORDS
central retinal vein occlusion, Paracentral acute middle maculopathy, optical coherence tomography.
Abbreviations: CRVO = central retinal vein occlusion, INL = inner nuclear layer, OPL = outer plexiform layer**,** OCT=optical coherence tomography**,** PAMM = Paracentral acute middle maculopathy**.**

ABSTRACT:
Paracentral acute middle maculopathy (PAMM) represents a type of ischemic maculopathy. These lesions were associated with branch and central retinal arterial occlusion, central retinal vein occlusion (CRVO), diabetic retinopathy, sickle cell retinopathy, and Purtscher retinopathy. Flu-like illness, and transient orbital compression are the symptoms. A typical presentation is sudden onset paracentral scotomas with or without decreased vision, parafoveal greyish-white lesions, and on spectral domain optical coherence tomography (SD-OCT) shows hyperreflective bands localized to the middle layers of the retina, mainly the outer plexiform layer (OPL) and inner nuclear layer (INL). It is painless and sudden, mimicking optic neuritis and ischemic optic neuropathy. We present a patient with this rare disorder with its diagnostic challenges and management strategies.

Introduction

Paracentral acute middle maculopathy (PAMM) is a type of ischemic maculopathy affecting intermediate and deep retinal capillary plexuses. PAMM is characterized by greyish-white parafoveal lesions. Spectral domain optical coherence tomography (SD-OCT) is the diagnostic modality, wherein focal or multifocal lesions are seen as bands of hyperreflectivity in the inner nuclear and outer plexiform layers. The characteristic lesions are due to ischemia of the intermediate and deep capillary plexuses (1).

CASE PRESENTATION

A 45-year-old male, patient came to us with history of loss of vision in the left eye in October 2022 being diagnosed as optic neuritis from referring doctor.

History revealed a sudden presentation of vision impairment in his left eye while finishing his duty at his work place, however he was able to drive with this complaint, consulted an ophthalmologist who diagnosed him as optic neuritis.

He is a middle-aged person on long term medication for obsessive compulsive disorder with antipsychotics and SSRI (Selective serotonin reuptake inhibitors). He is a known hypertensive on medication.

He is non-diabetic. He was treated with intravenous high dose solumedrol for 3 days, followed by oral steroids presuming a diagnosis of optic neuritis pending other investigations.

MRI revealed prominent perioptic hyperintensities on both the sides enhancing with contrast with questionable significance and did not correlate well with the presumptive initial diagnosis of optic neuritis.

He was investigated with MR angiography of the neck vessels to rule out dissection of the neck vessels which was normal. He is a smoker. His Serum antinuclear antibody (ANA), Anti neuromyelitis optica (NMO) antibodies, Anti myelin oligodendrocyte associated antibody (MOGAD antibodies) were also negative, ESR was 14 mm/hr and CRP levels are 6 mg/dL.

Meanwhile a repeat eye consult was done a day after starting therapy.

His eye evaluation revealed that;

Vision Right eye 6/7.5, Left eye 6/38 (did not improve further), no pain on ocular movements, left eye mild RAPD, anterior segment quiet, color vision missed few plates due to central scotoma.
Posterior segment-Right eye unremarkable, Left eye No evidence of disc edema or hyperemia however his macula had mild greyish white appearance.
The clinical symptoms were quite atypical of optic neuritis as it was sudden in onset with no pain on ocular movements.
Ocular Coherence Tomography (OCT) was performed. On the left eye hyper -reflectivity was seen in inner retinal layers (Figure 1).
His visual field showed left eye few paracentral Scotomas.
Fundus Fluorescin Angiography (FFA) did not reveal disc leakage on both the side. There was no gross abnormal fluorescence (Figure 2).
OCT Angiography showed abnormal vasculature in the capillary plexus with multiple, diffuse, fern-like hyper-reflective lesions in retinal layers (Figure 3).
His vision post steroids was only 1 Snellens line better and did not improve much despite full regimen at 2 week.
On the basis of atypical clinical presentation, minimal response with steroid and fundus and OCT angiography pictures, a diagnosis of Left eye Paracentral acute middle maculopathy (PAMM) was made, which happens due to a local ischemic event in the retinal layers.
Going back to the history, a hyperacute involvement fits very well with a diagnosis of PAMM. There is no direct treatment of this condition.
He was thereupon counseled and given topical NSAIDs only without any other active intervention. On follow up, his vision is slowly recovering with fading of scotomas and is now at Snellens 6/15 left eye.

DISCUSSION

PAMM is described as intraretinal ischemia mainly of the intermediate and deep retinal capillary plexuses located in the inner nuclear layer (INL) (2). This pathophysiology of disease is supported by OCT-A findings (3). Since its a relatively rare and unknown entity, it may be frequently missed and patient may be diagnosed as optic neuritis and may be given unnecessary systemic steroids etc.
Acute visual loss is commonly encountered scenario with many diverse etiologies and different management approaches. Awareness of rare conditions and correct diagnosis is important to achieve the final goal of freedom from disease. PAMM is a condition that mimics optic neuritis and ischemic pathologies of the optic nerve.
Such encounters are common where diagnosis remains ambiguous and a specialist ophthalmic perspective should always be sought (4).
Counseling of the patient and risk factor management plays an important part in the management.
Because these lesions are often associated with other vasculopathic conditions, it is important to screen patients presenting with PAMM for both local and systemic diseases, including retinal vessel occlusions, carotid artery disease, diabetes, hypertension, Giant cell arthritis or other vasculitides (5).
Management includes identifying and minimizing vascular risk factors. Careful retinal evaluation in follow up is also important, as these lesions can be harbinger of occult retinal vessel occlusions that further threaten vision (6).
The purpose of this article is to highlight common features of an uncommon and often missed retinal condition and role of retinal imaging including OCT angiography in reaching the diagnosis.

CONCLUSIONS

Paracentral acute middle maculopathy (PAMM) should be kept in mind in a patient with an acute vision loss which can mimic optic neuritis.

Conflict of Interest Statement

The authors report no conflicts of interest.

Informed Consent and Human and Animal Rights Statement

A signed written informed consent taken from the patient. This is a study case report, which was performed according to the principles of safety, ethical attitude and application of the rules of working with patients following the “Bioethical Regulations of the Declaration of Helsinki on the ethical regulation of medical research”.

Acknowledgments
None.
Sources of Funding
No funding was received for this research.
Disclosures
None.
REFERENCES

1. Scharf J, Freund KB, Sadda S, Sarraf D. Paracentral acute middle maculopathy and the organization of the retinal capillary plexuses. Progress in retinal and eye research. 2021 Mar 1;81:100884.
2. Shah A, Rishi P, Chendilnathan C, Kumari S. OCT angiography features of paracentral acute middle maculopathy. Indian J Ophthalmol. 2019;67(3):417-419. doi: 10.4103/ijo.IJO_1249_18. PMID: 30777975; PMCID: PMC6407391.
3. Shah D, Saurabh K, Roy R. Multimodal imaging in paracentral acute middle maculopathy. Indian J Ophthalmol. 2018;66(8):1186-1188. doi: 10.4103/ijo.IJO_173_18. PMID: 30038173; PMCID: PMC6080439.
4. Interlandi E, Pellegrini F, Giuffrè C, Cirone D, Brocca D, Lee AG, Casalino G. Acute-Onset Retinal Conditions Mimicking Acute Optic Neuritis: Overview and Differential Diagnosis. J Clin Med. 2023;12(17):5720. doi: 10.3390/jcm12175720. PMID: 37685787; PMCID: PMC10488521.
5. Colcombe J, Mundae R, Kaiser A, Bijon J, Modi Y. Retinal Findings and Cardiovascular Risk: Prognostic Conditions, Novel Biomarkers, and Emerging Image Analysis Techniques. J Pers Med. 2023;13(11):1564. doi: 10.3390/jpm13111564. PMID: 38003879; PMCID: PMC10672409.
6. Louie, E., Tang, A. & King, B. Paracentral acute middle maculopathy presenting as a sign of impending central retinal artery occlusion: a case report. *BMC Ophthalmol* 23, 268 (2023). https://doi.org/10.1186/s12886-023-02990-6.

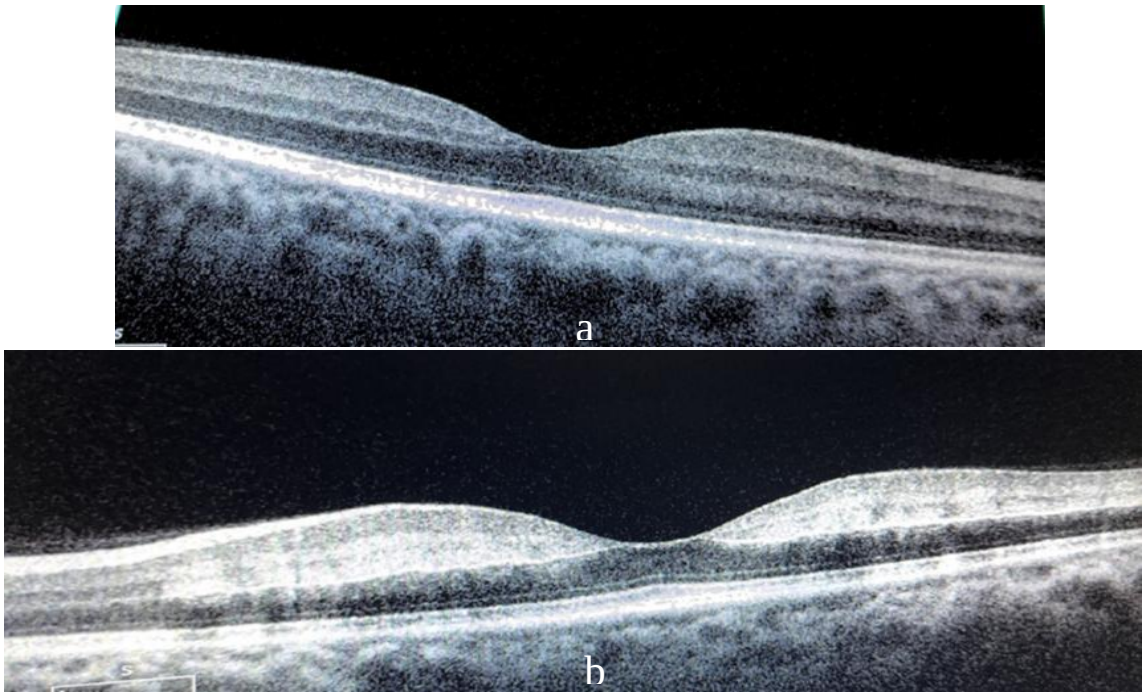

Figure 1: OCT scan of macula. a.Normal Right eye retinal layers. b. Left eye macular scan shows fuzzy and indistinct hyperreflective intra retinal layers.

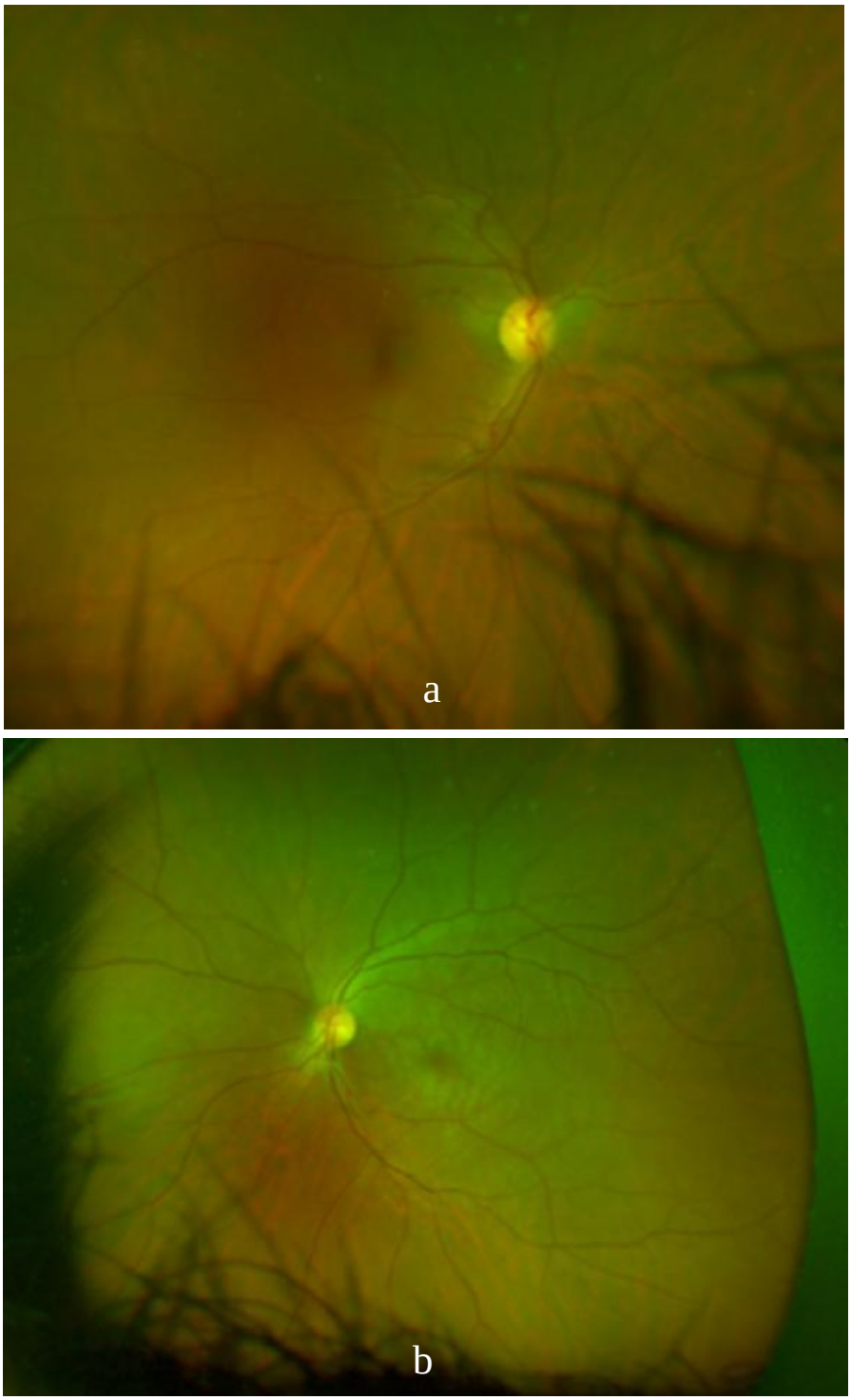

Figure 2. Fundus photograph. a. Fundus right eye grossly normal appearance . b. Fundus left eye- shows edematous, hypopigmentary appearance of macula.

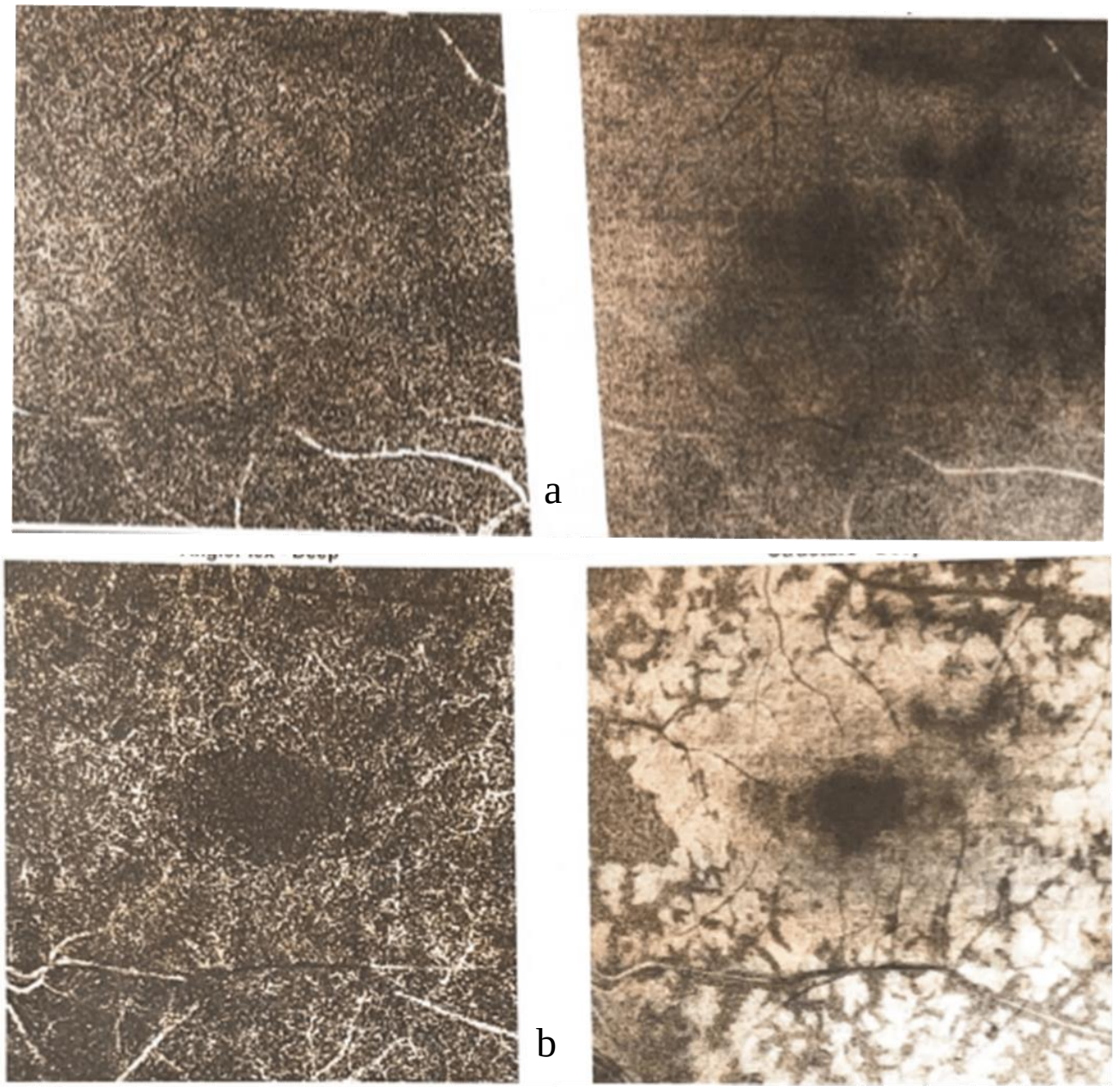

Figure 3. OCT angiography. a. Right eye-normal and unremarkable. b. Left eye - multiple diffuse fern like structures in deep retinal layer.